Case Reports

Management of Spinal Cord Cavernous Haemangioma

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Abstract

Cavernous haemangioma (CH) is a vascular malformation described as sinusoidal vascular channels located within the neural tissue but lacking intervening neural elements. Although spinal cord is not a frequent site for these lesions, the spinal cord cavernous haemangioma is currently encountered more frequently with magnetic resonance imaging. Management of both symptomatic and asymptomatic intramedullary cavernous haemangioma is a matter of debate as these lesions may cause a devastating spinal cord dysfunction. We present a young patient with thoracic intramedullary cavernous haemangioma who first presented with two episodes of transient paraparesis before he had a major intramedullary bleeding that rendered him in paraparesis and incontinence. We emphasize to consider early surgical resection as soon as possible to prevent potentially irreversible spinal cord damage secondary to a large or recurrent small bleedings.

Keywords: Cavernous haemangioma (CH), intramedullary, haemosiderin, surgical removal.

(J Med J 2009; Vol. 43 (2):129-133)

Received	***Accepted***
May 6, 2008	*August 28, 2008*

Introduction

Cavernous haemangiomas (or cavernoma) are congenital vascular hamartomas located within the neural tissue. It can occur throughout the central nervous system. Cerebral hemispheres and brainstem are usually the favorable locations. Whereas, spinal cord lesions are uncommon. Vertebral body cavernous haemangiomas is encountered frequently and usually asymptomatic in the vast majority of cases; occasionally these lesions may extend into the extradural or even into the intradural spaces, hence; compromising the neural tissues. [1-7] Spinal cord CH may be silent and may be discovered incidentally; but they usually have propensity for rupture and causing haematomyelia. Old bleedings result in haemosiderin deposit and gliotic reaction around the lesion. The treatment of choice for these lesions is total surgical resection but this may not be applicable in all cases. [8- 10] In this communication, we present a case of thoracic haematomyelia due to cavernous haemangioma, the patient underwent a total surgical resection of the lesion, and we then discussed the spinal cord cavernous haemangioma presentation and management options.

Case Report

A 19-year-old male was referred to our neurosurgical services with a two-day history of sudden onset of severe low back pain and rapid

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progressive paraparesis alongside with urine retention. Over the last nine months, the patient described two occasions of sudden and transient both leg weakness and frequent falls with six month interval.

The work up at that time included full blood tests, brain and lumber MRI and CSF study, which were normal. Although he had no clear diagnosis, he made a full recovery with expectant treatment that consisted of steroids and physical therapy each time. The rest of the systemic review was normal.

To examination of his cranial nerves and upper extremities were intact. He had his legs paralyzed with strength graded as 1/5 all over and hypotonic. He was areflexic. There was a decreased sensation on both legs compared with his upper limbs with a sensory level estimated at dermatome T8. The anal tone was also weak.

MRI scanning showed a hyper-intense intramedullary mass lesion surrounded by hypo intensity and occupying the anterior part of spinal cord at T6/7 level, expanding the spinal cord slightly in a fusiform manner (Figure 1a & 1b). Furthermore, the spinal angiogram was negative.

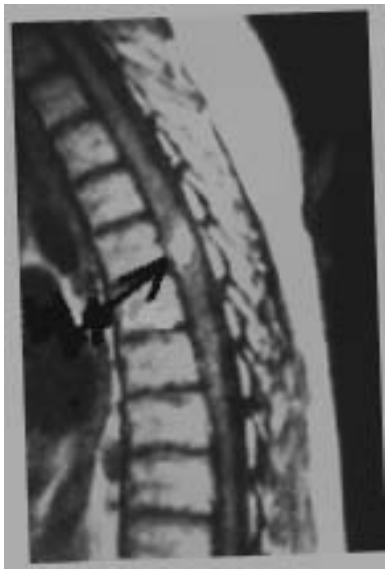

Figure 1(A): Sagittal T1- weighted.

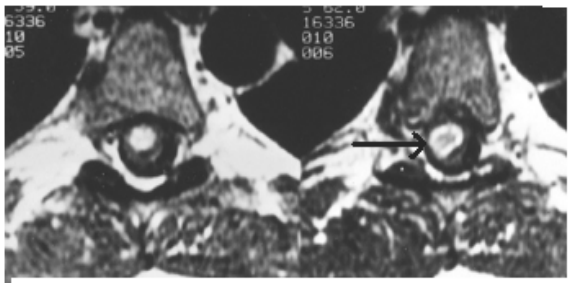

Figure 1(B): Axial T1- weighted MR scans of the Thoracic spine showing a hyper-intense lesion surrounded by hypo intense area involves the anterior 2/3 of the spinal cord at T6/7 with a fusiform spinal cord expansion. Represents a fresh clot surrounded by haemosiderin layer.

Surgical Procedure

The procedure was performed after placing the patient in prone position and receiving a high dose of steroids. A mid line thoraco-lumber skin incision was made. Laminotomy from T5-8 was then accomplished. The Thecal sac was full and bulging out. Under microscopic guidance, durotomy was affected. Tortuous small blood vessels were evident on the surface of the pale spinal cord. Marked expansion of the spinal cord was also noted. The myelotomy was then carried out. A well-demarcated lesion with a bluish – brown discoloration was identified with multiple small cysts and old clotted blood contents. Having dissection in a haemosiderin plane, the tumour was resected. Histopathological examination of the specimen revealed the presence of large sinusoidal blood spaces lined by endothelial cells, which are characteristics of cavernous haemangioma (Figure 2). Dura was then closed tightly. Lamina was replaced and fixed in situ, Closure was then affected in a watertight fashion.

Postoperatively, the patient made a relatively good recovery. Postoperative MR scanning revealed no residual mass left and the spinal cord looked normal (Figure 3). The patient was discharged ten days after surgery to rehabilitation. Neurologically, his strength now is almost 3/5 with improved sensation. His bowel and bladder function however had resumed some function.

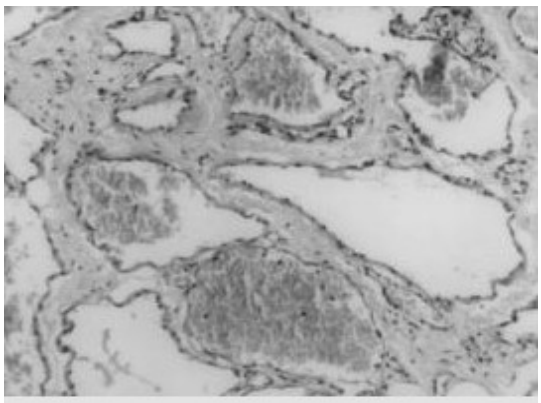

Figure (2): (H&E histopathology 400x). Typical histologic appearance of cavernous haemangioma. Note the irregular thick and thin sinusoidal vascular channels of variable sizes with some thrombosed blood vessels, it is lacking of intervening neural tissue.

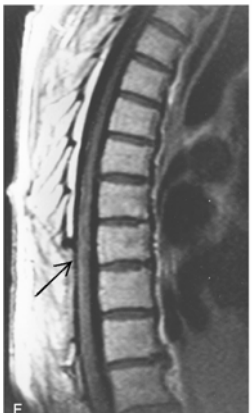

Figure (3): Post op sagittal MRI scan with GAD of thoracic spine shows adequate removal of intramedullary cavernous haemangioma with no residual lesion left.

Discussion

Intramedullary spinal cord cavernous haemangioma is uncommon and it occurs in females more than males with a ratio of 2: 1. Any age can be affected but it is usually presented in adults. Intramedullary cavernous haemangioma often presents with variable neurological conditions that depend on the location and size of the lesion which may vary from few millimeters to several centimeters and may be multiple in about half of cases. Symptomatic history may range from months to years. Most patients present with a long standing progressive myelopathic features, pain, motor deficit, gait disturbances, sphincter dysfunctions and sensory deficits.

These symptoms are usually attributed to the mechanical pressure effect on the spinal cord or to the repeated minor bleedings; these bleedings are usually venous and not arterial to cause a devastating spinal cord insult. Because of the insidious nature of these lesions, they are easily missed or may be picked up late after episodes of recurrent minor bleedings; besides bleeding some lesions may calcify or thrombose. In contrary, extremes may be encountered; acute symptoms caused by a new and significant hemorrhage within or around the lesions or silent lesions may be discovered incidentally on the MRI organized for other purposes. [1, 2, 7,10, 11]

The neurological deficits may have sometimes variable degrees of recovery after each ictus; others continue to display a gradual clinical decline. To clinical correlation; four patterns are recognized: (I) acute worsening with small but repeated haemorrhages or with thrombosis of the sinusoidal vessels, (II) slow progression due to progressive extension of the vascular lesion with thickening of the malformed vessels and thrombosis, (III) acute onset with rapid decline due to tissue haemorrhages, (IV) acute onset with gradual decline attributable to the changed microcirculation due to intraparenchymal haemorrhage. [12- 15]

The present case had spontaneous recovery on two events before experiencing a rapid deterioration of neurological status, which is attributed to a significant bleed and rapid enlargement of the cavernoma with spinal cord swelling.

Imaging appearance of spinal cord cavernous haemangioma is usually characteristic; it appears as mixed intensity on T1WI and T2WI, the core of the lesion appears as hyper intense and is surrounded by a low signal intensity; this ring of hypo intensity represents haemosiderin deposits secondary to old and may be recurrent bleedings; spinal cord enlargement may present and this is correlated with a new bleeding or edema, the enhancement is usually modest.

However, cavernous haemangioma is occult on the angiogram. [1, 16, 17] The MRI of this case revealed hyper-intense lesion within the parenchyma with a ring of decreased intensity and is associated with a spinal cord expansion. This hyper-intensity is correlated to a fresh bleeding.

Microscopical and early total surgical resection, particularly in those with acute symptoms, is considered to be the optimal treatment for all symptomatic intramedullary cavernous haemangioma with satisfactory results. However, to achieve this goal, meticulous dissection must be affected within the haemosiderin stained gliotic tissue commenced with by making myelotomy directly over the bulged bluish discoloration of lesion and keeping it limited to the abnormal looking tissue to stay away from normal surrounding neural tissues. As a result, when these lesions can be completely removed, the risk of regrowth or rebleeding is permanently eliminated. Incomplete removal may lead to recurrence of symptoms and may continue progressive deterioration as a result of bleeding from residual malformation. Radio surgery has no role in the treatment of intramedullary cavernous angioma. What is more, management of asymptomatic, incidentally discovered lesions should be dealt with on individual bases. [11, 13, 18,19]

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علاج الورم الوعائي الكهفي في النخاع الشوكي

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الملخص

الورم الوعائي الكهفي هو توسع خلقي في قنوات الأوعية الدموية ضمن مادة النسيج العصبي، غير أنه لا توجد عناصر عصبية ضمنه. وهو نادر الحدوث في النخاع الشوكي، ولكن مع وجود صورة الرنين المغناطيسي ازداد تشخيصه، وما زالت طرق علاجـه موضـع نقـاش. ونستعرض هنا حالة شاب عانى هذا الورم في النخاع الشوكي في منطقة الظهر، وأدى إلى حدوث شلل واحتباس بولي؛ مما تطلّب إجـراء مداخلة جراحية سريعة لمنع حدوث أذية شديدة في النخاع الشوكي بسبب تكرار الـ يف.

الكلمات الدالة: ورم وعائي كهفي، مادة النخاع الشوكي، ترسب هيموسيدرين، استئصال جراحي.