

Spinal intramedullary cavernous hemangioma

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ABSTRACT

The spinal cord can be involved in a variety of disease processes. These can be congenital or acquired. An acute onset of symptoms usually allows a defined set of causes to be considered including trauma, ruptured vascular anomalies, demyelination, and myelitis. Intramedullary cavernous hemangioma of the spinal cord is a congenital or acquired vascular malformation, and one of the rare causes of hematomyelia. We present such a case, and discuss the symptoms, diagnosis, and suggested best treatment options based on a review of present day literature.

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Spinal intramedullary cavernous hemangioma (SICH) is a congenital or acquired vascular malformation, which can bleed at any given point in time.¹ It accounts for approximately 5% of all intraspinal lesions.¹ The resultant hematomyelia causes para-or quadriplegia in the affected individual, and dictates surgical resection to ameliorate the permanence of the resultant neurological status.² To emphasize the need to offer patients the benefit of surgical intervention, we present our patient, who after presenting with paraplegia, declined the surgical treatment option, with dire consequences.

Case Report. A 30-year-old, single, Filipino, male mechanic, presented to the accident and emergency department of Riyadh Medical

Complex, Riyadh, Saudi Arabia complaining of lower limb weakness for one week. The illness started with a “funny” sensation in the abdomen and was followed by paresthesia and weakness in the right leg, followed 3 days later by weakness in the opposite leg as well as urinary retention. No history of upper respiratory tract infection, febrile illness, or spinal trauma could be elicited. He was a social drinker, and his last sexual contact was a year ago, heterosexual, and had been protected. No similar illness had occurred in his immediate family, and he had not come in contact with any ill person, particularly none with tuberculosis. He denied any exposure to animals or consumption of raw milk. On examination, the vital signs showed a pulse of 72/minute, blood pressure 130/80 mm Hg, respiratory rate 12/minute, and temperature of 37°C. No lymphadenopathy, raised jugular venous pressure, jaundice, or lower limb edema were noted. Examination of the chest, cardiovascular system, and abdomen were normal except for a distended urinary bladder. Central nervous system examination revealed a conscious alert and oriented patient with no cranial nerve abnormality and no neck stiffness. Neurological examination of the upper limbs was normal. The lower limbs showed an increased tone, with up going plantar response and sustained clonus bilaterally, hyperreflexia and decreased power of grade 3/5. Heel to shin test was impaired due to weakness. A sensory level could be elicited just above the umbilicus corresponding to a lesion around thoracic spine 7 or 8. Urinalysis was normal. Based on the above history and physical examination a tentative working diagnosis of transverse myelitis was made and work-up commenced. Complete blood count revealed white blood cell count of 16.6×10^3 with 83% neutrophils, hemoglobin of 14.4 g/dl, and platelet count of 258×10^3 . Urea and electrolytes showed a glucose of 5.5 mmol/l, urea 11.2 mmol/l, creatinine 110 μ mol/l, sodium 133 mmol/l, potassium 3.7, calcium 2.1 mmol/l, phosphate 1.18 mmol/l, magnesium 0.83 mmol/l. Amylase was 73 u/l, cardiac enzymes showed a lactate dehydrogenase (LDH) of 179 u/l, and a creatinine kinase of 95 u/l. Liver function test, and coagulation profiles were normal. Cerebrospinal fluid (CSF) analysis showed glucose 73.8 mg%, protein 48.8 mg%, and LDH of 33 u/l. The CSF cell count showed clear fluid with red blood cells $240/\text{mm}^3$ and white blood cells $2/\text{mm}^3$ and latex test negative. Blood, urine, sputum, and CSF cultures did not grow a pathogen. The CSF viral screen was negative for herpes simplex, Epstein-Barr and cytomegalovirus. The CSF and blood



Figure 1 - Magnetic resonance image of the spinal cord showing an increased intensity lesion on T2 images at T7-T8.

treponema pallidum hemagglutination test (TPHA) and venereal disease research laboratory test (VDRL) and Brucella titers were negative. Serology showed C-reactive protein of 3.19 mg/l, rheumatoid factor negative, anti-nuclear antibody and anti-double stranded antibody negative, anti-cardiolipin and anti-phospholipid antibody negative. Brucella abortus and melitensis titers were negative. Anti-human T cell lymphotropic virus (HTLV) and anti-human immunodeficiency virus (HIV) screens were negative. Electrocardiogram was normal. Chest radiograph revealed no abnormality. An MRI of the dorsal spine revealed an intra-spinal lesion of one cm at the level between thoracic spines 7 and 8 with a hypo-intense signal on T1 and T2 with increased intensity on the inferior aspect, indicating hemorrhage of most likely a small cavernous malformation. (Figure 1). On obtaining the result of the initial CSF investigations and MRI, the diagnosis was revised to bleeding of a cavernous hemangioma of the spinal cord, and he was offered neurosurgical intervention. However, he declined although it would have been performed at no cost since his case was an acute emergency. He opted to seek further treatment in his home country to the detriment of the medical team. There was no improvement in his condition in the following 2 weeks, while he awaited his departure, and eventually he was lost to follow-up.

Discussion. Except for trauma, a really acute onset of paraplegia is rare and suggests a vascular cause, which

when spontaneous is probably often due to rupture of a small congenital abnormality.³ These cavernous malformations also called cavernous hemangiomas are developmental vascular hamartomas.⁴ The resultant hematomyelia, meaning blood in the spinal cord, causes the immediate effects of a space occupying lesion with an acute onset of neurological compromise or a chronic progressive myelopathy due to micro hemorrhages and resulting gliotic reaction to blood products.^{1,3} Apart from rupturing, expansion of these lesions is well documented despite the absence of growth by mitosis.⁴ Congenital forms can be familial and acquired forms of SICH usually follow spinal irradiation for the treatment of malignancies.⁵ In a new classification proposed by Spetzler et al,⁶ where spinal cord vascular lesions are classified into 3 primary categories: neoplasms, aneurysms, and arteriovenous lesions, cavernous hemangiomas fall into the category of neoplasms.⁶ History of a ruptured or rapidly expanding SICH reveals an abrupt onset with additional deterioration in subsequent days as our case demonstrates.¹ Neurological examination, usually fairly accurately determines the level of the lesion due to the presence of a sensory level and urinary retention also points to cord compression as was the case in our patient.³ Diagnosis can nowadays be readily achieved with the use of MRI, which is thought to be diagnostic.¹ The lesions about a pial surface and have a characteristic imaging pattern.¹ Our patient's MRI revealed a hyperintense lesion intraspinally (Figure 1). Other diagnoses are to be excluded by appropriate investigations including serology, connective tissue screen, lumbar puncture, visual evoked potentials, and MRI brain. A MRI brain has to be performed in addition to MRI of the spine because of the known co-existence of both intracranial and spinal cavernous malformations.⁷ All modalities are effective, but must in every way be tailored to the specific needs and condition of the individual patient.⁴ They include observation, embolization, radiation, and surgical resection alone and in combination.⁴ Conservative treatment is instituted, when SICH are small, deep seated, and do not present with bleeding.⁸ Radiosurgery did not yield favorable results so far, and cannot be recommended.⁸ In the last 5 years, however, laminectomy and microsurgical resection have been advocated as the treatment of choice to avert or at least ameliorate the resultant neurological deficit, and thereby improving the quality of life for the patients.^{1,8-11}

The overall consensus is that surgery is better for patients with symptoms especially if paraparesis is not severe and of recent onset, particularly in view of the significant risk of re-hemorrhage with a prospective re-hemorrhage risk in one series of 66% per patient/year.⁹⁻¹¹ Unfortunately despite all efforts by the managing team, the patient declined surgery and most likely has suffered permanent neurological damage.

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REFERENCES

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