Neuro-endovascular Therapy of Carotid-Cavernous Fistula

Dr. Kin-ming Cheng  MBBS(HK), FRCS(Edin), FCSHK, FHKAM(Surgery)
Department of Neurosurgery, Queen Elizabeth Hospital

Introduction
Carotid-cavernous fistula (CCF) is an abnormal arteriovenous communication in the cavernous sinus. The cavernous sinus is a venous cavern between layers of the dura of the skull base (Fig 1). It receives venous drainage of the eye and the brain via the ophthalmic veins and the superficial middle cerebral vein. There are also venous communications with the opposite cavernous sinus, the clival venous plexus and the transverse sinus. CCF was first described by Travers (1809) who recognised pulsating exophthalmos to be due to an arterio-venous shunt and treated it with common carotid ligation.1 Although surgical ligation often produced initial good results, collateralisation from the arterial branches of the external and internal carotid artery produced high recurrence rate. Later, Brooks (1931) surgically opened the internal carotid artery in the neck and floated a strip of muscle into the fistula and Gardner (1932) described internal carotid artery ligation as a method of treatment.1 It was Serbinenko (1974) who described the use of detachable balloon technique to obliterate the fistula and leave the carotid artery patent.2 In the modern era, neuroendovascular therapy offers a safer and more effective treatment for CCF.3, 4, 5

Aetiology
Carotid-cavernous fistula can be classified into traumatic and spontaneous types. The traumatic type is caused by severe head injury after high velocity traffic accident, major skull base fracture or penetrating wound through the orbit. The spontaneous type can be congenital, secondary to rupture of carotid aneurysm into the cavernous sinus or acquired with multiple arterial branches of the carotid arteries that shunt into the cavernous sinus. The later type is the commonest and is usually called dural CCF. In Hong Kong, traumatic cause is uncommon and most are spontaneous in origin. In Queen Elizabeth Hospital (1997-2005), a total of 80 patients with CCF were treated. Seventy-six (95%) cases are spontaneous and only 4 cases (4%) are of the traumatic type.

Pathophysiology
The abnormal arteriovenous shunt creates a high venous pressure in the cavernous sinus that impairs venous drainage of the ophthalmic veins and causes eye congestion and increase in intraocular pressure (Fig 2). The increase in cavernous sinus pressure also causes pressure effects on the third, fourth and sixth cranial nerves that resulted in ophthalmoplegia. In addition, CCF can create back pressure in the cerebral veins that can lead to intracerebral haemorrhage (Fig 3).

Clinical features
Carotid-cavernous fistula usually presents with chemosis, proptosis, diplopia and visual impairment of the eye. Rarely, CCF can cause cerebral dysfunction (hemiplegia, dysphasia, etc). The clinical presentation of traumatic CCF is acute but some of the cases can present one week after the injury. Spontaneous CCF is usually insidious in onset. Physical signs include eye bruit, pulsatile exophthalmos, cranial nerve palsy (3rd, 4th, 6th), and impaired vision.

Investigations
Carotid-cavernous fistula can be diagnosed with imaging studies. CT scan with contrast may show skull base fractures, a dilated superior ophthalmic vein and venous engorgement in the region of the cavernous sinus. MRI and MR angiography are also useful tools for screening and follow-up of the patients. Cerebral angiography is a more invasive form of imaging study which provides the best diagnostic study for CCF. Selective internal and external carotid angiograms are essential to define the anatomy and develop the treatment plan.

Treatment
In the old days, CCF was treated with surgical operations with significant morbidity and mortality. In the modern era, neuroendovascular therapy offers a safe and effective treatment for patients with CCF and has replaced open surgery as the treatment of choice.

Most of the endovascular procedures for CCF can be done under local anaesthesia. The procedure starts with arterial or venous puncture in the groin region. Catheters and co-axial microcatheters are introduced via the femoral route to the intracranial circulation. The endovascular approaches can be transarterial or transvenous. The embolic materials include polyvinyl alcohol particles, histoacryl glue, platinum coils and Guglielmi detachable coils. Although technically more difficult to perform, the transvenous approach is safer and more effective than the transarterial approach and is recommended in most cases of dural CCF. The
transvenous embolisation routes includes the inferior petrosal sinus, inter-carvenous sinus, superior ophthalmic vein (Fig 4, 5, 6) and rarely, cortical vein, superior petrosal sinus and pterygoid venous plexus. Clinical cure can be achieved in more than 90% of the patients and the procedure-related morbidity and mortality is very low.5,6

Conclusion

Carotid-cavernous fistula is a well-known disease entity that can be encountered in daily clinical practice. CCF can cause visual and neurological deficits when the condition is left untreated. Diagnosis of CCF can be made by clinical examination and imaging studies. Neuroendovascular therapy is safe and effective and is the treatment of choice in CCF.

References