CASE REPORT

A Report on an Unusual Presentation of Autonomic Dysreflexia

Siti Suhaida Sudin, MD(UKM), Julia Patrick Engkasan, MRehabMed(Mal)

Department of Rehabilitation Medicine, University of Malaya, Faculty of Medicine, Lembah Pantai, 59100 Kuala Lumpur

SUMMARY

A 48-year-old male with complete tetraplegia C6 presented with sweating and flushing of the right half of the face and neck that recurred when lying in supine and left lateral positions. The symptoms subsided immediately upon sitting upright or lying in a right lateral position. The symptoms were associated with occasional mild head discomfort rather than headache and were accompanied by marked elevation of blood pressure, which was 190-200/120-130 mmHg compared to his previous baseline blood pressure of 80-90/50-70 mmHg, and he had a heart rate of 60-70 beats per minute. We believe that post-traumatic syringomyelia, found upon further investigation, was the cause of the Autonomic dysreflexia (AD) in this patient. He was advised to avoid the positions causing the symptoms and the progression of symptoms was monitored regularly. AD might not have been diagnosed in this patient because of the atypical and unusual presentations. Therefore, knowledge and a heightened level of awareness of this possible complication are important when treating individuals with spinal cord injury (SCI).

KEY WORDS:
Spinal Cord Injury; Autonomic Dysfunction; Hyperhidrosis; Syringomyelia

INTRODUCTION

Autonomic dysreflexia (AD) is an acute syndrome of excessive, uncontrolled sympathetic discharge with potentially serious consequences that can occur in individuals with spinal cord injury (SCI) above T6 level[1]. Exposure to noxious stimuli below the level of injury triggers vasoconstriction and a subsequent increase in blood pressure[1]. AD typically presents with bilateral pounding headache, flushed sweaty skin above the neurological level, nasal congestion, malaise, nausea and blurring of vision[1]. The common triggers are from urinary tract, bowel and musculocutaneous pathology, deep vein thrombosis, pulmonary embolism and syringomyelia[2].

A raised blood pressure of 20-40mmHg above the patient's baseline is considered significant[3] and as the normal systolic blood pressure for T6 is 90-110 mmHg, not all patients will present with markedly elevated readings. Failure to properly manage AD can result in severe consequences such as cerebral or subarachnoid hemorrhages, seizures, atrial fibrillation, neurogenic pulmonary edema, retinal hemorrhage, and coma or death[4]. Herein, we present a patient with tetraplegia who developed recurrent AD with unusual presentations secondary to post-traumatic cervical syringomyelia.

CASE REPORT

A 48-year-old man with a traumatic C6 American Spinal Injury Association (ASIA) Impairment Scale (AIS) A, sustained 29 years previously, presented with profuse sweating and flushing of the right half of the face and neck for 2 months. The symptoms only occurred when lying in supine and left lateral positions. The symptoms subsided immediately on sitting upright or when lying in a right lateral position. Occasionally, symptoms were associated with mild head discomfort rather than headache.

In supine position, there was a prominent right-sided facial hyperhidrosis and flushing accompanied by marked elevation of blood pressure, ranging from 190-200/120-130 mmHg, and he had a heart rate of 60-70 beats per minute. From previous records, his baseline blood pressure was 80-90/50-70 mmHg. Despite the high blood pressure, he remained otherwise asymptomatic. When he changed to lying in a right lateral position or was seated upright, the sweating and flushing ceased. This was followed by reduction in blood pressure. On the next visit, this signs were reproducible with the same positional changes.

No precipitant of AD was found during physical examination. Physical examination ruled out musculocutaneous pathology such as pressure ulcer, in-growing toe nail or spaticity. Laboratory and radio-imaging studies were negative for urinary tract infection, urinary system pathology or intra-abdominal pathology, and a recent urodynamic study showed a hypocompliant bladder with low pressure. Magnetic resonance imaging (MRI) of the spine revealed the old C5 and C6 vertebral fracture and a transected cord at the C6 level. There was a syringomyelia noted from level C7 to T1 (Figure 1). As we have ruled out other common causes of AD, we concluded that his AD symptoms were caused by the syringomyelia. He was treated conservatively at the time and was advised to avoid precipitating positions to prevent the recurrence of AD. During his out-patient follow up 2 months later, his symptoms were still well controlled at home with the avoidance of the precipitating posture. There was no deterioration in his neurological or functional status.

This article was accepted: 22 June 2012
Corresponding Author: Julia Patrick Engkasan, Department of Rehabilitation Medicine, University of Malaya, Faculty of Medicine, Lembah Pantai, 59100 Kuala Lumpur Email: julia@ummc.edu.my

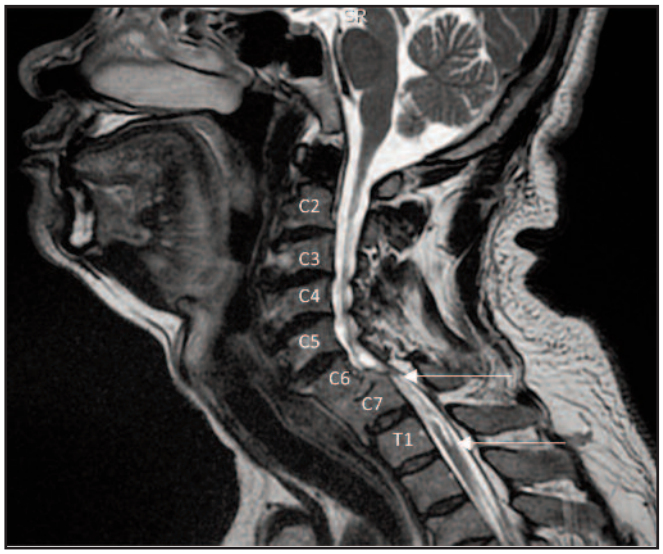

Fig. 1: T2 weighted non-contrast MRI result showing syringomyelia (line arrow) from the C7 to T1 levels and the presence of old vertebrae fractures (C5 & C6) with cord transection.

DISCUSSION

Here we present an unusual cause of AD in a chronic traumatic spinal cord injured individual who presented with atypical symptoms. Hyperhidrosis is one of the complications that can occur after a SCI either as an isolated etiology, as part of AD, as a manifestation of post traumatic syringomelia (PTS) or posttraumatic myelomalacic myelopathy (PPMM)[4,5]. We believed that the unilateral facial hyperhidrosis observed in our patient is due to AD rather than due to PTS or PPMM.

The most important clinical feature that distinguishes presentation of AD in this case from early PTS or PPMM is the presence of blood pressure (BP) changes during the event. PTS or PPMM pathology are not associated with blood pressure changes. Another point to support our diagnosis of AD is the pattern of hyperhidrosis. In patients with PTS, hyperhidrosis occurred during the upright position and changing to supine or would instantly stop on recumbent position [4]. However in our case, hyperhidrosis occurred on lying down and ceased upon upright position. Although PPMM itself can cause AD, there was no radiological feature such as cord lengthening or cord tethering to suggest the presence of PPMM seen in this patient's MRI scan [5]. Furthermore, most patients who had AD as the results of PPMM would have evidence of neurological deterioration which was not present in our case [5]. Although somewhat speculative, we think his position dependent AD was related to changes in pressure with different positions related to the syringomyelia. By lying supine or in left lateral position, the syrinx in the spinal cord could have irritated the spinal cord and during upright position, this irritation or pressure was eliminated.

Headache is an established symptom of AD and patients usually present with a bilateral pounding headache as a result of high blood pressure[1]. This patient, however, only experienced mild head discomfort despite the markedly elevated blood pressure. This presentation of AD could have been easily missed if the index of suspicion was low. Therefore, it is imperative that clinicians rule out AD when high risk patients present with symptoms that are not yet explained. They must also be aware of an array of possible presentations with this condition.

Pain and an ascending sensory level was the most common presentation of PTS, whereas increasing motor deficits, hyperhydrosis and increased spasticity were present less often[4]. The clinical diagnosis of PTS should be confirmed by MRI, which gives an accurate non-invasive assessment of the longitudinal extent and distension of the syringomyelia[4]. Surgery is most likely useful when there is a progressive deficit and should be deferred if the patient is not deteriorating. Our patient is able to control his symptoms by avoiding certain positions. Therefore, we decided to manage his condition conservatively with regular monitoring for any signs of neurological deterioration.

REFERENCES

1. Blackmer J. Rehabilitation Medicine: 1.Autonomic dysreflexia. CMAJ. 2003; 169(9): 931-5.
2. Lindan R, Joiner F, Freehafer A, Hazel C. Incidence and clinical features of autonomic dysreflexia in patients with spinal cord injury. Paraplegia. 1980; 18: 285-92.
3. Karlsson AK. Autonomic dysreflexia. Spinal Cord. 1999; 37: 383-91.
4. Glasauer FE, Czyrny JJ. Hyperhydrosis as the presenting symptom in the post-traumatic syringomelia. Paraplegia 1994; 32: 423-9.
5. Falcone S, Quencer RM, Green BA, Patchen SJ, Post MJD. Progressive posttraumatic myelomalacic myelopathy : Imaging and clinical features. AJNR Am J Neuroradiol. 1994; 15: 747-54.