High Cervical Spinal Cord Vascular Malformation masquerading as Guillian- Barre Syndrome: A Case Report

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Citation

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Abstract

Cervical spinal cord vascular malformations are very rare. They can mimic other neurological diseases involving the spinal cord. We present a case report of a 43-year-old male diagnosed and treated initially as Guillian Barre syndrome due to progressive ascending paralysis with intermittent exacerbations was referred to tertiary care facility for investigations. T2- weighted MRI revealed a vascular malformation in the upper cervical spinal cord. Postmortem examination revealed a vascular malformation with myelopathic changes and hemosiderin deposits in the upper cervical cord. A diagnosis of Intramedullary AVM was made. Spinal and MRI angiography could not be done to due the poor condition of the patient and hence precise delineation of angioarchitecture was not possible. In this report we emphasize the importance of accurate early diagnosis with utilization of appropriate imaging techniques and prompt management resulting in better treatment outcomes in spinal vascular malformations.

INTRODUCTION

Hebold in 1885 and Goup in 1888 described the vascular malformations of the spinal cord[1]. They are rare in occurrence and are inadequately studied. The disorder is quite complex and confusion remains about the appropriate classification Earlier the classification of spinal cord vascular malformations was based on autopsy findings.[1] In 1992, Anson and Spetzler classified spinal cord vascular malformations into 4 categories. Later, a modified system of classification of the vascular lesions of the spinal cord was proposed in 2002.This disorder could present as an acute, subacute and chronic spinal cord syndrome. They are not only difficult to diagnose but also mimic a wide variety of neurological disorders[3]. Vascular malformations are a treatable cause of spinal cord disease[5]. Recent advances in radio-imaging techniques delineating the angioarchitecture of spinal vascular anatomy has lead to better management of spinal vascular malformations[4].

CASE REPORT

A 43-year-old male was referred to our tertiary care hospital with features of high quadriparesis and a clinical diagnosis of Guillian- Barre Syndrome for MRI. He was also treated with Immunoglobulin. He was on and off ventilation for the past 3 months.

Patient suffered cardio-respiratory arrest while undergoing MRI scans. He was resuscitated and shifted to ICU. His blood pressure and pulse normal and his oxygenation was maintained by ventilatory support. However his GCS was 4/15. He had no spontaneous reflexes, no gag reflex. Pupils were non reactive. Lower limb had no reflexes and unresponsive to pain stimuli. Upper limbs showed only a flicker of movement to pain. He was diagnosed to have developed anoxic brain damage following cardio-respiratory arrest.

Patient had pallor, but no cyanosis, jaundice, clubbing or pedal edema. His hemoglobin was decreased (71 g/l). WBC count was 24.8x10e12/l. Platelet count was normal Serology for HIV and HTLV-1 were negative. While in intensive care unit he had episodes of bradycardia, fluctuating blood pressure, blood glucose levels, fluid and electrolyte balance. Later on his blood culture and urine were positive for Candida species and tracheal aspirates for Pseudomonas aeruginosa. He was transfused and treated with a combination of antibiotics.

Upon enquiry about his past history, his brother revealed that he had head injury following accidental fall 10 years ago and was suffering from recurrent right sided weakness and needed support on and off for walking. His condition progressed gradually with intermittent exacerbations.

Radiological studies: 1) MRI study of the cervical spine

done on the day of admission showed a heterogeneous area of approximately 5cms(LS) and 1.5cms (AP) having low signal on both T1W1 and T2W1 in the lower medullary and upper cervical region of the spinal cord. This region showed post IV contrast enhancement. These features are consistent with a vascular malformation with evidence of hemosiderin deposition due to chronic hemorrhage. The remainder of the cervical spine appears normal. (Fig1 &2)

Figure 1

Figure 1: pre (a) and post (b) contrast sagittal T1 weighted images of the cervical spine showing a 2 cm predominantly low signal area in the anterior cervical spinal cord with patchy tubular enhancement and focal dural thickening consistent with AVM

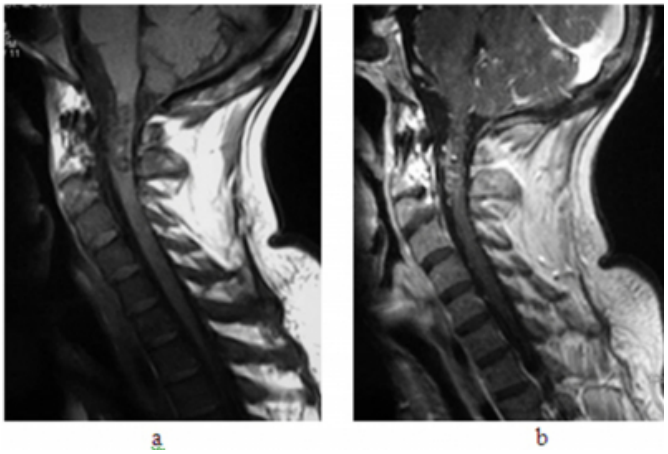
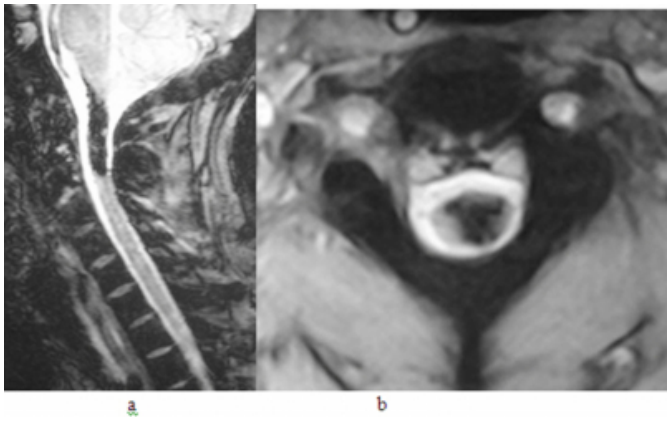


Figure 2

Figure 2: T2 sagittal (a) and axial (b) images showing hypointense areas within the anterior cervical spinal cord and dura consistent with haemosiderin of chronic haemorrhage



2) CT scan of the brain: Performed on the third day following the episode of cardiac arrest revealed cerebral edema. Hypodense areas in the basal ganglia, pons and occipital lobes were consistent with hypoxic/ischemic injury.

3) Chest x-ray showed right midzone consolidation.

Patient died after nine days of admission. A postmortem was requested to confirm the cause of death.

Postmortem examination revealed bilateral pneumonic consolidation with an area of infarct in the right mid zone. Cerebral edema was present and histology confirmed ischemic changes in the brain. There was no subarachnoid hemorrhage. Brain stem, cerebellum and cervical and upper thoracic spinal cord was taken out as a single unit through posterior approach. Upper cervical cord show clusters of vessels that were prominent on the dorsal surface. Thick walled blood vessels were seen within the cord substance with brownish areas of discoloration due to hemosiderin deposits (Fig3&4). Histology showed prominent and thickened blood vessels with hemosiderin deposits, ischemic changes and gliosis in the cord parenchyma (Fig5&6). These features indicate myelopathic changes with previous hemorrhages.

Figure 3

Figure 3: Specimen shows tortuous blood vessels on the dorsal and lateral surface of upper cervical cord and lower medullary region.

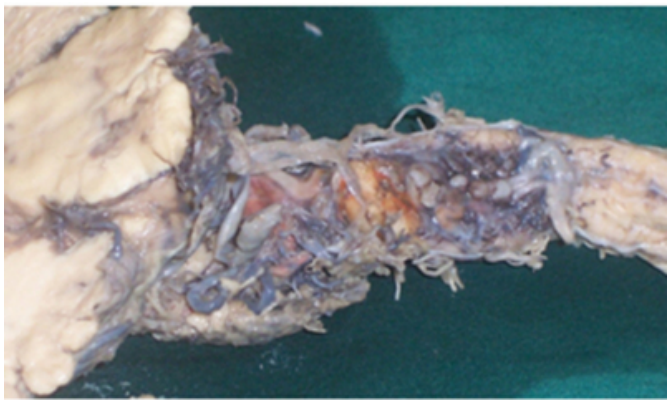


Figure 4

Figure 4: Cross section of the cervical cord showing prominent blood vessels in the parenchyma along with brownish discoloration

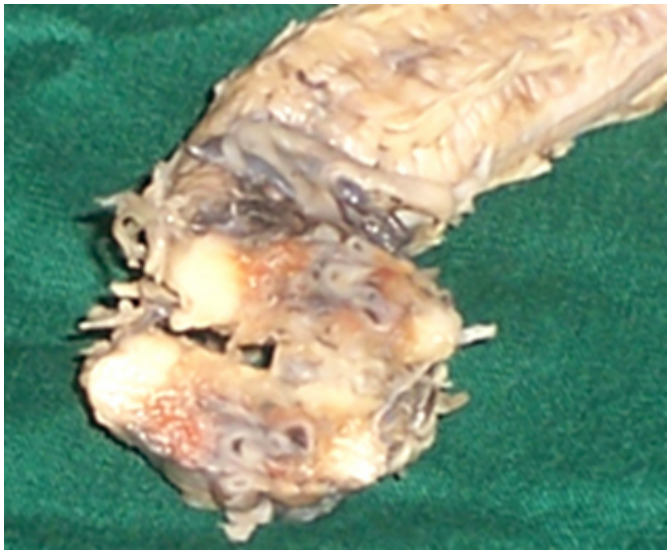


Figure 5

Figure 5: Section of the cervical cord showing large blood vessels, degenerating spinal cord parenchyma and wide spread hemosiderin deposits. H&E, 20x

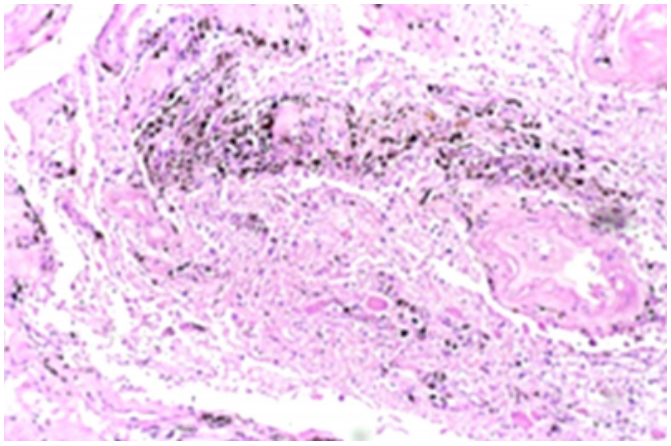
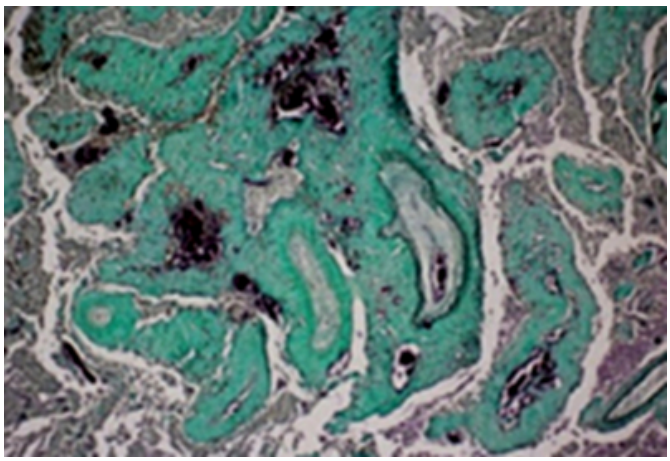


Figure 6

Figure 6: Section of the cervical spinal cord stained with Masson's Trichrome to highlight prominent thick walled blood vessels. Intervening degenerating parenchyma and hemosiderin deposits are also identified



A diagnosis of high cervical intradural cervical AVM (intramedullary) was considered based on clinical, radiological and pathology findings. However, Spinal and MR angiography were not done to precisely delineate feeder and draining vessels due to poor clinical condition of the patient.

DISCUSSION

Spinal vascular malformations consist of an abnormal connection between the normal arterial and venous channels[6]. A-V malformations (AVM) of the spinal cord are considered to be developmental abnormality composed of clusters of blood vessels[3]. The occurrence of vascular malformations of the spinal cord and dura are uncommon and compose 3-4% of spinal cord masses and are least common in the cervical region. They are more common in males than females [3,7]. For a long period of time there had been difficulties in proper classification and understanding of vascular malformations of the spinal cord. In 1992, Anson and Spetzler classified spinal cord vascular malformations into the 4 categories based on the angioarchitecture and flow patterns. AVM's are classified as Type 1 to Type 4. Type I, dural arteriovenous fistulas (AVFs); Type II, intramedullary glomus AVMs; Type III, juvenile or combined AVMs; and Type IV, intradural perimedullary AVFs [6]. Spetzler et al have updated and have proposed a modified system of classification, which divide the vascular lesions into three broad categories: Neoplasm's, Aneurysms and Arteriovenous lesions. Arteriovenous lesions are sub classified into AV fistula and AV malformations with further subdivisions [6,8,9].

The natural history of Spinal cord A-V malformations is important to make an early diagnosis and initiate prompt management of these cases. The characteristic clinical course reported by Wyburn –Mason states that the A-V Malformation of the spinal cord is associated with progression of symptoms by a series of apoplectiform stages or episodes with almost complete recovery between the attacks and eventual progression to having permanent symptoms. Aminoff and Logue in their study indicated that spinal AVM cases have progressive rather than relapsing deterioration.[2, 18]. Our case, the patient had relapsing deterioration with eventual permanent disability. This intermittent step like deterioration can be explained by episodes of hemorrhage, pressure effects of cluster of vessels and parenchymal changes of the cord[2]. The histological features in our case were characteristic and were comparable to those described in the literature [13]. A number of factors like Valsalva maneuver, trauma, pregnancy, postural changes, rise in body temperature, muscular effort have been associated with aggravations and precipitation of symptoms[3]. History of trauma in this case might have precipitated the condition or could be the cause of this lesion. The association of trauma and spinal AVM's is well documented[10]. It has been reported that clinical deterioration following intravenous contrast material could result from leakage due to altered cord-blood barrier in intramedullary AVMs [15,16]. Cardiac arrest during the imaging in our case could have resulted from a similar complication or due to his clinical condition.

There are number of reports stating that the spinal cord vascular malformations present as myelopathy[11]. Some neuropathic conditions may mimic myelopathy, most commonly GBS. Guillain-Barré syndrome (GBS) is one of the most common causes of weakness-inducing neuropathy[14]. The diagnosis of Guillian – Barre syndrome in this case was made due to the nature of presentation of the case as progressive quadriplegia. Past history was either not available or overlooked at the time of evaluation. The clinical presentations in this condition may mimic other conditions like multiple sclerosis, prolapsed intervertebral disc, demyelinating diseases neoplasia and myelitis.[2, 3,13]

CT scan, MRI and myelography are inadequate to delineate angioarchitecture and classification of the vascular malformations of the spinal cord. Advanced imaging techniques like MRI angiography and Digital Subtraction Angiography are essential to identify the lesion and also to precisely delineate the vascular anatomy of the lesion for appropriate treatment [3,17].Accurate and timely diagnosis is crucial for the treatment outcome. A number of reports have mentioned significant neurological deterioration in the course of the disease. Early detection and prompt surgical management is essential to preserve the neurological function and also to control the progression of the disease.

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