

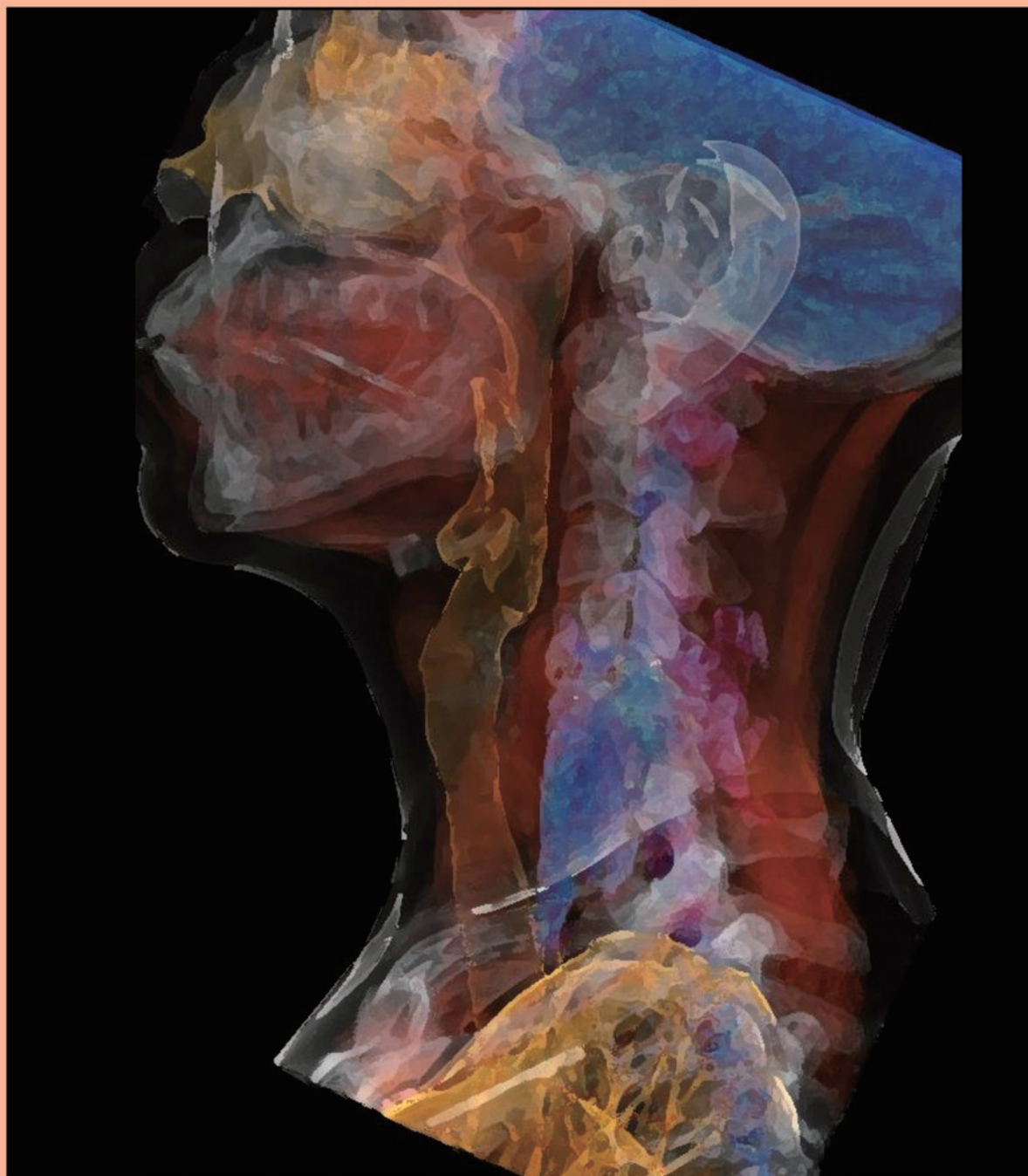


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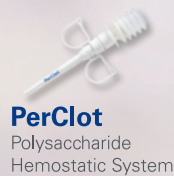


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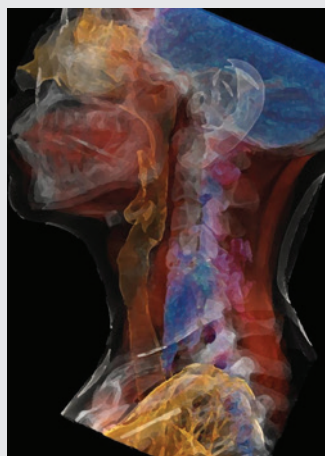
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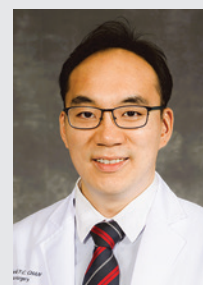
Human Brain, Spine & Cranial Cervical Junction (CCJ)

This is an aesthetic reconstruction of a human brain, spine and the cranial cervical junction (CCJ).

Time is Brain.
Time is Spine.

We are fighting against time to save the Brain and Spine.
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This is the time to remember who we are, where we are coming from, and where we are going.



Dr David YC CHAN

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The Way of the Dragon

Prof Bernard MY CHEUNG

President
The Federation of Medical Societies of Hong Kong



Prof Bernard MY CHEUNG

龍爭虎鬥成過去 風平浪靜迎新歲

Last year was the 50th anniversary of the iconic Bruce Lee (李小龍) film 'Enter the Dragon (龍爭虎鬥)'. The film was scripted and directed by a Hollywood team, but featured an international cast, including future stars such as Jackie Chan (成龍) in minor parts. As the Chinese New Year begins, exit the rabbit and enter the dragon!

In every culture, ancient and modern, the dragon is awesome and powerful. The dragon in China comes from the heavens and Chinese people are said to be descendants of the dragon, presumably carrying its DNA! As is often the case, non-Chinese cultures look at things the opposite way. In the case of the dragon, it is a fearsome monster to be slain by the hero, St George of England, Siegfried in Wagner's Ring or Indra in Hinduism. The dragon has to tread carefully, or risk being stabbed in the underbelly.

During the pandemic, the economy took a tumble and many giants, including the Cathay Dragon, succumbed. However, Napoleon famously said that China was a sleeping giant, which when awake would shake the world. The dragon is resting at present but I am sure will wake up soon.

The crest of the Federation of Medical Societies of Hong Kong shows two dragons, reflecting our two founding members, symbolically guarding our professions. The dragon is therefore our mascot, invoking authority, energy and agility. Like the dragon guarding its treasure, health professionals should be guarding the health of our patients. Under the leadership of our Chief Editor, Dr Raymond Lo, who is the President of the British Medical Association Hong Kong Branch, the Medical Diary in the coming year is sure to be a reliable and authoritative source of updates on the best clinical practice in Hong Kong.

On behalf of the Federation, may I wish all of our readers good fortune and good health in the new year. I am convinced that the Year of the Dragon would be a special year for the Medical Diary, the Federation and the Hong Kong Special Administrative Region. After all, nine dragons live here!

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Editorial

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Dr Michael WY LEE

This issue of the Hong Kong Medical Diary is the conjoint efforts of a group of specialists in neurosurgery who contributed in their areas of interest and subspecialties with regard to the latest advances in neurosurgery. I would like to express my sincere gratitude to all the authors and everyone involved for their hard work and unwavering support.

Hello! Hong Kong

Hong Kong is back to busy as usual. World renowned neurosurgeons Prof Michael Fiehlings from the University of Toronto, Canada and Prof Peng Zhao from the Beijing Tiantan Hospital, Capital Medical University attended in person our 30th Anniversary Scientific Meeting of the Hong Kong Neurosurgical Society on 17th and 18th November 2023. Our theme was "Spine & Craniocervical Junction". In addition to Time is Brain, we learnt that *Time is Spine*. Spinal diseases are definitely a prevalent primary health care concern, particularly for Hong Kong which is a place amongst the highest longevity counterparts in the world.

In the review and update on the management of degenerative spine, basic knowledge and various management approaches, including minimal invasive surgery (MIS), are described. Promising direction and development include early detection, monitoring, rehabilitation, and neuroprotection of the disease using microRNAs biomarkers, advanced MRI techniques, transcranial magnetic stimulation (TMS), as well as the sodium channel blocking anticonvulsant Riluzole.

For the management of recurrent dumbbell spinal

neurogenic tumours, the Triple F approach - *Facetectomy, Fusion and Function preservation* is advocated. Illustrative steps are distinctly spelled out and multi-disciplinary care should be adopted to preserve and restore neurological functions.

Spinal arteriovenous fistulae are great mimickers of degenerative spinal diseases as they can cause similar perplexing clinical features. Vigilance, early detection and prompt proper management, with both open approach or endovascular intervention, can give excellent neurological outcomes.

The cranio-cervical junction was a place difficult to approach without significant morbidities and mortalities. With the latest advent of endoscopes with improved image quality and neurosurgical tools, it is now possible to have a fine dissection of the tumours next to the cranial nerves and brainstem and achieve excellent clinical outcomes.

Besides being one of the commonest intracranial tumours, the pituitary tumour is also one of the commonest skull base tumours that are most effectively treated by the endoscope. It pays to study the latest 5th Edition of the WHO classification of Endocrine and Neuroendocrine Tumour (ENDO5) that was adopted and published in 2022. A neurosurgical perspective on the new WHO classification for pituitary tumour is absolutely timely.

Last but not least, Dr Teresa Tse, a mother of two kids while training in neurosurgery, introduces to us the life of Women in Neurosurgery and how to strike a fine balance between family, profession as well as personal hobbies.



Use of Endoscope in Craniocervical Junction

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INTRODUCTION

The craniocervical junction is a complex anatomical region that serves as the interface between the skull and the cervical spine. Pathologies affecting this region, such as tumours, congenital abnormalities, trauma, and infections, often require surgical intervention. Traditional open surgical approaches, such as transoral or transcranial procedures, have conventionally been employed to access and treat pathologies in this region. However, these approaches are associated with significant morbidity, including long surgery time, long hospital stays, postoperative pain, and potential complications. Surgeries are often done through a deep narrow corridor with very limited illumination. In recent years, endoscopic techniques have gained attention as a less invasive alternative for managing craniocervical junction pathologies.

ENDOSCOPIC TECHNIQUES IN CRANIOCERVICAL JUNCTION PATHOLOGIES

Endoscopic approaches in the craniocervical junction utilise thin rigid endoscopes to visualise and access the region. Recent advance of technology in neuroendoscopy extended its application in neurosurgical procedures. One of the use was in the skull base and the craniocervical junction. The initial application was for use in endonasal transsphenoidal surgery at the University of Pittsburgh¹. With the initial success, the application subsequently extended to parasellar region, anterior skull base and eventually to the craniocervical region².

The endonasal endoscopic approach involves accessing the craniocervical junction through the nasal cavity. This technique, also known as the expanded endonasal approach (EEA), provides direct access to the ventral aspect of the craniocervical junction, allowing for the removal of tumours, decompression of neural structures, and repair of cerebrospinal fluid leaks. It avoids the need for facial incisions or brain retraction, resulting in reduced postoperative pain, shorter hospital stays, and faster recovery. The lower limit of the approach is defined by the nasopalatine line.

The transoral approach had been utilised to assess the craniocervical region. It is particularly useful for treating odontoid pathologies to allow direct decompression of neural structures. Common pathologies include basilar invagination and odontoid fractures. The

shortcoming is, surgery was done through a deep, long corridor and the soft palate was frequently split for better exposure. The application of an endoscope in the approach improved the visualisation of the region and also reduced the need for splitting of the soft palate. Approach related complications were reduced with the use of an endoscope in transoral surgery for the craniocervical junction.

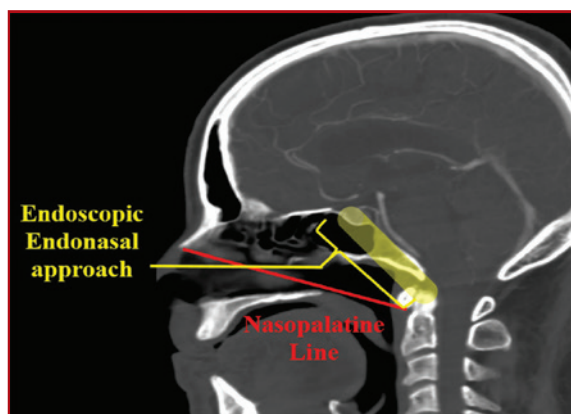


Fig. 1: Sagittal image depicting extent of approach from the nose using rigid instruments from sella to craniocervical junction. The red nasopalatine line depicts the lower limit of the approach through the nose. This resulted in the use of the endoscope in the transoral approach. (Personal collection)

BENEFITS AND LIMITATIONS OF ENDOSCOPY IN CRANIOCERVICAL JUNCTION PATHOLOGIES

Endoscopic techniques offer several advantages over traditional open surgical approaches in the management of craniocervical junction pathologies. These include reduced tissue trauma, shortened hospital stays, decreased postoperative pain, and faster recovery. Endoscopic procedures also provide enhanced visualisation of critical structures, allowing for precise surgical maneuvers.

However, endoscopy in the craniocervical junction is not without limitations. The limited working space and the need for specialised instruments and training pose challenges to surgeons. Additionally, the learning curve associated with endoscopic techniques may result in longer operative times initially. Patient selection



is crucial, as not all cases are amenable to endoscopic approaches, particularly those with extensive pathology or significant anatomical variations.

EFFICACY OF ENDOSCOPIC TECHNIQUES IN CRANIOCERVICAL JUNCTION PATHOLOGIES

Numerous studies have demonstrated the efficacy and safety of endoscopic techniques in the management of craniocervical junction pathologies. The multicentre study reported successful endoscopic excision allowed adequate decompression of pathologies, with low complication rates and favourable outcomes³. Systematic reviews concluded that endoscopic approaches are effective and minimally invasive for treating basilar invagination, with satisfactory clinical and radiographic outcomes⁴.

THE USE OF ENDOSCOPIC TECHNIQUES IN THE MANAGEMENT OF SPECIFIC CRANIOCERVICAL JUNCTION PATHOLOGIES

1. Basilar Invagination

Basilar invagination refers to the cranial migration of the odontoid process into the foramen magnum, resulting in compression of the brainstem and upper cervical spinal cord. Endoscopic techniques have shown promise in the treatment of basilar invagination. The transoral endoscopic approach allows for direct visualisation and decompression of the neural structures. It enables the resection or reduction of the odontoid process, alleviating compression and restoring normal anatomical alignment. The minimally invasive nature of endoscopy reduces postoperative morbidity and facilitates faster recovery compared to traditional open surgical approaches. After ventral decompression, posterior fixation would be needed for stabilisation of the craniocervical junction.

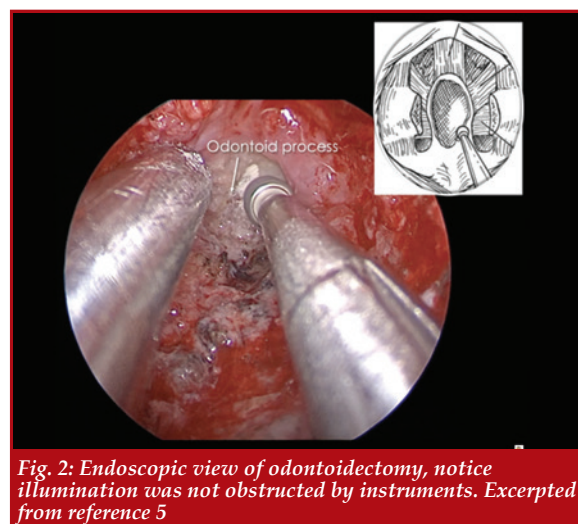


Fig. 2: Endoscopic view of odontoidectomy, notice illumination was not obstructed by instruments. Excerpted from reference 5



Fig. 3: Microscopic view of odontoid during transoral odontoidectomy, note the visualisation was limited by the deep narrow corridor, which require two retractors for one instrument to be used. Excerpted from reference 6

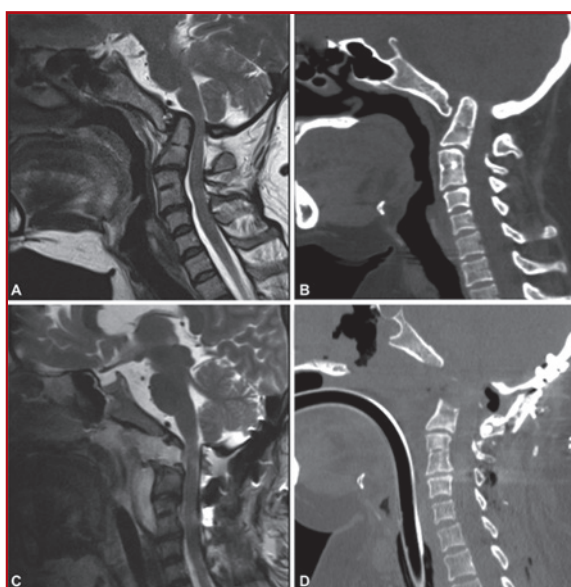


Fig. 4: MRI and CT showing pre-op imaging of a patient with basilar invagination and after endoscopic odontoidectomy and posterior fixation. Excerpted from reference 5

2. Tumours

Endoscopic approaches have been successfully employed in the resection of craniocervical junction tumours. These techniques provide excellent visualisation and access to the ventral aspects of the craniocervical junction, which enabled safe tumour removal while preserving critical neural structures. Tumours in the region include chordoma and chondrosarcoma. Studies have reported improved extent of excision and low complication rates with endoscopic tumour resection⁷.

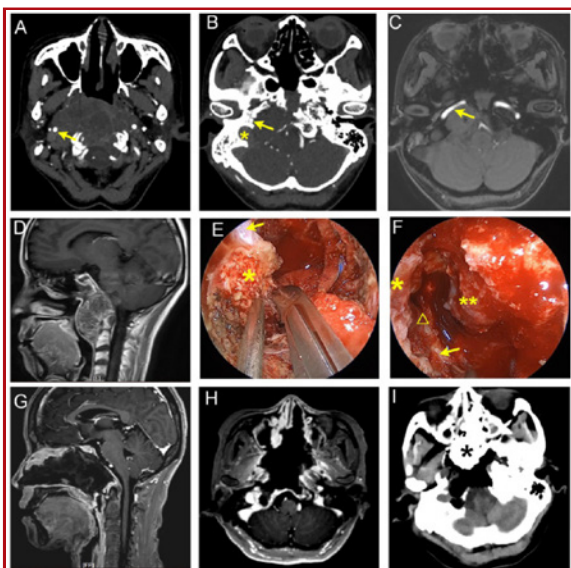


Fig. 5: Giant recurrent chordoma involving the lower clivus. The tumour shows proximity to the internal carotid artery (arrow), and the jugular foramen (asterisk in B). Endoscopic view (E and F) allows excellent visualisation of the region and allows dissection of tumours from critical structures. Postoperative images (G, H, I) showed the extent of removal of the tumour. (asterisk in image I was iodoform gauze for nasal packing). Excerpted from reference 8

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3. Post-Radiotherapy Radionecrosis

Post-radiotherapy radionecrosis is a potential complication following radiation therapy for head and neck cancers, especially nasopharyngeal carcinoma in our locality. It can lead to tissue fibrosis, vascular damage, and radiation-induced necrosis in the craniocervical junction. Radionecrosis would often be complicated with infection due to breakage of overlying mucosa and exposure of underlying bone to the bacteria in the aerodigestive tract. Endoscopic techniques, particularly the endonasal endoscopic approach, can play a role in the management of post-radiotherapy radionecrosis. The endoscope allows for precise visualisation and removal of infected and necrotic tissue, facilitating debridement and promoting wound healing. In some cases, reconstructive procedures, such as nasoseptal flap repair, can be performed using the endoscopic approach to aid in the healing process.

CONCLUSION

Endoscopic techniques have emerged as valuable alternatives in the management of craniocervical junction pathologies. These minimally invasive approaches provide direct visualisation and access to the pathology, allowing for precise surgical manoeuvres while reducing tissue trauma and postoperative morbidity. The benefits of endoscopy include better visualisation, less approach related morbidities, shorter hospital stays, and decreased postoperative pain. However, appropriate patient selection and surgical expertise are crucial for optimal outcomes. Further technical advancements in endoscopy would likely expand the applications of endoscopy in the management of craniocervical junction pathologies.

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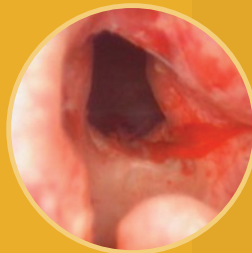
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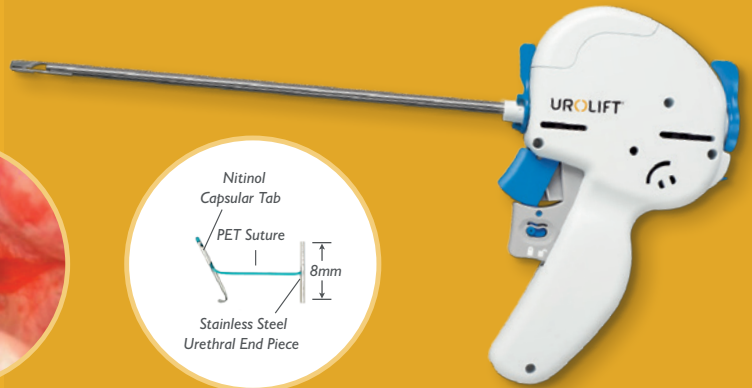
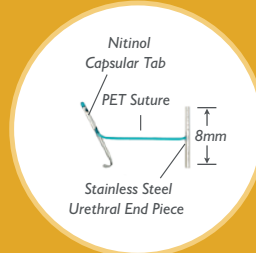
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Indicated for the treatment of symptoms of an enlarged prostate up to 100cc in men 50 years or older. As with any medical procedure, individual results may vary. Most common side effects are temporary and include hematuria, dysuria, micturition urgency, pelvic pain, and urge incontinence.⁹ Rare side effects, including bleeding and infection, may lead to a serious outcome and may require intervention. Consult the Instructions for Use (IFU) for more information.

*No instances of new, sustained erectile or ejaculatory dysfunction in the LIFT pivotal study.

1. Sønksen, 2015 Eur Urol, BPH Study; 2. Roehrborn, 2015 Can J Urol, 5 yr results of PUL LIFT study; 3. Roehrborn, Can J Urol 2017 LIFT Study; 4. AUA BPH Guidelines 2003, 2020; 5. Naspro, Eur Urol 2009; 6. Montorsi, J Urol 2008; 7. McVary, J Sex Med 2016; 8. Roehrborn, Can J Urol 2017; 9. Roehrborn J Urology 2013 LIFT Study; 10. Roehrborn et al. Can J Urol 2017; 11. Shore Can J Urol 2014; 12. Bachmann, European Urol 2013; 13. Mollengarden, Prostate Cancer Prostatic Dis 2018; 14. Gilling, J Urol 2017.

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Challenges in Recurrent Dumbbell Spinal Neurogenic Tumours and Treatment Strategy

Dr David YC CHAN

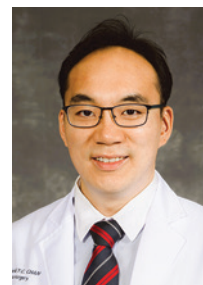
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INTRODUCTION

Dumbbell spinal neurogenic tumour is a Neurosurgical challenge. The majority of the Dumbbell spinal tumours are slow-growing and benign. At the same time, there are risks of tumour progression with cord compression and neurological deficits. Dumbbell spinal tumours can even progress with bony erosion affecting the facet joints, pedicles or the vertebral bodies. (Fig. 1) Depending on the surgical goals, there is a spectrum of treatment options. This clinical challenge required multi-disciplinary care involving different surgical subspecialties. From the neurosurgical point of view, the key surgical goal is to preserve and restore neurological function. Treatment strategies will be discussed in this article, ranging from 1) observation with close monitoring, 2) MIS Laminotomy or Hemi-laminectomy for decompression, with or without radiotherapy, to 3) a more durable and potentially curative strategy with the Triple-F (3F) approach: Facetectomy, Fusion and Function preservation.

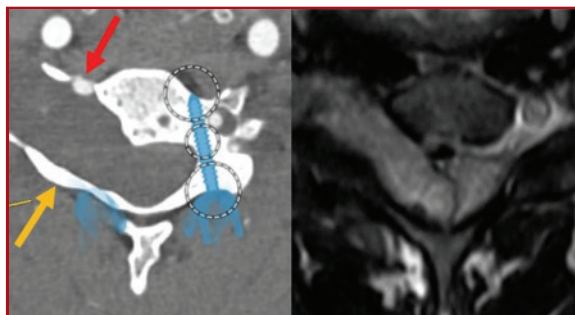


Fig. 1: Axial CT C-spine with CT Angiogram (left) and contrast MRI (right) showing a cervical Dumbbell neurogenic spinal tumour with bony erosion of the ipsilateral facet joint and lateral mass (orange arrow), as well as the pedicle and the vertebral body. The vertebral artery (red arrow) was also displaced. An illustrative pedicle screw (blue) is planned on the contralateral side for fusion. (Personal collection)

BACKGROUND

Neurogenic spinal tumours are nerve sheath tumours such as schwannomas, as well as neurofibromas. Schwannomas are composed of neoplastic Schwann cells¹. The "classic" pathology of a schwannoma included the Antoni A and Antoni B histological patterns². For neurofibromas, unlike schwannomas, they have additional components, such as fibroblasts,

perineurial cells, mast cells and residual axons¹. For the dumbbell spinal neurogenic tumours, they can be intradural, with foraminal and extraforaminal extradural components. They can also be completely extradural³.

Most spinal schwannomas are potential curative when gross total resection can be achieved⁴. Dumbbell schwannomas had shown to have significantly higher risks of recurrence, requiring re-operations⁵. The dumbbell spinal tumours, including intradural extramedullary with extradural extension were independent risk factors for recurrence⁶.

CLOSE MONITORING/OBSERVATION

The majority of the Dumbbell spinal tumours are slow-growing and benign⁷. For asymptomatic patients with no red flag signs or symptoms, close monitoring might be an option, especially if the spinal tumour is incidentally diagnosed upon workup for other conditions. At the same time, clinicians ought to warn the patients of the potential risks of tumour progression and neurological deficits. Moreover, biopsy or additional imaging (such as PET-CT) or other workup shall be offered to rule out malignant transformation or metastasis. Surgical treatment should be offered if the patient is symptomatic. In general, for most benign spinal schwannoma, the growth rate is around 5 % increase in the tumour volume per year⁸. Medical advice should be given for patients to re-attend if there is any deterioration or any red-flag signs or symptoms arise.

LAMINOTOMY/ HEMI-LAMINECTOMY (FACET-SPARING) FOR DECOMPRESSION AND TUMOUR EXCISION

Hemi-laminectomy or Laminotomy for excision of the intradural-extramedullary spinal tumour can achieve the surgical goal of decompression and obtaining specimens for the histological diagnosis⁹. The operation can be performed as 1) open operation, 2) mini-open, 3) minimally invasive surgery (MIS) microscopic operation, or even 4) endoscopic in selected patients¹⁰. Laminotomy is a safe and effective surgical treatment option for most dumbbell spinal tumours, especially during the first operation. Spinal stability can also be preserved with this facet-sparing surgical approach¹¹.



On the other hand, this facet-sparing approach with posterior decompression alone potentially had higher risks of recurrence, as compared to the radical surgery with facetectomy and gross total resection¹². In one cohort, the median time interval for re-operation is 3.5 years after Laminotomy¹³. (Fig. 2)

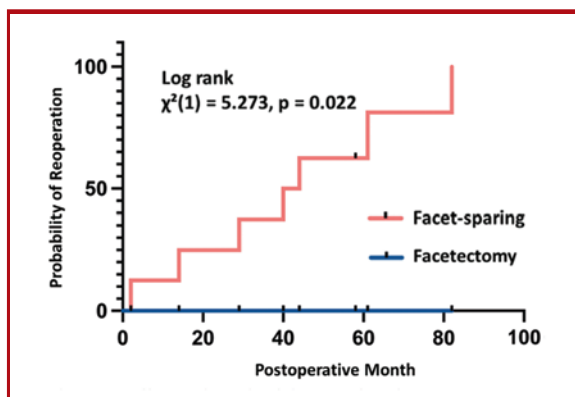


Fig. 2: Illustration cited from Schwake M, Maragno E, Gallus M, et al. Minimally Invasive Facetectomy and Fusion for Resection of Extensive Dumbbell Tumours in the Lumbar Spine. *Med.* 2022;58(11). Cumulative risk of re-operation is shown. The facet-sparing group was in the red line and the facetectomy group was in blue. Kaplan-Meier analysis showed a significantly higher re-operation risk in the facet-sparing group ($p = 0.022$). Reference: Schwake M, Maragno E, Gallus M, et al. Minimally Invasive Facetectomy and Fusion for Resection of Extensive Dumbbell Tumours in the Lumbar Spine. *Med.* 2022;58¹¹.

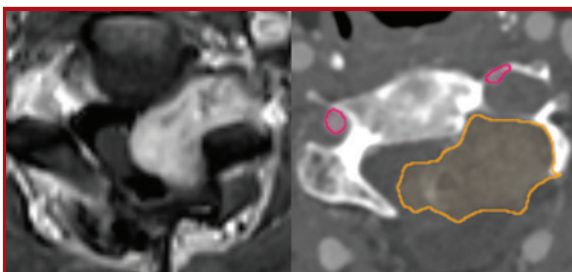


Fig. 3: Axial contrast T1 MRI (left) and CTA (right) of recurrent cervical dumbbell neurogenic spinal tumours 3 years after Laminotomy (facet-sparing). The recurrent dumbbell spinal tumour is herniating out from the old laminotomy site and the spinal cord is compressed. (Personal collection)

ENDOSCOPIC OPERATIONS

In well-selected patients without spinal instability, concomitant endoscopic operations can be considered. Thoracoscopic operations can be considered for thoracic dumbbell spinal tumours¹⁴. For lumbar dumbbell tumours, laparoscopic operations and retroperitoneal operations can be considered¹⁵. Robotic procedures had been reported in selected patients¹⁶. This approach is facet-sparing and can be considered in dumbbell spinal tumours without significant facet bony erosion. From the neurosurgical point of view, the key surgical goal is to preserve neurological function and to decompress the spinal cord. Water-tight dural closure is crucial to avoid leakage of cerebrospinal fluid (CSF) into the thoracic or retroperitoneal cavity. For dumbbell spinal

tumour with intradural extension, thoracoscopic or laparoscopic operations might have a risk of tension pneumocephalus¹⁷.

TRIPLE F (3F) APPROACH: FACECTOMY, FUSION and FUNCTION PRESERVATION.

In recurrent dumbbell neurogenic spinal tumours, the surgical challenge and the potential considerations include 1) inadvertent dural tear or spinal cord injury during incision and subperiosteal dissection, due to the lack of normal lamina or other bony landmarks at the index level (Fig. 3), 2) difficulty in identifying the normal dura, if any, at the old laminotomy site, as recurrent dumbbell tumours can re-grow with intradural and extradural components, within the spinal canal and the neuro-foramen at the dural sleeve of the spinal nerve roots⁵, 3) lack of a clear dissection plane at the interface between the spinal cord and the recurrent tumour, due to arachnoid adhesions or scarring, 4) eroded facet joint with distorted bony anatomy during subperiosteal dissection at the lateral margin, 5) close proximity to vascular structures, such as the vertebral artery (VA), or the aorta, with risks of vascular injury, 6) planning of the lateral mass screws or pedicle screws, due to bony erosion of the lateral mass or the pedicles by the recurrent dumbbell tumour¹⁸, 7) instability and the extent of the fusion, ranging from 1 level above and below, to 2 levels or even 3 levels above and below the index level, 8) pedicle screws insertion at the ipsilateral side only, or insertion at the contralateral side (bear in mind the contralateral VA can be the dominant one supplying the brainstem, as the ipsilateral VA is usually compressed by the giant dumbbell tumour, 9) deformity correction and restoration of alignment (Fig. 4), 10) water-tight dural closure and prevention of CSF leak, especially at the dural sleeves at the neuro-foramen junction with dural erosion by the recurrent dumbbell tumours, 11) the sequence of procedures, whether to perform fusion first or tumour excision and dural reconstruction first.

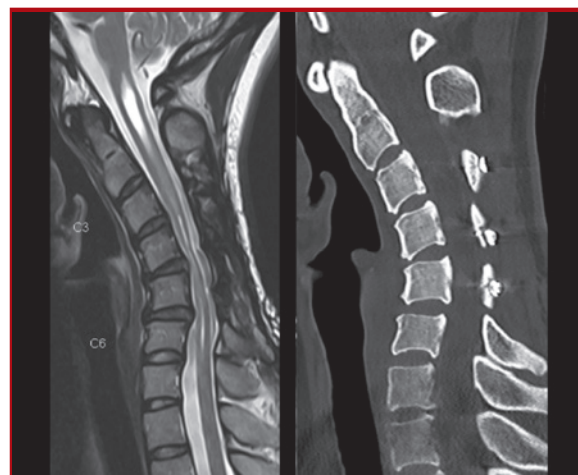


Fig. 4: Sagittal T2 MRI (left) and plain CT C-spine (right) illustrating the long-term potential risk of kyphotic deformity after multi-level laminoplasty for cervical intradural spinal tumour excision without fusion. (Personal collection)

Illustrative steps of the proposed 3F Surgical Methods

Intraoperative monitoring and positioning

- Step 1: Expose the Lateral mass, the Facet, and the Lamina Edge above and below the index level.
- Step 2: Lateral mass screws and/or pedicle screws insertions, above and below the index level. (Fig. 5) Rods insertion at the contralateral side first, if needed.
- Step 3: Facetectomy¹⁹ (ipsilateral side)
- Step 4: T-shaped dural opening, opening up the dural sleeve.
- Step 5: Tumour excision, both the foraminal and intraspinal parts²⁰.
- Step 6: Protect the spinal cord and dissection along the tumour-cord interface.
- Step 7: Protect and delineate the VA, such as by intraoperative Doppler and/or indocyanine green (ICG).
- Step 8: Water-tight dural closure and dural reconstruction.
- Step 9: Decortication (after dural closure) and insertion of the rod at the ipsilateral side and bone graft.

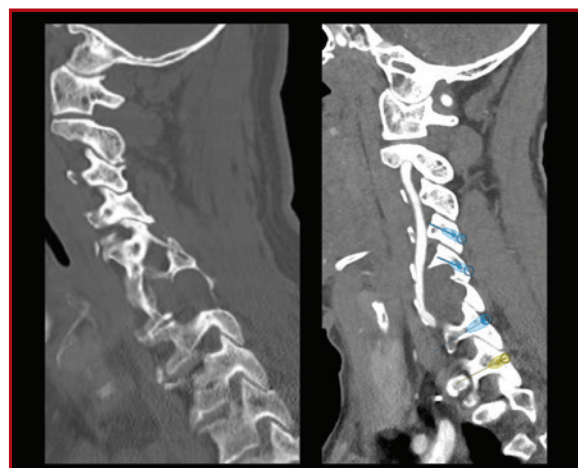


Fig. 5: Sagittal plain CT C-spine (left) and CTA illustrating dumbbell spinal tumours with facet joint erosion and subluxation. Illustrative surgical preparation for Fusion with lateral mass screws and pedicle screws are shown. Be mindful of VA injury during tumour resection or pedicle screw insertion. (Personal collection)

ADJUVANT THERAPY: RADIOTHERAPY

Radiotherapy and radiosurgery can be considered in selected patients with intradural spinal tumours. Good long-term outcomes have been reported in a well-selected group of patients²¹. On the other hand, radiation toxicity might occur, such as radiation myelopathy²², or hemi-cord syndrome²³.

CONCLUSION

The clinical challenge with Recurrent Dumbbell Spinal

Tumours required multi-disciplinary care involving different surgical sub-specialties. From the neurosurgical point of view, the key surgical goal is to preserve and restore neurological functions.

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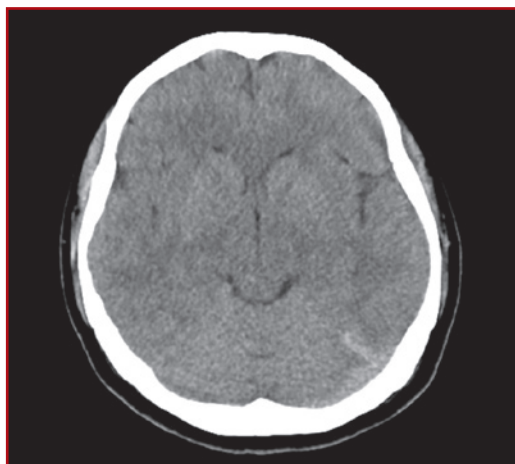
Radiology Quiz

Dr Carol PY CHIEN

MBBS, FRCR, FHKCR, FHKAM (Radiology)



Dr Carol PY CHIEN



A 50-year-old woman with good past health presented to the Accident & Emergency Department with symptoms of headache and vomiting. Limb power and sensation were unremarkable. There was no history of trauma. CT brain was performed.

Questions

1. What are the CT brain findings?
2. What is the differential diagnosis of this CT brain?
3. What are the common risk factors for this condition?
4. What are the important complications to be aware of?
5. What is the next step of the investigation?
6. What is the next step of management?

(See P.36 for answers)

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CHF: congestive heart failure; SIADH: syndrome of inappropriate antidiuretic hormone secretion.

Reference:

Abbreviated Prescribing Information

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A Review and Update on Management of Degenerative Cervical Spine Disease

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This article has been selected by the Editorial Board of the Hong Kong Medical Diary for participants in the CME programme of the Medical Council of Hong Kong (MCHK) to complete the following self-assessment questions in order to be awarded 1 CME credit under the programme upon returning the completed answer sheet to the Federation Secretariat on or before 29 February 2024.

INTRODUCTION

Degenerative cervical spine disease is a complex, multifaceted health concern, affecting a significant proportion of the population, particularly those of middle to older ages. It encompasses a spectrum of pathological changes within the cervical spine due to age-related degeneration and wear-and-tear. The rising prevalence of this disease can be attributed to several factors, including an ageing population and shifts in lifestyle patterns. Factors such as prolonged use of mobile phones or computers, poor posture, and lack of physical activity, all contribute to the development and progression of these degenerative changes.

Early and effective detection and management play a pivotal role in preventing long-term complications and improving the quality of life for patients. The objective of this article is to provide an overview of the recent developments in various aspects such as pathophysiology, clinical symptoms, differential diagnosis, and treatment options for degenerative cervical spine disease.

PATHOGENESIS

Degenerative changes in the cervical spine often begin at the intervertebral disc. With age, the disc reduces its water content and the nucleus pulposus becomes fibrosis. The disc becomes less compliant and loses its ability to distribute pressures over endplates equally. It leads to bone remodelling at the endplates, osteophyte formation, and structural changes in the vertebrae. Subsequently, ligamentous changes, including ligamentous hypertrophy or ossification can also occur in response to these degenerative changes, potentially resulting in spinal canal narrowing and spinal nerve root or spinal cord compression.

The static and dynamic compression of the spinal nerve root or spinal cord may lead to radiculopathy and myelopathy, respectively. On a microvascular level, compression can lead to ischemia and neuroinflammation, triggering an apoptotic pathway that results in progressive neuronal and oligodendroglia cell death¹. This explains the subsequent disease progression and irreversible neurological recovery in advanced cases.

Despite the controversy surrounding the attribution of axial neck pain to these degenerative changes, in general, it is believed nerve fibres and unencapsulated nerve endings in these structures make discs and facets potential pain generators.

Besides radicular pain, axial neck pain from a degenerative cervical spine can be debilitating. The pathophysiology of axial neck pain is even more complex, and many controversies exist. However, it is often believed that the discs and facets are potential pain generators. The axial neck pain begins when tissues are irritated by facet deterioration and disc degeneration. Then, the irritation activates nociceptors around these structures. Inflammation follows, and a vicious circle arises with inflammation, leading to even more pain². In addition, muscle spasms often occur as the neck is repositioned to avoid pain or because the pain stimulus directly causes the involuntary muscle contraction.

CLINICAL MANIFESTATIONS

Patients with degenerative cervical spine disease can present a range of symptoms, varying in both severity and duration. The most common symptom is neck pain, which can range from dull and achy to sharp and stabbing. This pain may be localised to the neck or radiate to the shoulders, arms, or even the fingers, often exacerbated by movements or prolonged periods of sitting or standing.

In addition to neck pain, patients may experience radicular symptoms, such as radiating arm pain, numbness, tingling, or weakness. These symptoms typically follow a specific dermatomal pattern corresponding to the compressed or irritated nerve roots.

Patients may also experience symptoms of degenerative cervical myelopathy, including upper and lower limb muscle weakness, paraesthesia, urinary incontinence, and gait disturbance.

While many degenerative cervical spine diseases are not acutely serious and gradual in progression, there are instances where urgent evaluation or intervention may be necessary. The following "red flags" may suggest more severe pathology and warrant prompt referral to a neurosurgeon or spine surgeon for further evaluation



and management:

- Progressive neurological deficits, including increasing weakness, loss of coordination, or bowel and bladder dysfunction
- Severe or worsening neck pain that is unresponsive to conservative measures
- Symptoms suggestive of cervical myelopathy, such as gait disturbances, difficulty with fine motor skills, or loss of balance
- History of trauma, especially high-energy or significant mechanism of injury
- Unexplained fever or suspected infection
- Unexplained weight loss or suspected of malignancy

DIAGNOSTIC TESTS

Radiological imaging plays a crucial role in the diagnosis and evaluation of degenerative cervical spine. Simple radiography can reveal disc space narrowing, sclerosis, osteophytes at the involved endplate, uncovertebral and facet joint hypertrophy, and overall sagittal alignment and balance.

Computed tomography (CT) scans provide more detailed images of the bony structures, particularly in cases of suspected fractures or when evaluating the integrity of facet joints. It is also a good choice of imaging in evaluating the ossification of the posterior longitudinal ligamentum.

Magnetic resonance imaging (MRI) is the gold standard for assessing soft tissue structures, including intervertebral discs, nerve roots, and the spinal cord. It can identify disc herniations, spinal stenosis, cord compression, hypertrophy of ligamentum flavum, and other pathologies. Numerous research findings have indicated a relationship between alterations in spinal cord signals and the severity of cervical myelopathy. Specifically, decreased signal intensity at T1 and increased signal intensity at T2-weighted images have been associated with poorer surgical outcomes.

Electromyography (EMG) and nerve conduction studies (NCS) are helpful in evaluating the electrical activity of muscles and nerves, respectively, and can provide further diagnostic insight into degenerative cervical spine disease.

CONSERVATIVE MANAGEMENT

The first-line approach to managing degenerative cervical spine disease typically involves conservative treatment options aimed at relieving symptoms and improving the patient's quality of life.

Pain Management

Effective pain management is a crucial aspect of treating degenerative cervical spine disease. Multimodal agents, such as acetaminophen, nonsteroidal anti-inflammatory drugs (NSAIDs), gabapentin, and mild opioid analgesics, are commonly prescribed to alleviate pain

and reduce inflammation. In some cases, short-term use of muscle relaxants or low-dose tricyclic antidepressants may be considered for managing muscle spasms or neuropathic pain³.

Physical Therapy

Structured physical therapy programmes can enhance range of motion, strengthen neck and shoulder muscles, and improve posture and body mechanics. Therapeutic exercises, manual therapy techniques, traction, ultrasound, transcutaneous electrical nerve stimulation (TENS), thermotherapy, and modalities like heat or ice may be utilised to reduce pain and improve function. Home exercises are also an important aspect of treatment, aiding in pain reduction, strengthening neck muscles, and maintaining cervical range of motion⁴.

Activity Modification

Patients are often encouraged to modify activities that exacerbate their symptoms, helping to prevent further strain on the cervical spine. Adjusting workstation ergonomics, avoiding heavy lifting or repetitive motions, and maintaining proper posture during daily activities are recommended.

Cervical Collars and Pain Injections

Short-term use of a cervical collar may provide support, limit motion, and alleviate symptoms. However, long-term immobilisation with a cervical collar is generally discouraged due to potential muscle weakness and further deconditioning⁵.

Local Injections

For patients with severe or refractory radicular pain, epidural steroid injections or selective nerve root blocks may be considered. These injections deliver corticosteroids and local anaesthetics directly to the affected nerve roots, providing temporary relief and reducing inflammation.

NATURAL HISTORY AND DISEASE PROGRESSION

Most patients with axial neck pain or cervical radiculopathy have a favourable prognosis following conservative management. A study demonstrated that 31.7 % of patients with symptomatic radiculopathy had symptoms recurred, and 26 % needed surgical intervention later⁶.

On the other hand, it is important to note that patients with canal stenosis or non-myelopathic compression of the spinal cord are at risk of developing degenerative cervical myelopathy (DCM). Research indicates that approximately 8 % of these patients develop DCM at a 1-year follow-up, with this figure increasing to 23 % at a median of 44-month follow-up⁷.

SURGICAL INTERVENTIONS

If a patient does not respond to conservative treatment or there is evidence of severe neurological compromise, surgical intervention may be warranted. The choice of surgery depends on several factors, including the patient's age, comorbidities, the type and site of the compression, the alignment of the spine, the state of the adjacent levels, bone density, and the surgeon's preference.

Anterior Approach

An anterior approach provides direct access to the disc, allowing for better control of the decompression and reconstruction of the height and the segmental lordosis of the disc. It entails procedures like anterior cervical discectomy and fusion (ACDF), anterior cervical corpectomy and fusion (ACCF), and artificial disc replacement (ADR).

Anterior Cervical Discectomy and Fusion (ACDF)

ACDF is a frequently performed anterior cervical surgery. The procedure involves the removal of the offending disc from an anterior approach, followed by the placement of a bone graft in the empty disc space for bony fusion (Fig. 1). The fusion process aims to provide long-term stability and prevent pathological motion that might perpetuate neurological impingement. ACDF is generally associated with high success rates in alleviating symptoms, although it might lead to a reduced range of motion and potential adjacent segment disease.



Fig. 1: A 55-year-old woman presented with neck pain and right radiculopathy. MRI C-spine (left) showed focal kyphosis at C4/5 with cervical spondylosis and bulging discs. Her symptoms resolved after a single level ACDF. Postop MRI (right) showed the reduction of focal cervical kyphosis. (Personal collection)

Anterior Cervical Corpectomy and Fusion (ACCF)

ACCF is a more aggressive procedure in which the entire vertebral body and the discs are removed, followed by the placement of a bone graft or cage device to restore cervical alignment and height. Subsequent fusion, similar to ACDF, aims for long-term stability. ACCF often carries a higher risk of complications, including dysphagia, nerve root injury, and graft-related issues.

Artificial Disc Replacement (ADR)

ADR represents a newer paradigm in cervical spine surgery, primarily aimed at preserving motion at the surgical level, thus potentially reducing the risk of adjacent segment disease associated with fusion procedures⁸. In ADR, the diseased disc is replaced with a mechanical device, allowing for continued motion in the disc space. It is typically indicated for younger patients with degenerative disc disease and without significant facet joint pathology. While ADR is associated with quicker recovery and a potential reduction in long-term adjacent segment disease, considerations such as device failure, wear debris, and the unknown long-term durability of the prostheses are noteworthy.

Hybrid constructs, using a combination of ACDF and ACCF, or ACDF and ADR (Fig. 2), have also emerged as a useful tool and may be superior over a long-segment degenerative cervical disease^{9,10}.

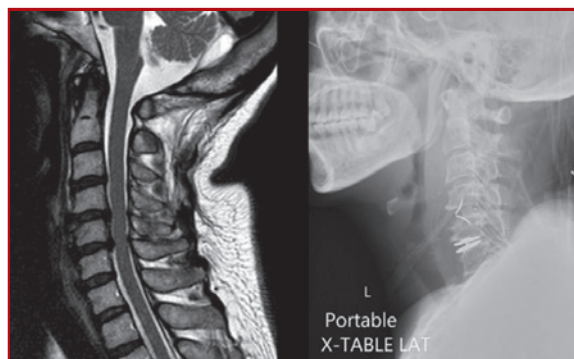


Fig. 2: A 62-year-old woman presented bilateral radiculopathy with pain and numbness. MRI (left) showed C4-6 spondylosis. Hybrid ACDF with ADR was performed (right). (Personal collection)

Posterior Approach

Posterior approaches can involve procedures like laminectomy with or without instrumented fusion, laminoplasty, or foraminotomy. Although a study comparing anterior to posterior approaches showed no significant differences in neurological outcome, we should advocate a patient-tailored approach based on the individual's condition and pathology¹¹. Recent research also has focused on more objective measurable cervical alignment, with radiological measures including C2-7 angle, C2-7 sagittal vertical axis, T1 slope, and modified K line supporting the decision between anterior and posterior approaches¹².

Laminectomy

Laminectomy involves the removal of the lamina to decompress the spinal cord or nerve roots. This procedure is typically performed on patients with significant spinal stenosis causing myelopathy or radiculopathy. It can be performed with fusion at the same time with the addition of pedicle or lateral mass screws and rods. The decision to add instrumented fusion depends on the stability of the spine and the potential of developing post-laminectomy kyphosis.

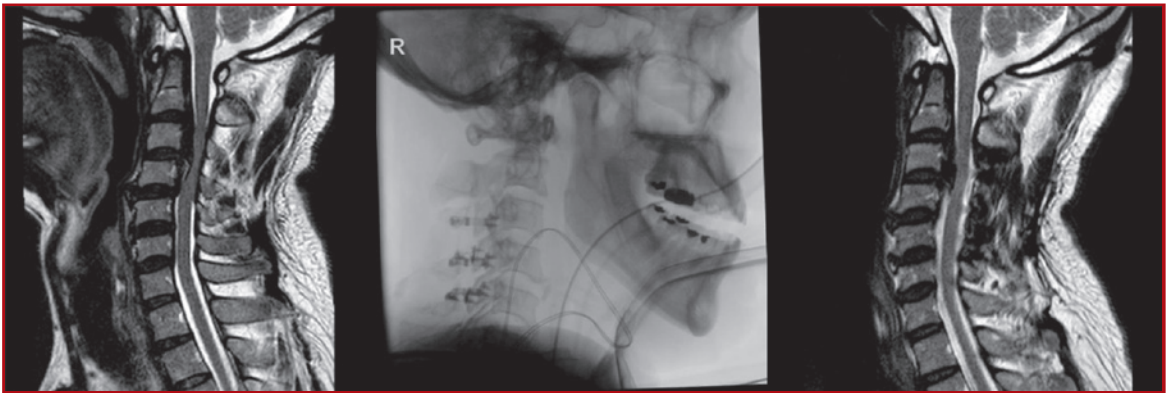


Fig. 3: A 58-year-old man complained of progressive neck pain and bilateral myelopathic hand signs. MRI C-spine (left) showed multilevel cervical OPLL causing cord compression. C3-5 open-door laminoplasty (middle) was performed. Postop MRI (right) showed adequate cord decompression. (Personal collection)

Laminoplasty

Laminoplasty is another posterior approach in which it maintains the stability of the spine, thus reducing the need for fusion and its associated risks. It also preserves neck motion better than fusion procedures. This procedure is especially beneficial for patients with multilevel cervical stenosis who do not have significant spinal instability or cervical kyphosis (Fig. 3). However, potential complications include loss of the achieved spinal canal expansion, and postoperative neck pain.

Foraminotomy

Foraminotomy involves the enlargement of the intervertebral foramen, the passage through which nerve roots exit the spinal cord. This procedure is typically indicated for patients with foraminal stenosis causing radiculopathy. It can be performed as a standalone procedure with minimally invasive tubular or endoscopy, or in conjunction with other procedures like laminectomy or discectomy.

Minimally Invasive Surgery and stereotactic surgery

In line with advancements in other fields of surgery, minimally invasive surgery (MIS) can achieve similar surgical outcomes while minimising complications. The adoption of minimally invasive or endoscopic techniques, such as endoscopic posterior foraminotomy and ACDF via tubular retractor, are gaining acceptance in cervical spine surgery.

Stereotactic techniques such as stereotactic navigation improve the accuracy and safety of both cervical pedicle and lateral mass screws. Robotic-assisted devices that utilise stereotactic navigation can aid with screw placement with more effectiveness and precision¹³.

FUTURE DEVELOPMENT

Biomarkers for Disease Detection

One of the most exciting prospects in the management

of degenerative cervical spine disease is the potential use of biomarkers for early detection and monitoring. Research has indicated that measuring serum microRNAs could help identify specific genes expressed during spinal cord compression.

For instance, the miR-21 is associated with neuroinflammation, and it plays a role in the progression of motor deficits and neuroinflammatory modulation in degenerative cervical myelopathy. The miR-10a modules the development of ossified posterior longitudinal ligament. These biomarkers could potentially improve diagnostic accuracy, guide treatment decisions, and provide insights into the disease's progression and response to treatment^{14,15}.

Advanced MRI Techniques

Advancements in magnetic resonance imaging (MRI) techniques present another promising avenue for the future. The grey to white matter ratio and fractional anisotropy in MRI might provide a more sensitive detection of the progression of myelopathy severity compared to traditional evaluation tools like the Modified Japanese Orthopaedic Association (mJOA) score. These advanced imaging techniques could allow for earlier intervention, potentially improving patient outcomes¹⁶.

A number of microstructural spinal cord MRI techniques have been identified for their clinical potential, namely diffusion tensor imaging (DTI), magnetisation transfer (MT), myelin water-fraction (MWF), and magnetic resonance spectroscopy (MRS). These advanced MRI techniques can qualify or quantify different microstructural components of interest, including myelin content, axonal integrity, gliosis, hypoxia and neuronal loss. In addition, studies utilising MRI T2*-weighted imaging have demonstrated its potential to measure tract-specific atrophy and microstructural changes¹⁷.

Transcranial Magnetic Stimulation (TMS)

Repetitive Transcranial Magnetic Stimulation (rTMS), a non-invasive electrical neuromodulation intervention

that uses a magnetic field to stimulate a region of the brain, has also shown promise in the management of degenerative cervical spine disease. Studies identified "corticospinal reserve capacity" as there is a reduction in activity within the cortical motor area, paired with a compensatory upscale in activity within the supplementary motor area for individuals suffering from degenerative cervical myelopathy (DCM). This discovery could open new avenues for therapeutic intervention, enhancing our understanding of the disease's neurological implications¹⁸.

Besides therapeutic usage, navigated TMS (nTMS) has shown diagnostic potential in the central nervous system. A significant number of studies have explored nTMS based neurophysiological assessments, indicating its role as a screening and prognostication tool in DCM. They are examining the value of TMS as a rapid, inexpensive and non-invasive technique for screening patients before further MRI studies¹⁹.

Riluzole: A Potential Therapeutic Agent

Riluzole is a sodium channel blocker used in motor neuron disease by inhibiting glutamate release. Studies in animal models have suggested that Riluzole might diminish neurological tissue destruction by reducing glutamatergic excitotoxicity and promoting functional recovery in degenerative cervical myelopathy²⁰.

A Phase 3, multicentre, double-blinded, randomised control trial has been completed, examining the benefits of Riluzole in surgical outcomes in DCM. While initial results showed no significant improvement in mJOA or Nurick scores, there appears to be a significant reduction in postoperative neck and neuropathic pain sustained 6 and 12 months after surgery²⁰. As such, Riluzole could constitute an additional tool in the future management of degenerative cervical spine disease.

CONCLUSION

Degenerative cervical spine disease is a prevalent health concern, particularly among middle-aged and older individuals. Its pathogenesis primarily starts with intervertebral disc degeneration and can progress to spinal canal narrowing and nerve root or cord compression, presenting various clinical manifestations such as axial neck pain, radiculopathy and myelopathy. Conservative management is the mainstay of treatment of most asymptomatic or mild cases. However, the disease's natural progression may necessitate surgical intervention in cases where conservative treatments fail, or severe neurological compromise is present.

Despite the complexities and challenges associated with degenerative cervical spine disease, advancements in understanding its pathophysiology, diagnosis, and management strategies continue to provide hope for improved patient outcomes. However, more research is needed, especially in the field of surgical interventions and their long-term effects.

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The Hong Kong Neurosurgical Society
30th ANNUAL SCIENTIFIC MEETING

Spine & Cranio-cervical Junction



Guest Speakers

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Professor of Neurosurgery
Chair in Brain and Spinal Cord Research, Department of Surgery
University of Toronto

Prof. Peng ZHAO, M.D. Ph.D.

Associate Professor
Neurosurgical Department Beijing Tiantan Hospital
Capital Medical University

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to arrive at rectitude





MCHK CME Programme Self-assessment Questions

Please read the article entitled "A Review and Update on Management of Degenerative Cervical Spine Disease" by Dr Alberto CH CHU and complete the following self-assessment questions. Participants in the MCHK CME Programme will be awarded CME credit under the Programme for returning completed answer sheets via fax (2865 0345) or by mail to the Federation Secretariat on or before 29 February 2024. Answers to questions will be provided in the next issue of The Hong Kong Medical Diary. (Address: Duke of Windsor Social Service Bldg., 4/F., 15 Hennessy Rd., Wan Chai. Enquiry: 2527 8898)

Questions 1 - 10: Please answer T (true) or F (false)

1. The ageing population and shifts in lifestyle patterns, such as prolonged use of mobile phones, contribute to more Degenerative cervical spine disease.
2. Static and dynamic compression of spinal neural structures can lead to ischaemia and neuroinflammation, triggering an apoptotic pathway that results in progressive neuronal and oligodendroglia cell death.
3. Unsteady gait, difficulty with fine motor skills, or loss of balance are red flags that indicate the need to refer to a neurosurgeon or spine surgeon.
4. Decreased signal intensity at T1 and T2-weighted MRI of the spinal cord is associated with poorer surgical outcomes.
5. Adjusting workstation ergonomics and maintaining proper posture during daily activities are good recommendations to patients.
6. The microRNA miR-21 is associated with neuroinflammation, which plays a role in the progression of motor deficits in DCM.
7. The miR-10a modulates the development of ossified posterior longitudinal ligament and may indicate a lower risk of developing DCM.
8. Quantitative MRI techniques, such as the grey to white matter ratio and fractional anisotropy, are useful for detecting the progression of DCM.
9. A recent phase 3, multicentre, double-blinded, randomised controlled trial showed that using Riluzole in DCM can have significant improvement in mJOA score.
10. Navigated TMS (nTMS) has shown diagnostic potential in neurophysiological assessments as a screening and prognostication tool in DCM.

ANSWER SHEET FOR FEBRUARY 2024

Please return the completed answer sheet to the Federation Secretariat on or before 29 February 2024 for documentation. 1 CME point will be awarded for answering the MCHK CME programme (for non-specialists) self-assessment questions.

A Review and Update on Management of Degenerative Cervical Spine Disease

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Contact Tel No.: _____ MCHK No. / DCHK No.: _____ (must fill in)

Answers to January 2024 Issue

Two Cases of Acute Retroviral Syndrome

1. **F** 2. **T** 3. **T** 4. **T** 5. **F** 6. **T** 7. **F** 8. **F** 9. **F** 10. **T**

A Review of Spinal Arteriovenous Fistulae

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INTRODUCTION

Spinal arteriovenous fistulae (AVF) are abnormal communications or shunting between arteries and veins without going through the normal capillary network. One of the most common AV shunt (AVS) occurs in the dura and is termed spinal dural arteriovenous fistula. It is a rare pathology, and due to its non-specific presenting symptoms, diagnosis may be delayed, which can lead to delayed treatment and considerable morbidity. The aim of this article is to review the pathophysiology, clinical presentation, classification, imaging findings and treatment approaches of spinal arteriovenous fistulae.

PATHOPHYSIOLOGY

The exact aetiology of spinal AVFs is unknown. It is presumed that spinal dural AVFs are from acquired origin. Spinal AVF can develop from one communication or multiple communications between arteries and veins without a capillary network in between¹. Without the interposition capillary network, the shunt results in increased flow with low resistance in the feeding arteries and increased pressure in the draining veins. If the transition from feeding artery(s) to the vein(s) is through a network of arterioles and venules, i.e., a nidus, this type of lesion is called an arteriovenous malformation (AVM). On the contrary, if the communication occurs directly between one or several arterial feeders and drains to a single vein, these lesions will be termed arteriovenous fistula (AVF). An AVF carries a single draining vein, and usually is fed by a number of feeding arteries¹.

Due to the abnormally high flow in the AVF, high pressure in the draining veins remodels both the arterial and venous components over time. The vasculature in the lesion often progressively dilates and becomes more tortuous and fragile, subsequently forming venous pouches and arterial aneurysms, which may later rupture or compress onto the spinal cord or nerves. In addition, due to the increased venous pressure created by the AVF, the competing normal venous drainage of the spinal cord is impaired, leading to venous congestive myelopathy.

CLINICAL PRESENTATION

The clinical presentation usually depends on the hemodynamic property of the AVF and can be categorised as follows:

Venous Congestive Myelopathy

Due to increased venous pressure directly from the arteriovenous shunting, the venous blood can reflux into the valveless perimedullary veins and also indirectly obstruct venous drainage of the spinal cord causing venous congestion. The lower thoracic spine has relatively fewer venous outflow channels than the cervical spine and thus is more prone to venous congestion and congestive oedema, which usually progresses in the caudocranial direction in the spinal cord. Patients with venous congestive myelopathy commonly present with dysfunctions of conus medullaris, including lower limb weakness, sensory disturbance, bowel or bladder or erectile dysfunction and, radicular pain and low back pain without radicular distribution. Both upper motor neurons and lower motor neurons can be involved, causing both physical signs to be present in the same patient, as has been reported originally by Foix and Alajouanine^{2,3}.

Haemorrhage

Under the chronic abnormal haemodynamics, the spinal AVF may rupture into different anatomical compartments depending on the anatomy of the AVF. If the spinal AVS occurs intradural, then it can cause subarachnoid haemorrhage (SAH) or haematomyelia due to bleeding from a weak point of the spinal AVF. The cause of spinal subarachnoid haemorrhage usually is perimedullary (i.e., pial) arteriovenous fistula (AVF). Sometimes, the dural AVF at the level of the foramen magnum with reflux towards the brain can cause SAH at the level of the foramen magnum or posterior fossa. Epidural AVF can result in epidural hematoma on rare occasions.

Spinal or Radicular Compression

In high-flow AVF, there may be significant dilation of the venous side, causing spinal or radicular compression⁴. Spinal artery aneurysms may compress spinal cord or nerve roots rarely⁴.

Vascular Steal and High-Output Heart Failure

Vascular steal of the spinal cord causing cord infarction was only reported to be a potential mechanism for neurological symptoms in high-flow fistulae in a few cases and is uncommon compared to brain AVS^{5, 6}.



High-output failure may occur in children with large or high-flow AVS⁷, but such a condition is exceedingly rare in Hong Kong.

RATE OF PROGRESSION

Overall, the spinal AVF is a progressive disease if left untreated. In a Netherland series of 80 cases, a gradual course occurred in 63 % of them, while in 26 % of them, an acute transient deterioration was observed to be superimposed on a gradual deterioration course. In 6 % of the patients, there was a stepwise deterioration, while in 5 % of cases, there was an acute onset of symptoms⁴.

CLASSIFICATION

Many classifications have been proposed. The Toronto Classification³ (Table 1) is one of the commonly used classifications to guide the understanding and treatment of different AV shunting lesions in the spinal cord. In this classification, the arteriovenous shunts are classified according to the anatomical location of the shunting point. The most commonly seen among these lesions are spinal dural AVF and spinal pial AVF which will be the focus of discussion in this article.

Table 1. The Toronto Classification of spinal arteriovenous shunts. Excerpted from reference 3.

Vascular Lesions	Topography of the Shunt
Spinal cord AVM	Intradural intramedullary
Pial AVF Microfistula Macrofistula	Intradural pial
Dural AVF	Dural
Epidural AVF	Epidural
Paraspinal AVF	Paraspinal space
Spinal Arteriovenous metamereric syndrome	Multiple compartments, involving the metamereric structures

SPECIFIC ENTITIES

Dural AVF

This is an acquired vascular lesion and accounts for around 70 % of all spinal vascular malformations⁸. It has a male predilection and usually occurs in elderly patients. The feeding artery(s) is usually from a radiculomeningeal branch of a radicular artery and drains into a radicular vein⁹. These radicular arteries may contribute to the spinal cord through their anterior spinal artery and posterior spinal artery branches and, therefore, should be considered during the treatment of such lesions (Fig. 1). The shunting pointing occurs within 1 cm of its dural portion at the dorsal surface of the root sleeve in the intervertebral foramen, located underneath the pedicle of the vertebral body¹. Venous congestive myelopathy is the most common presentation of spinal dural AVF, rarely spinal they can present with intracranial haemorrhage when they are foramen magnum level or can cause radiculopathy due to nerve root compression by a dilated radicular vein.

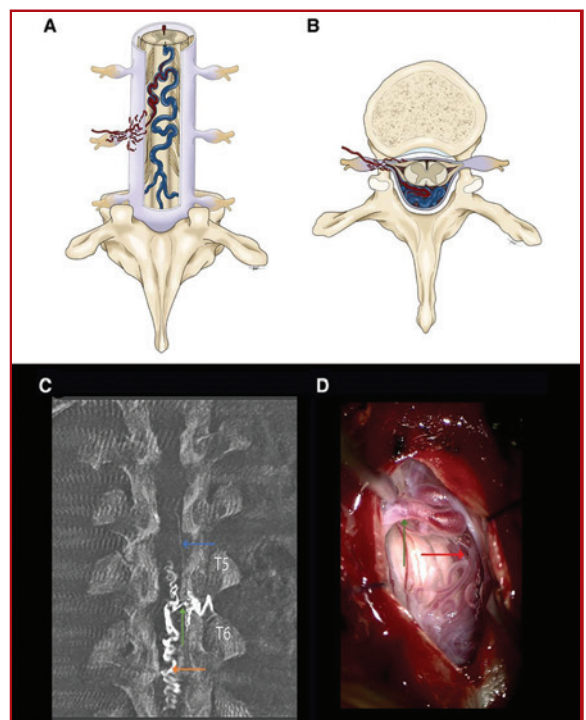


Fig. 1: 3-dimensional illustration (A), and axial illustration (B) showing the shunting point is between the radicular arteries and the radicular vein at the dorsal surface of the dural sleeve of a nerve root in the intervertebral foramen. C, Cone-beam computerised tomographic images showing the radicular artery of T5 supplying both the fistula (green arrow) and the posterior spinal artery (blue arrow) and, therefore, a contraindication for embolisation. The perimedullary vein was engorged due to the dural AVF (orange arrow). D, Open surgical view of the dilated radicular vein of the AV fistula (green arrow) after laminectomy and opening of the dura and the dilated posterior perimedullary vein (red arrow). (Ref: Stéphanie Lenck. Stroke. Spinal and Paraspinal Arteriovenous Lesions, Volume: 50, Issue: 8, Pages: 2259-2269, DOI: (10.1161/STROKEAHA.118.012783))

Pial AVF

Pial AVF is a group of AV shunts located at the subpial space, superficial to the spinal cord, or if they are situated at the filum terminale. They are usually directly fed by the anterior spinal artery (ASA) and rarely by the posterior spinal artery (PSA) and drain directly into the anterior or posterior medullary veins¹⁰ (Fig. 2). Patients with pial AVF most commonly present with congestive myelopathy (around 60 %), and some of them can present with haemorrhage (35 %). A flow-related aneurysm can be found in one-tenth of patients with this lesion. Pial AVF can be further classified into microfistulae and macrofistulae. Microfistulae occur usually in older male patients and are usually idiopathic or traumatic¹¹. On the contrary, macrofistulae usually occur in younger patients and children and are associated with genetic conditions such as Hereditary Haemorrhagic Telangiectasia, Kippel-Trenaunay-Weber syndrome, capillary malformation - AVM syndrome and Cobb Syndromes¹².

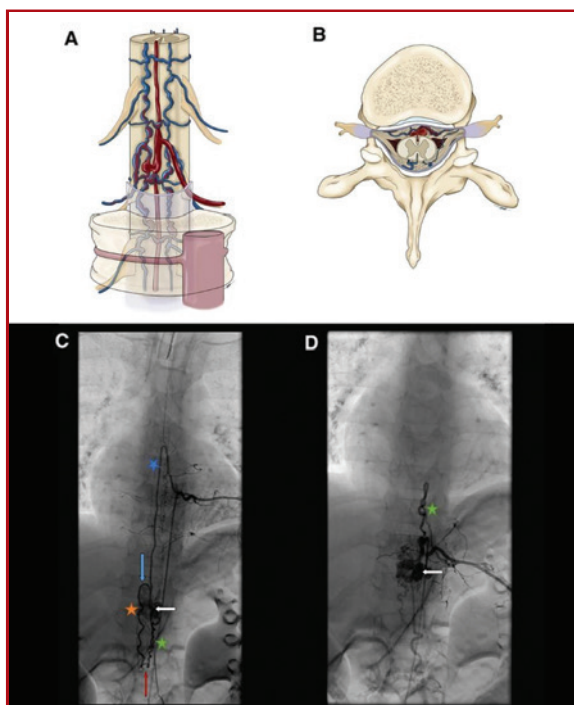


Fig. 2: Pial AVF. A and B, 3-dimensional (A) and axial (B) illustration of a pial AVF at the ventral surface of the spinal cord. The arterial supply is from an anterior spinal artery (ASA) and drains into the anterior perimedullary vein. C, selective angiogram of T8 segmental artery showing a thoracic pial AVF in an 8-year-old child with HHT. The main feeding artery is the left posterior spinal artery (PSA) (green star), and some contribution is from the right PSA (orange star). The ASA (blue star) originates from the left T8 segmental artery and anastomoses with PSA through a circumferential connection (blue arrow). The ASA and PSA connect to each other through the conus arterial basket (red arrow) and feed the pial fistula mainly through the PSA (green star). D, in another patient, the left T11 segmental artery selective angiogram showed the left PSA (green star) feeds directly to a pial AVF with a large venous pouch (white arrow). (Ref: Stéphanie Lenck. Stroke. Spinal and Paraspinal Arteriovenous Lesions, Volume: 50, Issue: 8, Pages: 2259-2269, DOI: (10.1161/STROKEAHA.118.012783))

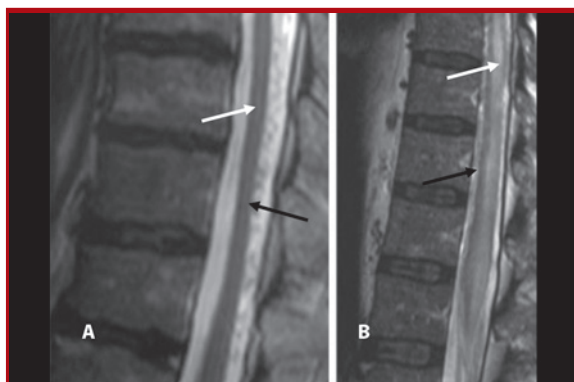


Fig. 3: 3A and 3B. Two different patients with cord oedema (white arrows) and serpentine vessels (black arrows). (Ref: Krings T, Geibprasert S. Spinal dural arteriovenous fistulas. AJNR Am J Neuroradiol. 2009;30:639-648. doi: 10.3174/ajnr.A1485)

IMAGING FEATURES

The initial imaging modality of choice is magnetic resonance imaging and should be followed by a digital subtraction angiogram for confirmation¹³. On MRI, the spinal cord can be visualised on T2-weighted sequences. The cord oedema is usually over multiple segments accompanied by a hypointense rim, which may be due to deoxygenated blood within the dilated capillary surrounding the oedema¹⁴. On T1-weighted images, the oedema may be slightly hypointense and enlarged. On T1-weighted post-contrast images, the cord may have diffuse enhancement, which can be a sign of venous congestion and a breakdown of blood-spinal cord barrier (Fig. 4). As the venous congestion progresses, the spinal cord will become atrophic.

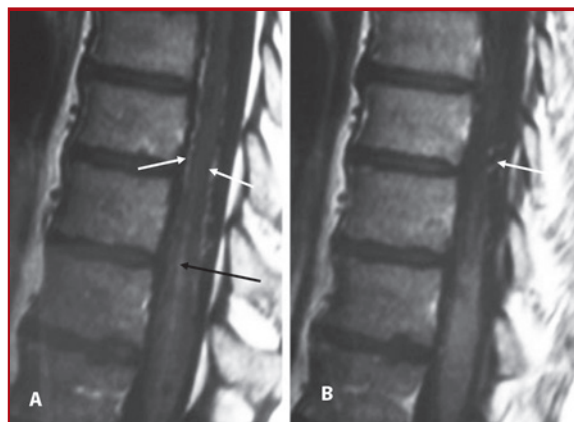


Fig. 4: 4A and 4B. Patients with dilated perimedullary vessels were shown after contrast enhancement. Cord enhancement can be seen in A and B (black arrow), which may imply there has been chronic venous congestion and breakdown of the blood-spinal cord barrier. (Ref: Krings T, Geibprasert S. Spinal dural arteriovenous fistulas. AJNR Am J Neuroradiol. 2009;30:639-648. doi: 10.3174/ajnr.A1485)

Apart from cord oedema, coiled or serpentine vascular structures can be commonly seen, usually on T2-weight images, as flow voids. The dorsal surface usually has more prominent vessels than the ventral surface. However, in the case of slow-flow shunts, there may be prominent coiled vessels after contrast enhancement. Heavily T2-weighted T2 sequences can demonstrate the serpentine vessels more clearly. (These may include 3D turbo spin-echo [3D-TSE] or Constructive Interference in Steady State [CISS], Fast Imaging Employing Steady-state acquisition [FIESTA] (Fig. 5).

Contrast-enhanced spinal MRA can help localise the lesions and avoid super-selective injections of all possible arterial feeders. First-pass gadolinium-enhanced MRA is a useful technique which can demonstrate early venous filling and suggest the location of the AV shunt, and help reduce the time spent in superselective angiography¹⁵ (Fig. 6).

On digital subtraction angiography (DSA), the AV fistula is identified to be the segmental artery having early venous drainage and retrograde contrast filling of radiculomedullary veins (Fig. 7). There is usually



a network of dilated perimedullary veins due to the venous congestion and hypertension. The anterior spinal artery and posterior spinal artery should be looked for during the DSA to observe if embolisation is dangerous in these cases. Even if the AV shunting has not been demonstrated yet, a delayed venous return after ASA injection can be a sign implying venous congestion and underlying AV shunting is causing it. Whereas if normal venous return following injection to ASA, then dural AVF is much less likely to exist¹⁶.

TREATMENT APPROACHES

The aim of treatment in spinal arteriovenous fistulae is to disconnect the shunting zone (i.e. to occlude the most distal part of the feeding artery with the most proximal part of the receiving vein). Proximal occlusion without reaching the shunting zone may improve the symptoms transiently, but will lead to recurrence of the fistula due to collateralisation of dura vessels. In pial AVF, proximal occlusion of ASA or PSA may cause significant neurological damage as these pial arteries have important supplies to the spinal cord. Two options are commonly available to treat these spinal AV shunts: microsurgical disconnection of the shunting zone or endovascular therapy.

Microsurgical disconnection is usually a safe and straight-forward procedure for fistulae and carries a high chance of cure. In endovascular therapy, liquid embolic agents or N-butyl cyanoacrylate are common agents used to obliterate the fistula point. The endovascular therapy has a high chance of cure if the embolic agents can pass the shunting zone to the venous side. However, the ASA and PSA supply must be carefully excluded before deciding on endovascular treatment. In thoracic dural AVF, the PSA can be quite difficult to visualise and, therefore, carries a risk of neurological injury from inadvertent embolisation.

PROGNOSIS

Following the complete occlusion of AV fistula, the progression of the disease is usually stopped. If a deterioration of symptom occurs after initial improvement, recanalisation of the shunt or a secondary shunt should be considered and investigated.

CONCLUSION

Spinal AV fistula is a rare group of neurovascular diseases. The presentation is often subtle and can lead to a delay in diagnosis and irreversible progression of disease. Knowledge of the spinal vascular anatomy is required to understand and manage these lesions. MRI and MRA are the first line investigations, followed by a complete digital subtraction spinal angiogram. Since these lesions are rare and sometimes complex, patients with these conditions should be referred to an experienced centre and often require multidisciplinary management.

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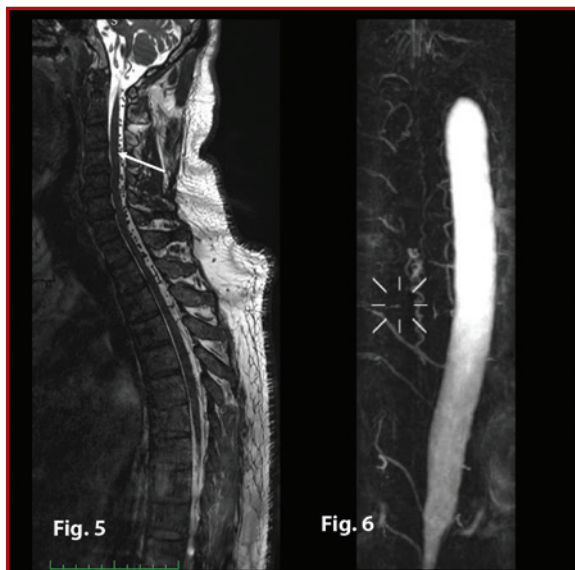


Fig. 5: Heavily T2-weighted image showing the prominent serpentine vessels on the dorsal aspect of the cervical to the thoracic cord (white arrow). In this patient, there was no cord oedema, and the patient had minimal myelopathic symptoms. (Personal collection)

Fig. 6: First-pass contrast-enhanced MRA can clearly illustrate the early venous drainage and thereby confirm the presence of an AV shunt in this patient and suggest the level T10 segmental artery to be the arterial feeder. This was later confirmed by spinal angiography. (Personal collection)

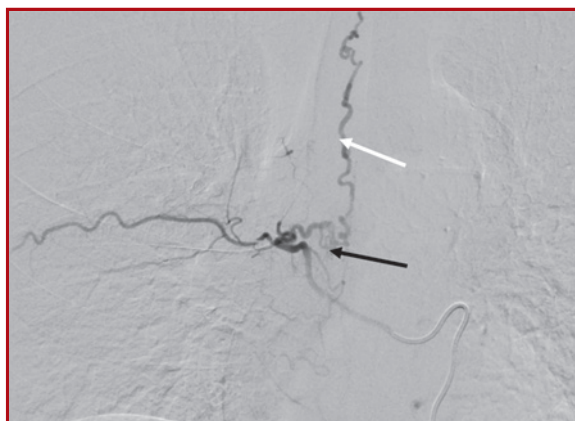


Fig. 7: Spinal dural AV fistula occurring at T10 root sleeve (black arrow). The arterial supply is from the T10 segmental artery and drains into the perimedullary veins, which are dilated and flowed in a retrograde direction (white arrow). (Personal collection)

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A Neurosurgical Perspective on the New WHO Classification for Pituitary Tumour

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Dr Michael MH YUEN

INTRODUCTION

Recently, some concerns were encountered by patients and their referring doctors regarding the diagnosis and prognosis of pituitary tumours. This concern arose from the new World Health Organization (WHO) classification and the updated International Classification of Disease Oncology (ICD - O). The key changes include reclassifying pituitary tumours as part of neuroendocrine tumours and the upgrading from 0 (benign) to 3 (malignant) in ICD - O coding. These changes led to a great debate among neurosurgeons, endocrinologists and pathologists.

BACKGROUND

Pituitary tumours are one of the commonest intracranial tumours, which account for 10 % of all primary intracranial neoplasms. Pituitary adenomas are the most common pituitary neoplasm and originate from the anterior pituitary gland. A meta-analysis found that around 14 % of autopsy studies and 22.5 % of radiological studies of the brain showed the presence of pituitary adenomas. Most pituitary adenomas are well known to be indolent and benign, though a minority of cases behave aggressively. Survival is usually not affected. Malignant pituitary adenomas, which are known as pituitary carcinomas, are extremely rare.

Pituitary adenomas can be classified into different categories according to the presence of overproduction of pituitary hormones (functional vs non-functional), size of pituitary adenomas (microadenomas vs macroadenomas, with 1 cm as the cut-off), anatomical extension (based on findings in magnetic resonance imaging or intraoperative findings; commonly used classifications include Hardy classification and Knosp classification) and histology +/- clinical aggressiveness.

Many patients are found to have pituitary adenomas while having neuroimaging for other purposes. This type of pituitary adenoma is also termed a pituitary incidentaloma. Those patients with pituitary incidentaloma are usually asymptomatic, and most of them only require regular surveillance of the tumours. Symptomatic patients with pituitary adenomas usually suffer from symptoms related to either the overproduction of certain pituitary hormones or the mass effect of the tumours.

Among the functional adenomas, prolactinoma is the most common type of functional tumour, and it is the only type of functional adenoma that uses medical

therapy (dopamine agonists) as the first choice of treatment. On the other hand, an attempt to have complete excision of the tumours is adopted as the first choice of treatment for functional adenomas other than prolactinoma. Adjuvant medical treatment and radiotherapy may be required for selected cases.

For non-functional pituitary macroadenoma with mass effect, surgery is the choice of treatment. Nowadays, endoscopic transsphenoidal surgery is usually adopted with the aims of adequate decompression.

LATEST UPDATE ON 5TH EDITION OF WHO CLASSIFICATION OF ENDOCRINE AND NEUROENDOCRINE TUMOUR (ENDO5) AND ICD - O CODING

The term "pituitary neuroendocrine tumour" (Pit NET) was first introduced by International Pituitary Pathology Club (IPPC) in 2017 to address the wide spectrum of clinical behaviours (from indolent to aggressive in some cases) and the potential severe morbidity arising from mass effect or hormonal over productivity^{1, 2, 3}. IPPC suggested Pit NET shared features of neuroendocrine cells and should be grouped into the family of neuroendocrine tumours. This proposal was adopted in ENDO5 which was published in 2022⁴.

In ENDO5, pituitary adenomas are replaced by Pit NET and pituitary carcinomas are replaced by metastatic Pit NET. ICD - O coding for all the Pit NET is upgraded to 3 (malignant). Atypical adenoma and the use of Ki67 and mitotic figures for risk stratification are abandoned. Further subtypes of Pit NET are based on pituitary cell lineage, namely PIT1 - lineage, TPIT - lineage and SF1 - lineage⁵.

The summaries of major changes are highlighted by Table 1⁴.

CONTROVERSIES

Since the proposed change of terminology by IPPC was published in 2017, there has been a great debate in the field worldwide. The Pituitary Society is among the international organisation of the field to show its objection to this new change. One of the main reasons for the objection is the difference in terms of clinical behaviours, proportion of malignancy and survival of patients of Pit NET when compared with patients

**Table 1: Comparison of major changes among ENDO3, ENDO4 and ENDO5**

	ENDO3 WHO 2004	ENDO4 WHO 2017	ENDO5 WHO 2022
Nomenclature	Typical adenoma	Adenoma (Subtype)	Pituitary neuroendocrine tumour (PitNET) (Subtype)
	Atypical adenoma	-	-
	Carcinoma	Carcinoma	Metastatic PitNET (Subtype)
ICD-O code*	8272/0 Typical adenoma	8272/0 Adenoma	8272/3 PitNET
	8272/1 Atypical adenoma	-	-
	8272/3 Carcinoma	8272/3 Carcinoma	8272/3 Metastatic PitNET
Terminology based on	Hormone secretion	Hormone secretion/pituitary cell lineage	Pituitary cell lineage
Grading	No grading	No grading	No grading; Ki67 and mitosis unproven
Staging	No staging	No staging	No staging; invasion unproven
Proliferation markers	Ki67: cut off 3%	Ki67 (hotspots count): no cut off	Proliferative rate unproven
	Mitosis: no cut off	Mitosis: no cut off	-
	P53 diffuse	(P53)	-
High risk of recurrence tumours	Atypical adenoma	Invasive tumours, highly proliferative (Ki67 & mitosis), rapid growth or Subtypes: - Crooke cell adenoma - Silent corticotroph - Lactotroph in men - Plurihormonal PIT1+ adenoma - Sparsely granulated somatotroph	Only accurate histological subtyping Subtypes: - Crooke cell adenoma - Silent corticotroph - Acidophil stem cell tumours - Immature PIT1-lineage tumours - Sparsely granulated somatotroph - Null Cell

Edited from Villa C, Baussart B, Assié G, et al. The World Health Organization classifications of pituitary neuroendocrine tumours: a clinico-pathological appraisal. Endocr Relat Cancer. 2023 Jun 22; 30; 8:e230021. doi: 10.1530/ERC-23-0021. PMID: 37068095.

with NET in other body parts^{6, 7, 8}. The upgrade of ICD - O coding may also cause unnecessary anxiety for the majority of patients with Pit NET that formerly belonged to the diagnosis of pituitary adenomas and might lead to unnecessary requests for active treatment. It may also lead to potential conflicts in insurance related issues on coverage of malignancy^{9, 10, 11}. Another criticism of the ENDO5 is that it lacks guidance on prognosis and risk factors prediction for fast growing tumours and recurrence.

NEUROSURGICAL PERSPECTIVE

The new classification does not affect the current neurosurgical practice for patients with pituitary tumours in terms of investigations and management, as the new classification does not impact the prognosis or risk factor stratification. However, more detailed patient counselling and education may be required, especially for those patients who are more anxious or misled by the new terminology.

CONCLUSION

The new classification for pituitary tumours does not shed light on prognostic predictors for pituitary neuroendocrine tumours. Further studies, including genetic studies and other immunohistochemical studies, are required to provide more clues in outcome prediction that may be adopted in future classification systems and ultimately lead to new standards of practice in managing this type of pathology.

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Women In Neurosurgery (WIN)

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Specialist in Neurosurgery

Associate Consultant, Department of Neurosurgery, Princess Margaret Hospital



Dr Teresa PK TSE



Despite women still being minorities in neurosurgery, in recent years, the number of female neurosurgeons has been increasing. Besides the usual hardship all neurosurgeons face, including the demanding nature of neurosurgery with long working hours, frequent on-call duties and work-life imbalance, women in neurosurgery face additional pressures related to family responsibilities, societal expectations and the underrepresentation of women in the field.



HOW DO I COPE WITH IT?

Family, Pregnancy and Childbirth

I got married when I was in my year three higher neurosurgical training. I gave birth after achieving a fellowship in neurosurgery. I have two kids, a 5-year-old daughter and a 2-year-old son. Balancing a career

in neurosurgery with motherhood can be challenging, as both roles require significant time and dedication. It also involves sacrifice and prioritisation. Women may also need to make choices regarding career opportunities and make adjustments to their work routines. I decided to step away from endovascular interventional procedures to avoid radiation exposure since I have had the plans for pregnancy. I am now back in the endovascular suite after the birth of my second child. With planning, a supportive husband and via open communications with colleagues, I successfully thrive as a neurosurgeon and a mother, contributing to both my profession and family.

HOBBIES

Singing

Taking care of one's mental well-being is crucial. I attend singing lessons every week and singing has become a very important part of my life. It's also an effective way to manage stress and maintain a healthy work-life balance. I have a strong passion for singing and pursue it as a professional interest. Enrolment in singing competitions and performances motivates me to enhance my vocal skills, which require discipline, regular practice and perseverance, qualities that can be transferred to the professional realm of neurosurgery. Effective time management allows me to carve out time for vocal practice, rehearsals and performances, which brings a sense of achievement, confidence and personal growth.



Tennis

I started to learn tennis with my husband just before we got married. We aimed to develop a common



interest which helped to strengthen our bond and provide opportunities for shared experiences and quality time together, in addition to the maintenance of one's physical health. Despite not being the winner, enrolling in tennis competitions provided me with valuable learning experiences. Competing against skilled opponents helps me gauge my own abilities and understand the level of play to succeed. It also motivates me to work harder on my technique, strategy and physical fitness.



LAST WORD

It's essential to note that every woman's experience is unique, and decisions regarding pregnancy, childbirth and career choices should be based on personal circumstances, values, and expectations. Prioritising self-care and well-being is essential for work-life balance. Engaging in activities that promote mental and physical health, such as exercise, music and spending quality time with loved ones, enhances overall satisfaction in both personal and professional life. Women in neurosurgery may need to redefine their perceptions of success to achieve work-life balance. Women in neurosurgery may also need to recognise that success is not solely determined by professional achievements; instead, it can be multifaceted and fulfilment in personal relationships, and hobbies can contribute to a sense of balance and satisfaction.



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Sunday	Monday	Tuesday	Wednesday	Thursday	Friday	Saturday
				1	2	3
4	5	* In-person / Zoom HKMA-HKSH CME Programme 2023-2024 Topic: Updates in Interventional Endoscopy	* In-person / Zoom HKMA-CUHK Medical Centre CME Programme 2024 Topic: To-be-confirmed	8	9	10
11	12		* The Hong Kong Neurosurgical Society Monthly Academic Meeting –Role of Next Generation Sequencing in brain tumor	15	16	17
18	19	20	* Zoom The Impact of Microbiome on Long COVID, Improvement of Health Outcomes, and Quality of Life: Insights from Recent Randomized Controlled Trials and Beyond	* In-person Topic: Recent Updates on Osteoporosis Management * FMSHK Executive Committee Meeting * FMSHK Council Meeting	* Zoom Lipid Management in Patients with Mixed-Dyslipidemia - How Can We Do Better?	24
25	26	27	* In-person Topic: Updates on Lipid Management	* In-person / Zoom Esophageal Squamous-Cell Carcinoma (ESCC) & Nasopharyngeal Cancer (NPC) and Potential Application of Patient-Derived Organoids	23	29
			28			



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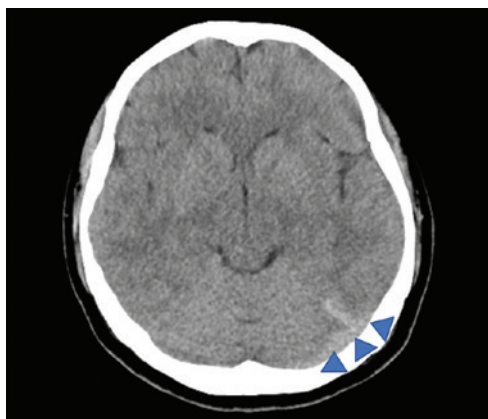


Date / Time	Function	Enquiry / Remarks
6 TUE 2:00 PM	In-person / Zoom HKMA-HKSH CME Programme 2023-2024 Topic: Updates in Interventional Endoscopy Organiser: The Hong Kong Medical Association and the Hong Kong Sanatorium & Hospital Speaker: Dr Anthony Yuen-bun TEOH Venue: The HKMA Wanchai Premises, 5/F, Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, HK	HKMA CME Dept. Tel: 3108 2507 1 CME Point
7 WED 2:00 PM	In-person / Zoom HKMA-CUHK Medical Centre CME Programme 2024 Topic: To-be-confirmed Organiser: The Hong Kong Medical Association and the CUHK Medical Centre Speaker: To-be-confirmed Venue: The HKMA Wanchai Premises, 5/F, Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, HK	HKMA CME Dept. Tel: 3108 2507 1 CME Point
14 WED 7:30 AM	The Hong Kong Neurosurgical Society Monthly Academic Meeting –Role of Next Generation Sequencing in brain tumor Organiser: Hong Kong Neurosurgical Society Speaker(s): Dr LAM Shek-ching Chairman: Dr Jason Kwan-ho CHOW Venue: Conference Room, F2, Department of Neurosurgery, Queen Elizabeth Hospital; or via Zoom meeting	CME Accreditation College: 1.5 points College of Surgeons of Hong Kong Enquiry: Name: Dr Calvin MAK Tel: 2595 6456 Fax. No.: 2965 4061
21 WED 2:00 PM	Zoom The Impact of Microbiome on Long COVID, Improvement of Health Outcomes, and Quality of Life: Insights from Recent Randomized Controlled Trials and Beyond Organiser: The Hong Kong Medical Association Speaker: Dr Martin Chi-sang WONG	HKMA CME Dept. Tel: 3108 2507 1 CME Point
22 THU 2:00 PM 7:00 PM 8:00 PM	In-person Topic: Recent Updates on Osteoporosis Management Organiser: The HKMA District Health Network Speaker: Dr YIP Wai-man Venue: Rich Garden Restaurant, C2/F, 114 Broadway Street, Mei Foo Sun Chuen Stage 8, Mei Foo FMSHK Executive Committee Meeting Organiser: The Federation of Medical Societies of Hong Kong; Venue: Council Chamber, 4/F, Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, Hong Kong FMSHK Council Meeting Organiser: The Federation of Medical Societies of Hong Kong; Venue: Council Chamber, 4/F, Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, Hong Kong	The HKMA District Health Network Dept. Tel: 2861 1979 1 CME Point Ms Nancy CHAN Tel: 2527 8898 Ms Nancy CHAN Tel: 2527 8898
23 FRI 2:00 PM	Zoom Lipid Management in Patients with Mixed-Dyslipidemia – How Can We Do Better? Organiser: The Hong Kong Medical Association Speaker: Dr Bernard Bun-lap WONG	HKMA CME Dept. Tel: 3108 2507 1 CME Point
28 WED 2:00 PM	In-person Topic: Updates on Lipid Management Organiser: The HKMA District Health Network Speaker: Dr KONG Chun-cheong Venue: Lei Garden, Shop 1130-1143, L1/F, YOHO MALL I, 9 Long Yat Road, Yuen Long	The HKMA District Health Network Dept. Tel: 2861 1979 1 CME Point
29 THU 2:00 PM	In-person / Zoom Therapeutic Strategies for Esophageal Squamous-Cell Carcinoma (ESCC) & Nasopharyngeal Cancer (NPC) and Potential Application of Patient-Derived Organoids Organiser: The Hong Kong Medical Association and the Hong Kong Science Park Speaker: Prof Dora Lai-wan KWONG Venue: The HKMA Wanchai Premises, 5/F, Duke of Windsor Social Service Building, 15 Hennessy Road, Wanchai, HK	HKMA CME Dept. Tel: 3108 2507 1 CME Point



Answers to Radiology Quiz

Answers:



1. Hyperdensity at the left posterior fossa along the left transverse sinus (blue arrowheads). No haemorrhage.
2. Dural venous sinus thrombosis.
3.
 - Hormonal: oral contraceptive pill, pregnancy, steroids
 - Prothrombotic conditions: protein S deficiency, polycythaemia, sickle cell anaemia
 - Systemic illness: dehydration, sepsis, malignancy, connective tissue disorders
 - Local factors: infections (especially mastoid sinus), skull fracture with dural venous sinus involvement
 - Idiopathic
4. Venous infarct, haemorrhagic venous infarct, dural arteriovenous fistula, intracerebral haemorrhage, increased CSF pressure.
5. CT cerebral venogram to look for non-opacification of the dural venous sinus for confirmation.
6. Consult a neurologist or neurosurgeon. Anticoagulation is the mainstay of treatment, even in the presence of a haemorrhagic venous infarct. Interventional neuroradiologists can perform catheter-directed thrombolysis by using targeted thrombolytics in the affected sinuses.

Dr Carol PY CHIEN

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