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Case Report

Incomplete Spinal Cord Injury With Concurrent Hypertrophic Obstructive Cardiomyopathy

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A B S T R A C T

Traumatic spinal cord injuries are significant contributors to the morbidity and mortality burden of trauma patients worldwide, and consume significant resources in both their acute and rehabilitative care. Another cause of mortality and morbidity is hypertrophic obstructive cardiomyopathy, which can cause syncope or sudden cardiac death in patients with no known prior cardiac disease. This case report describes a unique combination of these two high-risk pathologies in a scene trauma patient, and provides an overview of the pathophysiology and treatment of these high-risk disease processes.

Although relatively rare, traumatic spinal cord injuries (SCIs) are a cause of significant morbidity and mortality among trauma patients and represent significant resource utilization in their acute and rehabilitative care.[1] Similarly, hypertrophic obstructive cardiomyopathy (HOCM) poses a risk of syncope or sudden cardiac death in patients with no known prior cardiac disease.[2] This case report describes a unique combination of these 2 high-risk pathologies in a scene trauma patient and a review of the pathophysiology and treatment of these disease processes.

Case Report

A helicopter emergency medical service (HEMS) team was dispatched to rendezvous with an advanced life support emergency medical services (EMS) unit at an improvised landing zone in a community church parking lot. The dispatch was for a patient reported to have fallen at home, with no further information available at the time of lift off from the base. Upon arrival to the landing zone, the flight crew entered the waiting ambulance and discovered a 64-year-old woman secured to a long backboard with towel rolls beside her head as modified spinal precautions. The EMS providers reported that the patient's husband had witnessed her returning from the bathroom when she suddenly fell forward, striking her face against the bathroom door jam. The patient's husband stated he believed the patient had “passed out” before falling and that after landing on the ground, she experienced a period of unresponsiveness lasting approximately 5 minutes. Upon EMS arrival to the scene, they found the patient to be alert and oriented but lying prone in the position in which she had fallen. The EMS crew noted a heart rate (HR) in the 30s to 40s but did not have an opportunity to capture a 12-lead electrocardiogram. The patient reported being unable to move any of her extremities at this time. Recognizing the potential severity of the patient's condition, the ground providers elected to activate the nearest HEMS resource for rapid treatment and transport to the regional level 1 trauma center.

When the flight crew completed their assessment at the landing zone, they found a patient with a Glasgow Coma Scale score of 15 with equal and reactive pupils supine on a backboard with improvised cervical spine control present. The primary survey revealed an airway that was patent and intact without need for intervention, breathing that was regular and tachypneic at a rate of 24 breaths/min without the use of accessory muscles, profound bradycardia and hypotension with HR of 38 and blood pressure (BP) of 60/20 mm Hg, and evidence of poor perfusion with cold skin and diaphoresis. The patient was generally restless, crying aloud as providers touched anywhere on her body, and was able to correctly identify sensation to the bilateral lower limbs but was unable to move her limbs, wiggle her toes, or grasp with either hand. There was global hypotonia and flaccidity on a brief neurologic examination. The only obvious

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external trauma to the head or face was limited to an abrasion and swelling about the left orbit. Despite hypersensitivity to touch or palpation of her extremities, the patient did not complain of any pain or point tenderness of her cervical spine upon examination.

The patient was placed on the flight cardiac monitor, a rigid cervical spine collar replaced the improvised device, and the patient was carefully transferred to the aircraft stretcher. Once safely loaded into the aircraft, per flight program patient care guidelines, 1 L normal saline was hooked up to the patient's intravenous (IV) line and placed under pressure with the goal of improving the BP, and 4 L supplemental oxygen via a nasal cannula was initiated to correct for altitude-induced hypoxemia. In flight, a 12-lead electrocardiogram was obtained by the crew that showed ST-segment depression and deep T waves in the inferior and lateral leads.

There was a slight improvement in cardiac output after fluid challenge with HR of 47 and BP of 71/40 mm Hg. Despite this volume resuscitation, the patient had ongoing bradycardia and hypotension, so a 0.5-mg atropine IV dose was administered per patient care guidelines without effect. The patient was given 25 μg fentanyl for ongoing hyperesthesia of the limbs. A radio alert with trauma notification was sent to the receiving regional level 1 trauma center, and helipad availability and security presence were confirmed. In the last few minutes of flight, a further 0.5-mg atropine IV dose was given, with a noted increase in P-wave amplitude but no change in HR. The crew gave consideration to transcutaneous pacing per the advanced cardiac life support guidelines, however, they deferred because of the patient's profound hypersensitivity. Similarly, with the patient's appropriate mentation, the decision was made to focus on ensuring adequate volume resuscitation before commencing vasoactive agents. Upon landing at the receiving pad, the patient was taken immediately to the trauma bay where a bedside report was provided to the waiting trauma team.

On arrival to the emergency department, the patient was greeted by the trauma team, with rapid assessment and treatment provided. The patient was determined to have ongoing bradycardia and hypotension with HR of 42 beats/min, BP of 70/39 mm Hg, respiratory rate of 21 breaths/min, peripheral capillary oxygen saturation of 96% on room air, and a Glasgow Coma Scale score of 15. The patient continued to endorse hypersensitivity to all 4 limbs, most profound in the bilateral upper extremities, and a complete absence of motor function in the bilateral lower limbs. The trauma team continued fluid resuscitation with 1 L NaCl, initiated 1 unit of packed red blood cells because of equipoise about possible internal bleeding, and started a norepinephrine infusion to improve vascular tone. A focused assessment with sonography in trauma was determined to be negative, and a computed tomographic scan of the patient's head revealed no acute intracranial abnormality or acute fracture of the cervical spine; a stat neurosurgery consult was requested because of signs of neurogenic shock with quadriplegia. Neurosurgery attended and confirmed a diagnosis of spinal cord injury with resulting neurology and ordered stat magnetic resonance imaging (MRI). MRI revealed multilevel spinal stenosis with evidence of interspinous ligamentous injury between C4 and C7. She was diagnosed with an incomplete SCI with resulting described neurology attributed to acute traumatic central cord syndrome (ATCCS), with a predisposition to this injury pattern caused by radiologically detected age-related cervical stenosis.

In concert, the patient was investigated for the cause of her syncope. Echocardiography revealed left ventricular outflow tract obstruction (LVOT) and a 70% left anterior descending coronary artery lesion. This underlying HOCM was hypothesized to have caused the fall with the subsequent described trauma.

Discussion and Review of Incomplete SCI and HOCM

SCI

Traumatic SCI is a disruption of, or disturbance to, the central pathway of the body's nervous system and results in sensory, motor, and autonomic function changes.[2] These changes can have significant impacts on an affected individual's social, psychological, and physical health.[2] Protected by the bones of the spine, injury to the spinal cord is the result of significant kinetic injury forces transferred by trauma and is often associated with other injuries. The prevalence of SCI varies significantly by geographic region with a range of prevalence reported including 11.5 to 13.3 per million in the Republic of Ireland, 250 per million in France, and the highest prevalence in the United States at 906 per million, with average crude annual incidences between American states of 83 per million (Alaska) and 29.4 per million (Alabama).[2-4] The reported mortality associated with SCI is quite high, with 48.3% to 79% perishing before arrival at the hospital and a further 4.4% to 16.7% dying from their injuries during the course of their hospital stay.[5]

The mechanism of injury for SCI can be split into 2 separate processes described as primary and secondary mechanisms.[1] Primary SCI is the result of the initial impact, transfer of kinetic energy, and subsequent compression of the cord caused by fractured, dislocated, or subluxated vertebrae.[1,5] These vertebrae can be disrupted as a result of flexion, extension, axial loading, distraction, and rotational forces acting on the osteoligamentous spinal column with failure of the protective structures.[1,5,6] The secondary injury is a much more complex physiological pathway that involves glutaminergic mechanisms, the inhibition of intracellular protein synthesis, apoptosis, and the sequelae of the primary mechanism.[1,6] As a result of the compression forces acting on the spinal cord, there is localized cord ischemia, neurogenic shock causing worsening perfusion and increasing ischemia, microcirculatory changes, thrombosis, and vasospasm with subsequent loss of motor neurons.[5,7] This physical result of the trauma causes intracellular derangements including increased sodium permeability, increased intracellular calcium, decreased extracellular potassium, catecholamine and serotonin accumulation, and excitotoxic cell injury from extracellular glutamate.[5] There is also free radical and arachidonic acid production, inflammation, edema, impairment of the adenosine triphosphate −dependent process of cellular energy production, and ultimately apoptosis.[3,5,6]

The physical findings associated with SCI can vary greatly depending on the level of injury and the involvement of motor and/or sensory tracts, and it requires a savvy clinician to make the prehospital diagnosis in the absence of computed tomographic imaging or MRI. The most common form of SCI without complete transection of the cord is ATCCS and is the result of hyperextension of the neck.[8-10] ATCCS is most common in elderly patients with preexisting cervical stenosis and is characterized by limb weakness greater in the upper limbs than the lower, with weakness and loss of dexterity most profound in the hands.[10] Classically, there is a loss of thermal and pain sensation in a bilateral “cap” distribution over the shoulders and upper extremities, with intact anal sphincter tone because of sacral sparing and variable bladder involvement dependent on the neurons involved.[10] It is thought that the greater severity of weakness in the upper limbs over the lower is caused by the greater involvement of the corticospinal tract in arm and hand use than in leg and foot use.[8] It has been suggested recently that a true diagnosis of ATCCS requires a difference of 10 points in strength between the upper and lower limbs on the American Spinal Cord Injury Association scoring system.[8,10]

The specific primary mechanism for the subset of SCI patients experiencing ATCCS is the pinching of the cord by the narrowing of the canal as the anterior ligamentum flavum moves posteriorly

during a hyperextension event.[10] The cord is physically compressed, with subsequent central cord edema and lateral corticospinal white matter damage.[10] This damaging event in the younger population is likely to be caused by high-velocity blunt trauma and falls in the elderly.[10]

The diagnosis of SCI is classically made with radiographic imaging, with T2-weighted MRI being the modality of choice.[11] The use of advanced imaging is not available or practical in the prehospital arena, so knowledge of the prodrome of SCI and clinical gestalt is required for the provider to make appropriate immobilization and treatment decisions. Having made the determination that the patient is suffering from SCI, it is imperative that the providers make the decision to transport the patient to an appropriate receiving facility to avoid the delays to definitive care associated with a secondary transfer. It may be appropriate for the prehospital crew to treat hypotension with their neurogenic shock protocols, which can include crystalloid administration to fill vasoplegic vessels, alpha-agonist vasoactive drugs to improve peripheral vascular tone, and atropine or isoproterenol for increased heart rate.[12] It is appropriate to target a higher mean arterial pressure between 85 and 90 mm Hg to ensure adequate spinal cord perfusion.

The prognosis for patients suffering from ATCCS can be quite good.[10,13,14] One retrospective electronic chart review found that patients under the age of 65 years were more likely to have a good functional outcome, defined as the ability to walk unaided and the absence of neuropathic pain.[10,14] The ability to spontaneously void one's bladder was not correlated with age or sex but with a Functional Independence Measure score of greater than 81.[10] There is clinical equipoise in the literature as to whether surgical or conservative management provides for better patient-oriented outcomes in ATCCS; however, outcomes are reported to improve with intensive rehabilitation and therapy.[10]

HOCM

Hypertrophic cardiomyopathy is an inherited myocardial disease marked by ventricular wall thickness > 15 mm and is categorized into the more common HOCM and the less common nonobstructive hypertrophic cardiomyopathy (HCM).[15] The 2 subtypes are differentiated by the presence, or absence, of LVOT, either at rest or upon provocation on echocardiographic examination.[16] This myocardial disease is the most common hereditary cardiac condition and is not marked by clear geographic, sex, or ethnic patterns.[16,17] It is recognized that HCM and HOCM are significant causes of sudden cardiac death and heart failure pathology in not only the older populations more commonly associated with cardiac disease but also young healthy individuals and can lead to severe functional disabilities across the life span.[11] Echocardiographic population studies have shown a current prevalence for HCM of 1:500.[17]

The obstruction in HOCM is most often noted at the LVOT as the mitral apparatus moves anteriorly toward the hypertrophied septum during systole but can also occur similarly in the midventricular wall of the left ventricle (LV) as the ventricles contract.[15] It is estimated that approximately 30% of patients with HCM will have LVOT obstruction when at rest, with an exacerbation of the obstruction with exercise.[17] Those patients with severe LVOT or midventricular obstruction are more likely to have elevated ventricular diastolic pressures with dyspnea on exertion and can progress to florid heart failure.[17] It is these patients who can experience syncope during or immediately after exertion or exercise with or without a fatal or nonfatal ventricular arrhythmia.[17] Most patients with HCM will have varying degrees of mitral valve abnormality including elongation of the mitral leaflets, displacement of the papillary muscles anteriorly, and abnormal attachments of the chordae and structures, which can result in mitral regurgitation with a detrimental effect on their cardiac status.[18]

The signs and symptoms of HCM and HOCM are worryingly similar to other cardiac complaints; typically, the symptomology includes chest pain, dyspnea, palpitations, and syncope.[15] Patients with more progressed disease and severe outflow obstruction are prone to heart failure, atrial fibrillation, ventricular arrhythmias, and sudden cardiac death.[15,17] Patients with more severe disease have an annual mortality in the range of 1% to 5%.[19] Diagnosis is usually made first clinically and then confirmed with imaging such as cardiac MRI or echocardiography.[20] The most sensitive diagnostic test for HCM and HOCM is the 12-lead electrocardiogram; only 5% to 10% of patients with the condition have a normal electrocardiogram on assessment.[20] Typically, the electrocardiographic abnormalities will include left ventricular hypertrophy, ST-segment changes, left atrial enlargement, Q waves, absent or diminished R waves in the lateral leads, or T-wave inversion in 75% to 95% of cases.[20] Clinical suspicion is most often confirmed with focused cardiac ultrasound with examination of LVOT gradients, mitral valve anatomy and function, systolic and diastolic function, and left ventricular hypertrophy.[21]

Emergency management of HOCM revolves around increasing preload, afterload, and filling time because these processes allow for enlargement of the LVOT with an improvement in forward flow.[22] Similarly, interventions that decrease preload and afterload and shorten filling time all worsen the LVOT and reduce the blood flow out of the left ventricle.[22] As such, vasoactive agents that increase inotropy or chronotropy should be stopped or avoided, and the volume should be optimized with crystalloid solutions to ensure adequate preload. Afterload can be increased with alpha-agonists, such as norepinephrine or phenylephrine, and beta-blockers can be used with careful consideration to decrease HR to augment diastolic filling.[22]

Long-term treatment for HCM and HOCM exists on the spectrum from surgical therapy to lifelong pharmacologic management.[21] Patients who have a single episode of atrial fibrillation will be placed on lifelong anticoagulation, those who have experienced ventricular arrhythmias will likely have a cardiac defibrillator implanted, and patients with severe outflow obstruction will initially be placed on beta-blockers or calcium channel blockers and may progress to surgical myectomies and septal ablations to increase the LVOT lumenal space.[21] The goal of treatment is to relieve or improve patient symptoms, limit the progression of disease, and reduce the possibility of sudden cardiac death.[17,19]

Case Resolution

During the course of her hospital stay, the patient was admitted to the cardiac surgery intensive care unit and remained on SCI precautions in an Aspen cervical spine collar at all times, with a particular focus on avoiding hypotension to reduce the chance of secondary SCI. The patient was treated with dexamethasone 10 mg every 6 hours and gabapentin 300 mg every 8 hours and was fasted from midnight before an anterior cervical disc fusion with neurosurgery. Once stable from the trauma perspective, cardiothoracic surgeons took the patient to the operating room for a coronary artery bypass graft of the left internal mammary artery to the left anterior descending artery, septal myectomy, and mitral valve repair. Postoperatively, the patient was extubated and was found to have a stable valve repair and ejection fraction of 65% on echocardiogram. The patient remained under inpatient cardiac and neurologic care and residential rehabilitation for 5 months before being discharged home. Upon follow-up 1 month later, the patient was found to have incomplete motor tetraplegia with intermittent ability to ambulate with a walker, ongoing neurogenic bladder requiring straight urinary catheterization, and intermittent gastroparesis.

Conclusion

To the authors' knowledge, this case represents the first published report of combined incomplete SCI secondary to syncope from HOCM flown by HEMS. This case highlights the complexities of both pathologies and the challenges of prehospital recognition and management of these patients.

Supplementary materials

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1016/j.amj.2019.03.002.

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