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Ruptured spinal arteriovenous malformation: Presenting as stunned myocardium and neurogenic shock

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Abstract

Background: Neurogenic pulmonary edema (NPE) is a clinical syndrome usually defined as an acute pulmonary edema occurring shortly after a central neurologic insult. NPE was identified 100 years ago, but it is still underappreciated in the clinical setup. NPE usually appears within minutes to hours after the injury. It has a high mortality rate if not recognized early and treated appropriately. Similarly, neurogenic shock is a known complication of spinal cord injury reported incidence is more than 20% in isolated upper cervical spinal injury. But NPE is rare to occur, and stunned myocardium (SM) is not reported in spinal arteriovenous malformation (AVM) rupture. SM is a reversible cardiomyopathy resulting in transient left ventricular dysfunction which has been described to occur in the setting of catecholamine release during situations of physiologic stress. We report a case of high spinal AVM rupture presenting as SM, NPE, and neurogenic shock.

Case Description: A 32-year-old male who presented with sudden onset of pain and weakness in upper limbs. Imaging studies showed AVM rupture by imaging techniques. Initially, the patient had severe hypertension, respiratory distress requiring intubation and ventilation, then he developed hypotension, bradycardia, and asystole, which required immediate cardiopulmonary resuscitation and atropine. He remained with quadriplegia and suffered from frequent episodes of bradycardia and asystole.

Conclusions: Spinal AVM rupture can present as neurogenic shock, stunned myocardium, and pulmonary edema. Early recognition of AVM rupture and prompt surgical intervention, as well as aggressive treatment of shock, may enhance recovery and decrease the long-term morbidity.

Key Words: Neurogenic pulmonary edema, neurogenic shock, spinal arteriovenous malformation rupture

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INTRODUCTION

Hypotension with bradycardia caused by a spinal cord injury (SCI) is given the name of "neurogenic shock." Bradycardia and cardiac arrest is well-known complication of cervical spine injury due to reduced sympathetic activity, which usually settles down within 6 weeks of injury.[6] There are few case reports in the literature of

high cervical SCI requiring permanent cardiac pacemaker due to this complication.[5]

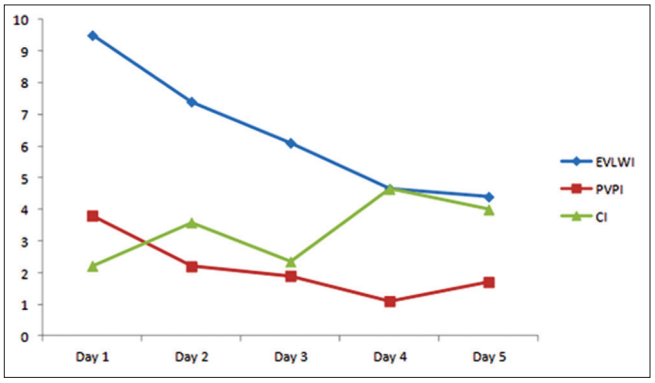
Immediately with the sudden rupture of an arteriovenous malformation (AVM) occurs release of huge amount of catecholamine which causes stunned myocardium (SM). Neurogenic SM (NSM) has most recently been explained by the "catecholamine hypothesis," which describes the underlying activation of sympathetic nervous system activity as the cause of reversible left ventricular (LV) dysfunction. Specifically, catecholamine release from sympathetic nerves innervating the myocardium leads to the development of reversible wall motion abnormalities, typically presenting as global hypokinesia and occurring with mild troponin elevations. The hypotension that results from neurogenic shock places patients at increased risk of secondary spinal cord ischemia due to impairment of autoregulation. Association of neurogenic pulmonary edema (NPE), SM, and neurogenic shock with spontaneous rupture of spinal AVM has never been reported in the literature. There are few reports of NPE and SM as an initial complication of central nervous system insult. Spontaneous spinal cord hemorrhage causing SM and pulmonary edema as a neurogenic shock requiring cardiac pacing is an unusual presentation of this case.

CASE REPORT

A 33-year-old Indian male developed sudden upper limbs weakness with neck pain while talking to his family on the phone. Emergency medical services (EMS) were called by one of his friends. On arrival of EMS, Glasgow coma score (GCS) dropped to 10/15 and oxygen saturation (SpO_2) 82%. On the way to hospital, GCS dropped to 3/15, blood pressure (BP) was 198/110, and he was intubated by EMS.

On arrival to emergency, BP dropped to 60/40 and heart rate (HR) 38 SpO_2 85%, he received atropine 0.5 mg and was started on dopamine 5 mcg/kg/min. His blood gases were pH 7.155, PO_2 61.2, PCO_2 53.2, HCO_3 18. Patient vitals stabilized BP 104/70 h 85 and SpO_2 98%. Computed tomography scan showed evidence of linear hypodense area noted in the visualized portion of the upper cervical spine. Chest X-ray showed bilateral diffuse infiltrates suggestive of early signs of pulmonary edema. The patient suddenly became bradycardic HR 36 followed by asystole which was revived by atropine and cardiopulmonary resuscitation (CPR) for 4 min according to ACLS guidelines. He was sent to the intensive care. Upon arrival at the Intensive Care Unit (ICU), he was resuscitated with fluids and central venous catheters line and pulse induced continuous cardiac output (PiCCO) line inserted. He had another episode of bradycardia HR 30 followed by asystole, being stabilized after atropine 3 mg and 4 min of CPR. Neurological assessment by GCS on admission was 3/15 (E1, M1, VT), pupils right 4 mm, left 5 mm, and reacting to light. The patient had no cardiac problems in the past and his electrocardiogram following both the episodes showed no abnormalities other than sinus bradycardia. Aminophylline, dopamine 5–10 mcg\kg\min, dobutamine 2.5 mcg\kg\min intravenous infusions were started. PiCCO® study results confirmed pulmonary edema and low cardiac index [Graph 1].

On the same day, echocardiogram was done which showed diffuse hypokinesia, mild impairment of left ventricle with apical wall motion abnormality ejection fraction (EF) of 40–45%, right ventricle systolic pressure (RVSP) of 41 mmHg. On the 2nd day, the patient was spontaneously opening eyes but complete quadriplegic though he was able to communicate with eyes and lips movements. It was documented as incomplete SCI as perianal sensation was intact. Magnetic resonance imaging study done showed sized area of hemorrhaging within the cervical cord extending from C1 to C4 secondary to rupture AVM [Figures 1 and 2]. On day 3, he developed severe ARDS requiring high ventilator support. Sputum Gram stains were negative. His electrolytes were within normal limits. On day 3, Holter monitoring showed the dominant rhythm of sinus bradycardia without any other critical arrhythmias. HR was varying from 35 to 89. On the 4th day, during routine ICU care, he again developed one episode of severe bradycardia responded to 3 mg of atropine. He continued to be on ventilator support at positive airway pressure. On the 5th day, he was weaned off from dopamine but continued on dobutamine and aminophylline infusion. Again on the days 6 and 7, he had oxygen desaturation (up to 77%) followed by bradycardia cardiac asystole resuscitated with atropine and CPR for 4 min. Neurologically, he did not improve, had repeated episodes of bradycardia. Surgical tracheostomy was done on the day 7. He developed high-grade fever, sputum Gram stains, and cultures showed Haemophilus influenza on the day 8, treated with meropenem. He had 12 episodes of severe bradycardia and asystole with desaturation



Graph 1: Advance hemodynamic parameters

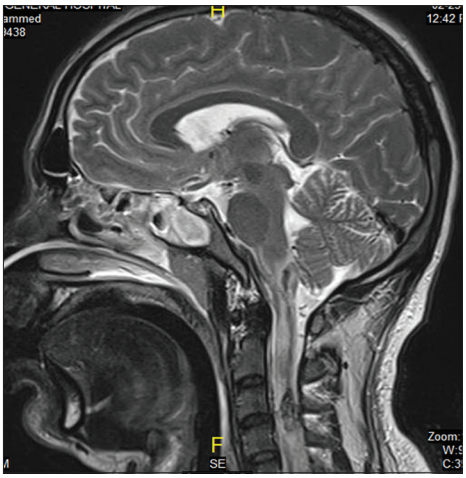


Figure 1: Magnetic resonance imaging showing abnormal signal intensity from C2 to C4

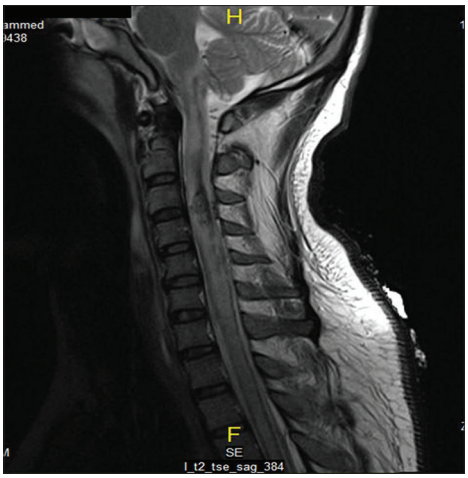


Figure 2: Magnetic resonance imaging showing hemorrhage from arteriovenous malformation

which required prompt treatment with atropine, CPR and high ventilator support during his stay of 21 days in ICU. This bradycardia was during routine ICU care, tracheal suctioning, and hypoxic induced. We decided to insert permanent cardiac pacemaker, but family wishes to transfer him to his home country for further management. He was escorted with a transcutaneous pacemaker. He remained quadriplegic and ventilator-dependent.

DISCUSSIONS

Our patient suffered insult in the upper cervical region of the spinal cord. We presumed that his hemodynamic instability is due to neurogenic shock, and further investigations showed the mild elevation of troponins with LV dysfunction which can be likely explained as NSM as the patient have no cardiac history before.

Neurogenic shock is almost always documented after an injury to the spinal cord.[4] The anatomic level of the injury to the spinal cord impacts the likelihood and severity of neurogenic shock. Injuries above the T1 level have the capacity of disrupting the spinal cord tracts that control the entire sympathetic system. Injuries occurring at the level from T1 to T3 may only partially interrupt the sympathetic outflow. The higher the level of insult, the more likely it is for the patient to exhibit severe symptoms.[6]

Hypotension with bradycardia caused by an SCI is named "neurogenic shock." Neurogenic shock is most common when the level of the injury is above T6. It can happen as an initial presentation or may be weeks after the initial insult. The incidence of neurogenic shock in children with SCI is unknown. However, reports indicate anywhere from 50% to 90% of adults with cervical SCI require fluid resuscitation and vasoactive infusions to achieve the adult parameters recommended (mean arterial pressure >85–90 mmHg for 7 days) by the Congress of Neurological Surgeons' guidelines for management of SCI. Adults with higher SCI (C1–C5) may be more likely to require cardiovascular interventions, such as vasoactive agents or cardiac pacing.[4]

Sympathetic innervation to heart arises from neurons in intermediolateral gray columns of cord segments T1–T4 which is under the control of higher centers via cervical spinal cord. Parasympathetic innervation comes via vagus nerve. Following any SCI above T1, the supraspinal control of sympathetic nervous system is lost, making the functioning spinal cord below the lesion independent of higher centers (known as "decentralization" of sympathetic nervous system), resulting in diminished sympathetic activity, while the parasympathetic control remains intact via vagus nerve, which results in relative parasympathetic dominance causing bradycardia and rarely cardiac arrest.[6]

This bradycardia will respond to atropine, glycopyrrolate, or vasoactive infusions with chronotropic, vasoconstrictor, and inotropic properties such as dopamine or noradrenaline.[6] In cases where patient demonstrates particular sensitivity to routine care cardiac pacing is strongly recommended.

In 1908, Shanahan reported cases of acute pulmonary edema as a complication of epileptic seizures. Localized ischemic insult in suspected brain trigger zones (vasomotor centers, pulmonary input and output locations: Medulla oblongata, area postrema, caudal medulla, solitarius tractus nuclei) has been explained as one of the pathophysiological causes. Several clinicopathologic paradigms have been proposed to explain the clinical syndrome of NPE: (1) Neuro-cardiac; (2) neuro-hemodynamic; (3) "blast theory;" and (4) pulmonary venule adrenergic hypersensitivity.[1]

The release of large amounts of catecholamine causes severe systemic vasoconstriction and concomitantly, reduced LV diastolic and systolic compliance leads to an increased ventricular volume and extremely increasing the LV workload leading to SM.[3] Care for SM involves

treatment of the underlying cause and supportive care.

NPE though uncommon can occur in patients with spinal cord injuries and an important co-factor of morbidity and mortality. There is evidence that the motor cortex is involved in cardiovascular adjustments associated with somatic motor activity, as it has functional connections with the caudal ventrolateral (CVL) medulla, a brainstem region critically involved in the control of BP and the regulation of plasma catecholamine levels. The CVL medulla CVL sends projections to the spinal intermediolateral nucleus, where preganglionic neurons take control of heart and blood vessels (T2 segment) and adrenal medulla (T8 segment). Injury to the middle thoracic spine cord or upper level can probably justify the occurrence of hemodynamic disturbances.[6]

Focusing on our case, as the patient had no previous cardiac abnormalities, sudden onset of neurological problems, typical initial severe hypertension, respiratory distress, and postarrest finding of diffuse hypokinesia with EF 40% and RVSP of 40 mmHg in combination PiCCO findings supports the diagnosis of NPE and SM. A reduced EF and SM have both been described in association with NPE.

CONCLUSION

Cervical AVM rupture despite being rare can present with neurogenic shock, NPE, and SM. Early recognition of AVM rupture and prompt surgical intervention, as well as aggressive treatment of shock, may enhance recovery and decrease the long-term morbidity. Cardiac pacing for long-term patient safety should be considered in such cases.

Ruptured cervical AVM reported to present as chest pain and intracranial subarachnoid hemorrhage.[2,7] To the best of our knowledge, it is the first case of such rare presentation, not reported in the literature.

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