Challenges in Palliative Care of Neurodegenerative Patients

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Introduction

Although palliative care needs of non-cancer patients are recognized, palliative care provision to these patients is still limited in Hong Kong. In this article, we shall focus our discussion on two patients with neurodegenerative diseases whom we cared for in Bradbury Hospice. These two cases demonstrate the challenges in the care process from the perspectives of health care workers and family caregivers.

Case history one

Mr X was a 52-year-old gentleman suffering from Machado-Joseph Disease since the age of 33 (1987). He presented with unsteady gait and the diagnosis was confirmed by genetic study. Mr X was married and had one teenage daughter. Mr. X’s father and younger sister also suffered from Machado-Joseph Disease.

Machado-Joseph disease (MJD) is a rare type of hereditary ataxia characterized by clumsiness and weakness of limbs, spasticity, a staggering lurching gait, difficulty in speech and swallowing, involuntary eye movement, double vision and frequent urination. The life expectancy of affected patient ranges from mid-thirties in severe forms of MJD to a normal life expectancy in mild forms. Aspiration pneumonia is often the cause of premature death in MJD.

Mr X had progressive deterioration and became chair bound since 2003. In 2004, he developed coughing and choking with meals. He also had progressive weight loss, double vision, urinary retention and terminal dribbling. In 2005, he was referred to Bradbury Hospice for palliative care.

Mr X was admitted to Bradbury Hospice in August 2007 for dysphagia, choking on feeding and persistent fever. Physical examination showed cerebellar signs and decreased air entry over right chest. Chest X-ray showed a large shadow over right lung with well-defined border and a suspected fluid level within the shadow. Mr. X was treated as aspiration pneumonia. However, he did not respond to antibiotic treatment initially and he underwent a protracted clinical course requiring chest drain insertion for draining of loculated pleural effusion. Mr. X was referred to speech therapist and tube feeding was suggested. The feeding tube had caused him much discomfort and he was further agitated by his inability to express his views and emotions effectively. The occupational therapist assisted Mr X in using pictures for better communication.

Eventually Mr X recovered from the acute infection. He requested oral feeding but was advised against by the team. He became emotionally labile and eventually he was resumed on oral feeding after long discussions with him and his family. Mr. X tolerated oral food well and continued to improve. The feeding tube was removed before he went home.

Challenges facing the palliative care team

In caring for Mr X, our palliative care team had to overcome barriers in communication and to work with the uncertainty of the clinical course and prognosis. The clinical course of Mr. X lasted for more than 15 years, and disease progression could be less predictable than that of cancer. Defining the end of life stage could be difficult. As in Mr X, his acute infection was initially thought to be a terminal event, but he recovered with some functional improvement. Without effective communication with Mr X, it would not be possible to elicit his concerns and arrived at the decision of resuming oral feeding.

In a questionnaire survey of 220 specialist palliative care units in the UK and Ireland, it was found that many hospices were involved only in the terminal stage of neurodegenerative diseases. It has been recommended that early stages involvement can be advantageous in easier communication with patient and for a clearer understanding of the patient’s views on their care. Moreover, involvement of the wider multidisciplinary team varied from hospice to hospice. For example, lack of access to speech and language therapist could impair service to patients with speech and swallowing problems.
For symptom palliation in neurodegenerative diseases, one should be aware of the possible differences in symptom profile as compared with cancer. In a systematic review of symptom prevalence among patients suffering from neurodegenerative diseases\(^3\), it was found that these patients suffered from more neurological symptoms than cancer. Problems with communication, symptoms of weakness, spasms, bowel and bladder dysfunction and swallowing problems were more prevalent. Palliative care providers have to acquire knowledge and skills in neurological symptom control. It is suggested that there is greater need for the involvement and integration of neurological and rehabilitation in the development of palliative care service.

**Case history 2**

Ms C was a 29 year-old-lady diagnosed to suffer from sporadic Type II neurofibromatosis in 2000. MRI of her brain and spine showed bilateral acoustic neuroma and spinal tumors at multiple cervical levels with spinal cord compression. She had multiple operations done in her spine from 2000 to 2006. However, Ms C continued to develop multiple neurofibromata. She was on long term Foley and her right lower limb was paralysed since 2006. She became deaf and then tetraplegic in 2008, and was then referred to Bradbury Hospice.

Ms C was admitted to Bradbury Hospice in April 2008 for symptom control and respite care. She had increased left upper limb weakness and became totally dependent. She had pain over multiple sites and frequent re-positioning only gave modest relief. Her sleep was disturbed and her elderly parents were exhausted.

After admission, pain was controlled with methadone and NSAID. Frequent turning was performed by nurses. Physiotherapist performed limb stretching exercise and occupational therapist modified a wheelchair to suit her needs. Medical social worker attended to the needs of her parents. Finally Ms C was discharged with home care support.

**Challenges facing the family caregivers**

Patient’s mother expressed worries about looking after her daughter in the future. She did not know what to expect and worried about what would happen to her. She did not know how long her daughter would live. She reflected on the many surgeries she had had and in the end it was useless. Her own health had suffered; she had aches and pains and difficulty sleeping. Now they had financial difficulties.

In a qualitative study on the responses of family carers of patients with muscular dystrophy and motor neuron diseases\(^4\), three key themes had emerged: including reactions and responses, health care system crossing points and reaching forward. Reactions and responses of family members included grieving everyday, fearing each crisis may mean the end and watching life in reverse. As for health care crossing points, family caregivers felt getting lost in the system and had to live with limits. Lastly, theme of reaching forward included sub-themes of holding on the big picture, needing help to plan the future and just getting on with it.

Results from another qualitative study also echoed with these findings\(^5\). In addition, family caregivers did not always seek help in particular with regard to their emotional needs.

Challenges facing the family caregivers are no less than the patients. Palliative care professionals have yet much to learn in caring for these patients with non-cancer diseases.

**References**