CASE REPORT

Orthodeoxia syndrome: a rare complication of ASD in spinal cord injury

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STUDY DESIGN: This is a single-subject case report.

OBJECTIVES: The objective is to describe and discuss a rare presentation of orthodeoxia after spinal cord injury in a previously healthy adult with an asymptomatic and undiagnosed atrial septal defect.

SETTING: This study was conducted at the Tertiary Care Spinal Injuries Center in Sheffield, UK.

METHODS: An 81-year-old woman who was otherwise healthy and active sustained a polytrauma following a road traffic collision. Her injuries were managed conservatively. As a complication of epidural analgesia, she developed a complete thoracic paraplegia secondary to an epidural hematoma. She was found to have repeated episodes of severe deoxemia, without any significant postural hypotension when attempts were made to sit her up in bed. Detailed investigations were done to ascertain the cause for this deoxemia.

RESULTS: Echocardiography revealed an atrial septal defect; agitated saline contrast transesophageal ultrasound done in various positions revealed interatrial defect with reversal of shunting when the patient was made to sit up in bed.

CONCLUSION: Orthodeoxia is an uncommon finding in spinal injury patients. Detailed investigations at an early stage of rehabilitation need to be done to rule out cardiac causes after ruling out the common medical complications seen in the acute and subacute phases following spinal cord injury.

Spinal Cord Series and Cases (2015) 1, 15005; doi:10.1038/scsandc.2015.5; published online 9 July 2015

An 81-year-old woman with controlled hypertension presented to the emergency department following a road traffic collision. She sustained a stable C2 and multiple rib fractures. On examination, she had no neurological deficits. As she was immobilized, she was placed on prophylactic anticoagulation with low-molecular-weight heparin. The pain was managed by epidural patient-controlled analgesia for 4 days. On the seventh day following injury, she developed complete flaccid paralysis of both lower limbs with bilateral sensory loss below T8 (ASIA Impairment Scale-A). An MRI confirmed an intradural extramedullary hematoma compressing the spinal cord between T6 and the conus with signs indicating a venous infarct. This was managed conservatively.

After 6 weeks of immobilization, reimagining confirmed satisfactory union of her C2 fracture. She was mobilized and racked up in bed to 30 degrees, during which the oxygen saturation (SpO₂) dropped to 76%. This was not associated with dyspnea, chest pain or dizziness. There were no changes to her blood pressure and heart rate to suggest postural hypotension. Oxygen saturation returned to normal with supine position. CBC, serum chemistry and chest X-ray were normal. D-dimer was within normal range. CT pulmonary angiogram ruled out pulmonary embolism. ECG ruled out ST-elevation myocardial infarction. The echocardiogram done in supine position showed an atrial septal defect (ASD) measuring 12 mm (Figure 1). Estimated pulmonary artery pressures were within normal limits. Pulmonary function tests were not done.

Saline contrast transthoracic echocardiogram was done with the patient lying down and sitting up. In sitting position, agitated saline load in the left heart in the third cardiac cycle was noted,

which suggested right-to-left shunting. SpO₂ dropped to 76% and improved to 96% on lying down. Transesophageal echocardiography showed an ASD with reversal of flow evident in sitting up position (Figures 2 and 3). This positional reversal of flow was the probable cause of the orthodeoxemia.

The patient underwent percutaneous closure of the defect with a 25 mm Intrasept device. After the procedure, she had complete resolution of her symptoms and completed her rehabilitation to become partially independent in a wheelchair.

DISCUSSION

Studies have shown that following acute spinal cord injury (SCI), patients develop some degree of postural hypotension when they are mobilized or tilted to vertical positions. After SCI, reflex vasoconstriction is blunted causing an excessive pooling of blood in the abdominal viscera and lower extremities, causing hypotension, reflex tachycardia and other symptoms of orthostatic hypotension but rarely a significant deoxemia.^{1–3} In our patient, sitting posture did not cause any postural hypotension, but there was deoxemia, which did resolve once she returned to supine position. One of the differentials considered was orthodeoxia, and was investigated.

Orthodeoxia syndrome is uncommon in SCI patients. Arterial desaturation and dyspnea in the upright posture, which resolves in the supine posture, are the hallmark findings in this syndrome. Our patient had both the anatomical and functional components of this syndrome, that is, the interatrial shunt and the fact that reversal of flow in the shunt was triggered by change to upright

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Received 4 July 2014; revised 18 December 2014; accepted 15 January 2015

Figure 1. ECHO showing interatrial septal defect.

Figure 2. ECHO in supine position without significant right to left shunting.

Figure 3. Echo in sitting position with significant shunting from right to left.

posture. Autopsy studies have reported an incidence of probe patent foramen ovale (PFO) as high as 27%. With transthoracic

echocardiography, the detection rates are between 10 and 15%.⁴ This increases to ~57% with agitated saline contrast imaging, and to 92% with transesophageal echocardiography with Valsalva maneuver.⁵ Positional right-to-left inter-atrial shunting through a PFO causing orthodeoxia has previously been described in able-bodied patients with normal pulmonary artery pressure and normal heart function.⁶ We do not know whether this patient had orthodeoxia before her SCI, as she was asymptomatic. Alteration in the positional relationship of the inferior vena cava (IVC) to the PFO with postural change from supine to upright has been postulated to be the cause for reversal of the shunt.⁶ Grutters *et al.*⁷ described a case in which the superior vena cava (SVC) was involved. They showed a change of blood flow direction from the SVC to the PFO when position changed from supine to upright position.

It is likely that the subject of this report had asymptomatic orthodeoxia before SCI. The injury at T8 level caused paralysis of abdominal musculature, resulting in a lowering of the diaphragm and mediastinum. This could change the orientation of the heart and the directional relationship of the IVC to the ASD, causing the deoxygenated blood from the vena cava to directly shunt from the right to left. As the SVC is generally intrathoracic and high up, it is hard to imagine it shifting in alignment. However, the IVC is intra-abdominal, and it is more likely to shift in alignment. IVC carries more desaturated blood, and hence it is more likely to precipitate symptoms of deoxemia in such instances. This could have been the cause for deoxemia to have become apparent when she was sitting.

To the best of our knowledge, this is the first case involving a person with SCI having positional orthodeoxia owing to an ASD. The orthodeoxia vanished after repair of the ASD, proving the cause-and-effect relationship. Consequently, we recommend screening all new SCI patients for orthodeoxia as soon as they are able to tolerate the sitting position.

COMPETING INTERESTS

The authors declare no conflict of interest.

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