Prolonged Coma in Cervical Myelopathy with Dysautonomia

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Abstract

The term dysautonomia is used when there is a derangement in the reflex mechanisms of the limbic system resulting in improper control of pulse and blood pressure and even syncope sometimes. Cervical myelopathy may be complicated by dysautonomia and may rarely cause syncope; however, prolonged coma as a result of autonomic dysfunction in traumatic cervical myelopathy has not been described in the literature. We describe two patients with cervical myelopathy with dysautonomia who had prolonged coma, one of whom had to intubated multiple times and kept on ventilatory support.

Key words: Cervical myelopathy, Coma, Detrusor muscle dysfunction, Dysautonomia, Syncope.

Introduction

Dysautonomia is the dysfunction of the autonomic nervous system resulting in the derangement of the reflex mechanisms in the limbic system of the brain and brainstem resulting in improper control of pulse, blood pressure, intestinal motility, and sometimes syncope[1]. The fault may lie in the brain, brainstem, or in neurotransmission in the peripheral distribution of autonomic nerves. Postural hypotension as a result of autonomic dysfunction has been reported in patients with spinal cord injuries and tumor[2], syringomyelia[3] and acute transverse myelitis[4]. Cervical myelopathy may be associated with dysautonomia, but cervical myelopathy with dysautonomia leading to prolonged coma is a very rare event. Here, we describe two patients with severe traumatic cervical myelopathy with autonomic dysfunction presenting as prolonged coma requiring ventilatory support.

Case reports

Case 1: A 47-year-old quadriparetic, Chinese gentleman was brought to the hospital complaining of drowsiness of a few hours' duration. On further questioning the family told that the patient felt giddy initially and progressively became drowsy over a few hours. He was brought to the hospital, unconscious. According to his caregivers he had similar episodes in the past but on a few occasions he recovered spontaneously at home. On examination he was unconscious, afebrile, his pulse rate was 30/min, systolic BP was 60 mm Hg and he maintained normal oxygen saturation at room air. Neurologically his GCS was 3 (E1V1M1), pupils were equal and reactive, he was hyperreflexic and hypertonic in all four limbs, power in the upper limbs 3/5 and Lower limb 1/5 (MRC grade). In his previous admissions to our hospital he had similar events with variable severity, however, the most consistent findings noted were unconsciousness (GCS varying from 3 to 6) with bradycardia and hypotension.

Many years ago he had a traumatic cervical cord injury which resulted in quadriparesis. Since then, he is bed ridden with an indwelling catheter, but has normal mentation. His general condition is good without any signs of malnutrition. As he was deeply comatose in accident and emergency department, he was intubated to protect the airways and subsequently investigated in detail for possible metabolic causes that could account for his recurrent episodes of unconsciousness. The investigations revealed a normal hematological, biochemical and endocrine profile that included epinephrine, metanephrine, cortisol, thyroid profile and synacthen test. An initial EEG

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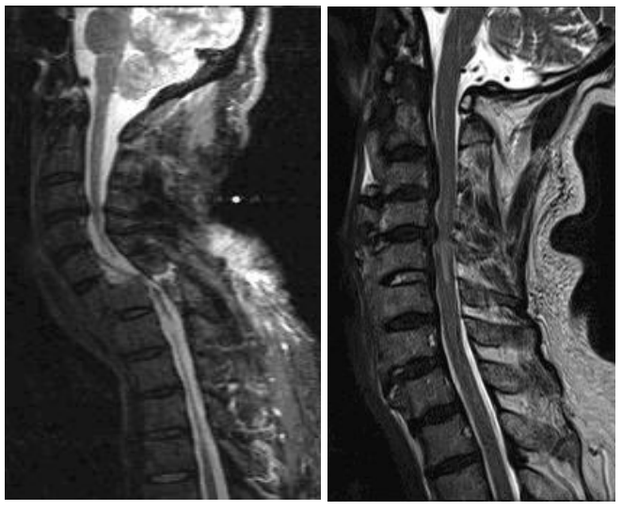

Fig 1. MRI Cervical Spine, Case 1 (left), Case 2 (right)

done at his presentation, revealed mild to moderate diffuse encephalopathy (suggestive of hypoxic or metabolic encephalopathy) and a repeat EEG after he regained full consciousness showed normal brain waves. The 24-hr Holter cardiac monitoring and the 2-D echocardiography were normal. DSE was negative for cardiac ischemia and MRI/MRA/DWI/FLAIR brain was normal. In all his previous hospitalization he had the same sequence of events: presenting initially with drowsiness followed by unconsciousness that required intubation and ventilation. After a few days with general supportive measures he became hemodynamically stable and regained consciousness. On reviewing his previous case notes it became obvious that the most probable cause of his repeated coma was autonomic dysfunction. This was subsequently proved with autonomic nerve testing which showed sudomotor dysautonomia. He could not complete autonomic nerve testing as he was not able to perform stand-up/tilt test. However, the temporal relation of hypotension with bradycardia and unconsciousness with reversal of mental state in presence of normal blood pressure and heart rate was in favor of dysautonomia as a cause of cerebral hypoperfusion leading to unconsciousness. During hospitalization, it was noted that manipulation and/or change of urinary catheter somehow improved his hemodynamic and mental status: his blood pressure and pulse rate improved with subsequent improvement in his level of consciousness. Urodynamic studies revealed detrusor instability. A therapeutic sphinchterotomy was performed with improvement of urine flow. Three years after sphinchterotomy he is symptom free and heamodynamically stable.

Case 2: A 68-year-old Malay gentleman was admitted with a history of dizziness which progressed to drowsiness and coma over a few hours. He was previously in good health and had a normal diet when he suffered from a road traffic accident leading to cervical cord injury 2 months ago. MRI of the cervical spine showed severe cervical spondylosis and cervical myelopathy with cord edema as a result of acute injury at C3 to C6. He underwent cervical laminoplasty of C3-C6 vertebrae. His weakness improved and his sensory loss resolved after the spinal surgery. However, about a week later he started having several episodes of hypotension with bradycardia requiring intravenous fluid challenges and inotropic support. He remained only mildly symptomatic throughout the episodes. He was subsequently maintained on salt supplements and fludrocortisone which was changed to Midodrin as he developed fluid retention. Following a period of rehabilitation, he was discharged with a urine catheter. However, two weeks after discharge, he was brought back to hospital in a state of stupor. He was found to be hypotensive and bradycardic. Neurological examination revealed a state of coma with upper motor neuron quadriparesis and urinary retention. He was investigated for possible cardiac, neurogenic, septic and metabolic causes of coma. The investigations revealed a normal hematological, endocrine and metabolic profile. The blood cultures were negative. The serial ECGs, cardiac enzymes and the 2-D echocardiogram were normal. General supportive measures with IV fluid resuscitation were started. Atropine was given and an indwelling catheter was inserted. A good hemodynamic response was noted; however he remained drowsy and unresponsive for more than 36 hrs. An MRI brain (stroke protocol) was done, DWI and ADC showed a faint diffusion restriction in both thalami and left mid brain (PCA territory) and MRA showed severe stenosis at the tip of basilar artery which was suggestive of a subacute 'tip of the basilar' syndrome. The transcranial doppler scanning of anterior and posterior circulation up to level of insinuation showed normal flow and velocities. The autonomic test later confirmed severe sudomotor dysautonomia by testing sympathetic skin response and inconclusive cardiovascular autonomic testing as he could not complete stand-up/tilt test due to severe weakness in the lower limb. The diagnosis of brainstem ischemia from hypoperfusion, secondary to autonomic dysfunction from a post-traumatic central cord syndrome was made. His mental status gradually returned to normal though he had weakness in all four limbs from previous insult to cervical cord.

Discussion

The cases described above show the occurrence of prolonged coma in the presence of autonomic dysfunction in two patients with traumatic cervical myelopathy. In the first patient, the state of prolonged coma was likely

due to failure of cerebral autoregulation as a result of autonomic dysfunction from cervical cord injury. The detrussor muscle instability caused retention of urine which subsequently resulted in worsening of dysautonomia. The dysautonomia improved after sphincterotomy. In this particular case the presentation of coma in the accident and emergency department was invariably followed by intubation. The coma resolved with inotropic support and catheterization of the bladder. If the initial condition of the patient was known it was possible that bladder catheterization and inotropic support only could have averted the invasive interventions. Laskari et al[5] described a patient with recurrent syncope in the context of a chronic dysautonomia but unlike our patient, neither was it associated with cervical myelopathy nor was there prolonged coma[5]. Symptoms of myelopathy had been present for the past several years, but the patient recently became severely symptomatic due to autonomic dysfunction that became worse with detrusor instability. Assessment of the autonomic function revealed impairment of circulatory reflexes and bladder instability which was consistent with pyramidal dysfunction.

In the second case, there was a stenosis of the tip of basilar artery which could have been a contributory factor to his prolonged coma, though Doppler study of posterior circulation was normal up to the level of insinuation. It is difficult to assess the contribution of a partially blocked catheter to coma, but the rapid improvement in mental status following bladder catheterization and improvement in hemodynamic status is suggestive of an association.

Other common conditions such as alcoholism, diabetes, uremia and associated malignancies can result in peripheral neuropathy and autonomic dysfunction. Amyloidosis can also result in prominent autonomic dysfunction in association with peripheral neuropathy.[6] Lack of vitamin B1 can exacerbate symptoms of dysautonomia that may improve with addition of vitamin supplementation. Mitral valve prolapse also can contribute to dysautonomia[7]. All these conditions were absent in both the patients described above.

Conclusion

These two cases highlight the importance of urinary retention as a cause of stupor and coma in patients with cervical myelopathy and autonomic dysfunction. Unblocking the urinary catheter /sphincterotomy and general supportive measures can be very useful in avoiding unnecessary investigations and invasive treatment such as intubation.

Take Home Message: Blocked urinary catheter can trigger dysautonomia in patients of traumatic cervical myelopathy. Prolonged coma as a result of autonomic dysfunction can easily be treated by just unblocking the catheter or by sphincterotomy which is a new observation. Hence it should always be taken into consideration to avoid unnecessary investigations and invasive treatment.

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