Autonomic dysreflexia in syringomyelia secondary to Chiari malformation

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Autonomic dysreflexia is a serious complication of high level spinal cord injury. Here, the authors present a case of autonomic dysreflexia in a patient with non-traumatic spinal cord pathology and review the incidence of such potentially fatal complications in other neurological conditions involving the high level spinal cord.

Autonomic dysreflexia is a well known complication of traumatic spinal cord injury. It is a phenomenon characterised by an imbalanced reflex sympathetic discharge that leads to potentially life-threatening hypertension. Although rare, this phenomenon has also been reported in the non-traumatic pathologies, including multiple sclerosis, and all patients with high level spinal cord involvement, from whatever pathology, should be educated about this condition. However, there is currently an under-recognition in the literature and clinical practice of autonomic dysreflexia presenting in the non-traumatic spinal pathologies.1

We present a case of a middle-aged gentleman referred to a neurorehabilitation department with poor mobility and autonomic dysreflexia that was linked to secondary syrinx Chiari malformation, which was a diagnosis that was initially missed by the acute medical teams.

Presentation
A 55-year-old man was referred for medical assessment from the community neurorehabilitation team. He had a thirty-year history of deteriorating mobility secondary to an extensive syrinx and Chiari malformation. Although he had previously had decompressive surgery and insertion of a syrinxopleural shunt, eventually he had become wheelchair bound and dependent for his care needs. A recent MRI scan had shown a syrinx extending throughout the cord, which was generally atrophic.

Prior to review, the patient had been admitted twice under the acute medical team at his local hospital with severe pounding headache, sweating, anxiety and flushing of the face. These symptoms were associated with a urinary catheter blockage at one incident and by severe constipation in the other incident. He was found to be hypertensive (BP 200/110mmHg) and his electrocardiogram showed atrial fibrillation. He was consequently diagnosed with paroxysmal atrial fibrillation secondary to a urinary tract infection. He was treated with bisoprolol and antibiotics, and discharged after a few days. He was to be followed up by his cardiologist with a view to starting anticoagulation.

The neurorehabilitation team recognised this phenomenon as autonomic dysreflexia. An alert card was given and the patient was educated about the condition. Nifedipine was prescribed as needed, and trigger factors, including his problematic neurogenic bladder, were addressed. His cardiologist was advised of the risks of anticoagulation, which would routinely be recommended in paroxysmal atrial fibrillation, but would be dangerous in autonomic dysreflexia (characterised by episodic severe hypertension) because of the increased risk of brain haemorrhage. After assessment of risk versus benefit of anticoagulation, including a discussion with the patient, a decision was made not to commence anticoagulation.

The patient managed his evolving autonomic dysreflexia attacks appropriately at home and has not needed any further inpatient admissions.

Discussion
Autonomic dysreflexia is an imbalanced reflex sympathetic discharge leading to potentially life-threatening hypertension and is a well known complication of spinal cord injury. Often it occurs in individuals with an injury at or above T6.

Autonomic dysreflexia is often precipitated by a strong sensory input from a level below the spinal cord injury. The intact peripheral nerves carry the signal to the spinal cord where it initiates a strong reflex sympathetic surge resulting in peripheral vasoconstriction leading to systemic hypertension.2 The cervical baroreceptors detect the increased blood pressure (BP) and alert the hypothalamic centres. The brain’s attempts to reduce BP via reducing the sympathetic surge are in vain as the down-bound impulses are blocked by the spinal injury. A simultaneous vagus nerve stimulation aiming at reducing the BP by slowing down the heart may lead to the classic autonomic dysreflexia presentation of severe hypertension in conjunction with bradycardia or other forms of arrhythmia.3

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Autonomic dysreflexia is a medical emergency and if left untreated can cause seizures, retinal haemorrhage, pulmonary oedema, renal insufficiency, myocardial infarction, cerebral haemorrhage and death. Autonomic dysreflexia is known to occur in those patients who have high spinal cord involvement from medical pathologies including multiple sclerosis. Nevertheless there is under-recognition of this risk in the literature and clinical practice.

We believe this is the first case in the literature linking autonomic dysreflexia with syringomyelia secondary to Chiari malformation. However, there is case report evidence of autonomic dysreflexia presenting in the non-traumatic pathologies of encephalomyelitis, spinal cord sarcoidosis and multiple sclerosis; with spinal cord involvement. Despite this, awareness of autonomic dysreflexia in the non-traumatic pathologies is limited. This is demonstrated by an interesting survey of multiple sclerosis clinicians, which found that although 10% had encountered autonomic dysreflexia complicating multiple sclerosis in their clinical practice, a much larger 45% were not even aware of the possibility of this occurring.

In this case, the patient also presented with atrial fibrillation. Autonomic dysreflexia is usually associated with bradycardia, although tachycardias are not uncommon. There is, however, only limited case report evidence suggesting that it can present in association with atrial fibrillation, and this is only in traumatic spinal cord injury. Recognition of autonomic dysreflexia in this patient was important since there was a high chance they could be anti-coagulated, which in combination with uncontrolled hypertension would likely result in devastating consequences.

In conclusion, autonomic dysreflexia can be difficult to diagnose by specialists who may not be accustomed to treating it, particularly if it presents in the non-traumatic spinal pathologies. However, although rare, this condition is serious and must be recognised and treated appropriately. Therefore, it is important that awareness of autonomic dysreflexia is disseminated throughout neurological and general medical practice.

**Declaration of interests**

No conflicts of interest were declared.

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**References**