

Chapter

46

Comparing and Contrasting the Approach to Advance Care Planning in Multiple Sclerosis, Case 1

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Clinical History

A 60-year-old woman with a long-standing history of secondary progressive multiple sclerosis (MS), hyperthyroidism, and depression presented to the emergency department with shortness of breath and drowsiness. Her MS history dated back more than four decades with emergence of secondary progressive disease at the age of 41 years, after which she accumulated substantial neurological disability. She required the use of a wheelchair starting at the age of 47, soon after which she became completely dependent in all of her activities of daily living. By the time of presentation, she had no purposeful movement of her limbs, severe spasticity and contractures, debilitating oscillopsia and poor visual acuity, an indwelling urinary catheter, intractable constipation requiring regular enemas, and significant cognitive decline. Recent years were marked by recurrent admissions to the hospital because of complications related to her MS, including multiple bouts of sepsis secondary to urinary tract infections, bronchopneumonia, and grade IV sacral pressure sores. She required several prolonged stays on rehabilitation wards for treatment and respite care, but she was primarily cared for by her husband at home with community support. Given her inability to maintain adequate nutrition, a percutaneous endoscopic gastrostomy (PEG) was planned in the upcoming days.

She presented to the emergency department because of shortness of breath, drowsiness, and anorexia. A week prior to presentation, she was noted to be increasingly lethargic. The morning of presentation, she started to make grunting noises and she gasped for air. All of her symptoms swiftly worsened during the course of the day, prompting a visit to the hospital that evening.

Examination

She was drowsy and made incomprehensible sounds. She appeared uncomfortable using accessory muscles

of respiration. She was diaphoretic, tachypneic (21 breaths per minute), and hypoxemic (SpO₂ 84 percent on room air). She was tachycardic (120 beats per minute) and had a blood pressure of 140/80 mmHg. Her temperature was 37.5°C, mucous membranes were dry with a low jugular venous pulsation (JVP), and dark urine was in her catheter bag. On auscultation of her chest, coarse crepitations throughout her right lung and at the left lung base were noted. Her abdomen was soft but distended. A 4 cm-deep sacral pressure sore was noted. The neurological examination was significant for a depressed level of consciousness with a Glasgow Coma Score (GCS) of 10 (baseline was 12). She inconsistently followed simple commands, but she could not communicate meaningfully. The motor examination was significant for severe contractures and disuse atrophy of both upper and lower limbs. Sensory, cerebellar/coordination, and gait examinations were deferred.

Palliative Domain of Care

Structure and Processes of Care – Interdisciplinary team assessment based on patient/family goals of care; prognosis; disposition (level of care – inpatient unit, home); referral to palliative care to guide decisions about the goals of care.

She had severe end-stage secondary progressive MS with significant disability. Given her complex needs, many health care professionals from a range of disciplines were involved in her care over the years, including physicians (general practice, neurology, psychiatry, rehabilitation medicine, plastic surgery, acute medicine, and intensive care), specialist nurses (multiple sclerosis, rehabilitation, and wound care), clinical psychologists, speech and language therapists, dietitians, physiotherapists, and occupational therapists. She was evaluated by multidisciplinary teams on at least seven separate occasions. However, despite her

extensive and progressive morbidity and low quality of life, at no stage was a referral to palliative care made.

Palliative Care Discussion

There had been some prior discussion about the goals of care. During her multiple admissions to the rehabilitation ward over the years, she had completed several life goals questionnaires before her cognitive dysfunction became too severe, which were appropriately considered in her package of care. On review of the medical record, the rehabilitation team had recommended full active care and treatment within the two years prior to the current admission. Although multiple health care providers were involved in her care subsequent to this, there was no further documentation about goals of care, even though her clinical condition continued to worsen. That being said, a do-not-attempt cardio-pulmonary resuscitation (DNA CPR) form was completed during an admission for bronchopneumonia the year prior and once again during the current admission after discussion by her admitting acute medical consultant with her husband and daughters.

During her last admission, the acute medicine team initially identified noninvasive ventilation for respiratory failure as the ceiling of care. Accordingly, she was admitted to the resuscitation room and her case discussed with an intensive care consultant. At that point, a discussion between the medical team and family about her baseline level of function and quality of life ensued with conflicting opinions between family members emerging. Some family members believed that she had some quality of life, thereby warranting full medical treatment, while others were less certain about this approach given her significant morbidity. After further discussion a few hours later and continued clinical decline, it was decided by the acute medicine and intensive care consultants (in consultation with the family) that noninvasive ventilation would not be in the patient's best interests. Early advance care planning with the palliative care team could have helped guide these discussions.

Clinical Course

She was admitted to the acute medicine service. A diagnosis of respiratory failure secondary to bronchopneumonia was made, prompting institution of

supplemental oxygen and intravenous co-amoxicillin /clavulanic acid. Given concern of pulmonary embolism, treatment dose dalteparin was also initiated. Initially, noninvasive ventilation was deemed as the ceiling of appropriate care should her respiratory failure worsen. However, given her significant debility and worsening clinical status, this was no longer seen as appropriate after a conversation with the family a few hours later. Her family also agreed that a DNA CPR with a focus on comfort was most appropriate at this stage.

Her respiratory function continued to deteriorate rapidly over the next several hours and she suffered respiratory arrest. Scant secretions were removed by suction and high-flow oxygen was administered. Cardiac arrest ensued. She was pronounced dead. Her family agreed to donate her brain and spinal cord for research and education

General Remarks

Early palliative care input and intervention may have helped focus her goals of care and outline coordinated strategies to facilitate them. Her quality of life was very poor. She lacked capacity to make her own decisions, could not interact meaningfully with those around her, and was completely dependent on others for her basic needs. Her care was fragmented and the overall goals of care were not clearly defined. The mobilization of a palliative care approach could have helped optimize symptom management and frame realistic expectations about care needs and outcomes. This strategy would likely have preempted emergent and distressing discussions about the use of noninvasive ventilation during the acute admission and may have likely put into context the value of PEG insertion at this stage of her disease. It might have also avoided a visit to the emergency department, with care instead being focused in the community. The palliative care team could have been an effective consulting service with regard to symptom management, goals of care, caregiver support, and end-of-life care (Figure 46.1).

Suggested Reading

1. World Health Organization. *World Health Organization Definition of Palliative Care*. www.who.int/cancer/palliative/definition/en/
2. Hawley PH. The bow tie model of 21st century palliative care. *J Pain Symptom Manage*. 2014 Jan;47(1):e2-5.

Section 6: Demyelinating Diseases and Autoimmune Neurology

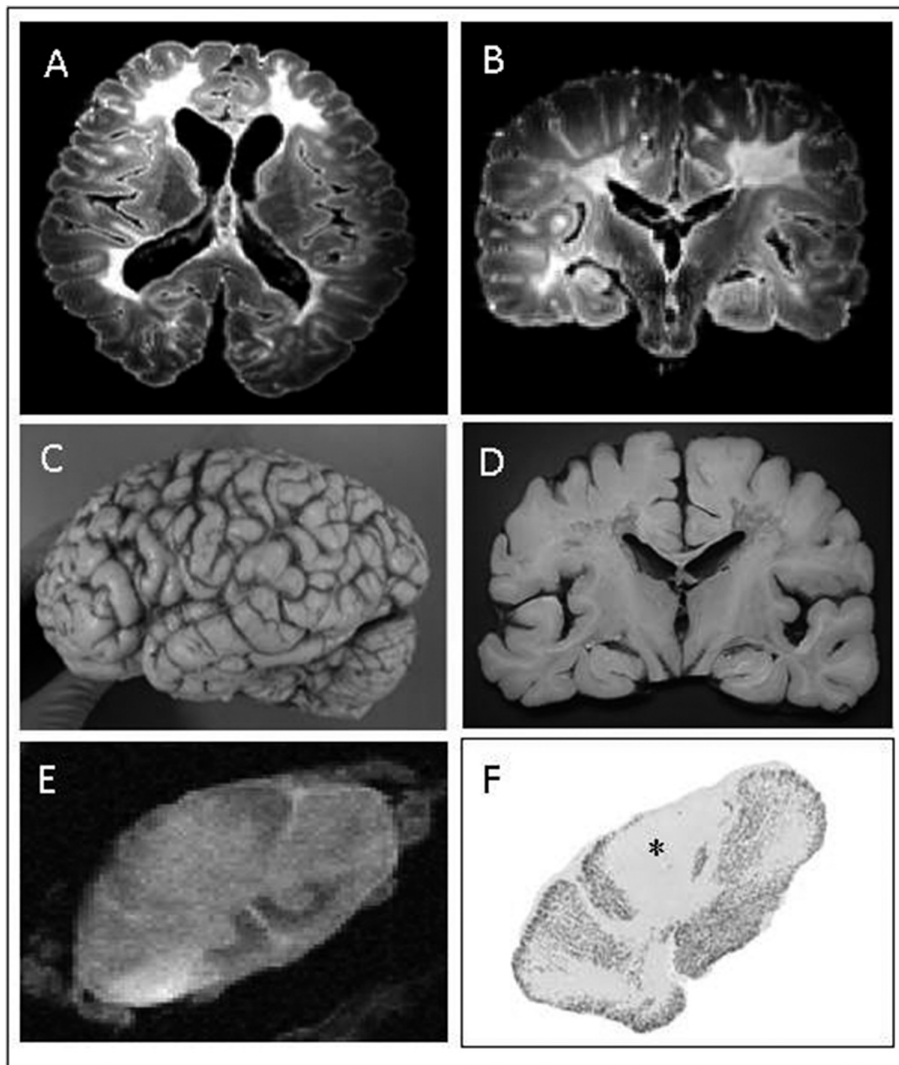


Figure 46.1 Radiographic, macroscopic, and microscopic views of the postmortem brain and spinal cord of the deceased patient featured in the case. T2-weighted MRI brain scans of the postmortem brain *ex situ* in the axial (A) and coronal (B) planes reveal extensive, confluent white matter lesions in the periventricular and juxtacortical regions along with considerable cerebral atrophy. Macroscopic view of the left lateral hemisphere (C) demonstrates diffuse cerebral atrophy with prominent sulci with the coronal view (D) showing greyish pallor of the periventricular white matter consistent with chronic demyelination characteristic of advanced, end-stage MS. Axial views of the postmortem spinal cord demonstrated by T2-weighted MRI obtained *ex situ* (E) shows extensive lesion burden involving both grey and white matter structures, which on histology (F) confirms demyelination (asterisk) by immunohistochemistry labeled with the myelin protein, proteo-lipid protein (myelin depicted in brown). The extensive MS lesion burden in both the brain and spinal cord was responsible for the significant, irreversible disability the patient experienced during life.

3. Lunde HMB, Assmus J, Myhr KM, Bo L, Grytten N. Survival and cause of death in multiple sclerosis: A 60-year-longitudinal population study. *J Neurol Neurosurg Psychiatry*. 2017 Aug; **88**(8):621–5. Epub 2017 Apr 1.
4. Strupp J, Romotzky V, Galushko M, Golla H, Voltz R. Palliative care for severely affected patients with multiple sclerosis: When and why? Results of a Delphi survey of health care professionals. *J Palliat Med*. 2014 Oct;**17**(10):1128–36.
5. The MS Society: MS and Palliative Care. www.mssociety.org.uk/sites/default/files/Documents/Professionals/MS%20and%20Palliative%20Care%20-%20guide%20for%20professionals.pdf
6. Veronese S, Gallo G, Valle A, et al. Specialist palliative care improves the quality of life in advanced neurodegenerative disorders: NE-PAL, a pilot randomised controlled study *BMJ Supportive & Palliative Care* Published Online First: July 16, 2015.