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CASE REPORT

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Multiple CNS cavernous haemangiomas presenting with spinal cord haematoma secondary to an intra-medullary cavernous haemangioma: A case report

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Summary

Background: Spinal vascular malformations are rare. Intra-medullary cavernous haemangiomas are very rare and only few cases have been described. Occasionally they may cause pain, myelopathy with sensory or motor deficit due to mass effect or hemorrhage.

Case Report: We report a patient who presented to the emergency department with acute retention of urine and lower limb weakness. He was diagnosed as a spinal intramedullary haematoma secondary to a cavernous haemangioma. In addition, multiple intracranial cavernous haemangiomas were found.

Conclusions: We are of the opinion that our case of spinal intramedullary cavernous haemangioma with multiple intracranial involvements deserves attention due to its rarity and it is also an important reminder to examine the whole neuroaxis in patients with spinal intramedullary cavernous haemangiomas, regardless they are symptomatic or asymptomatic.

Key words: cavernous haemangioma • intramedullary haematoma • MRI • spine • brain

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Background

Spinal vascular malformations are rare diseases that include true inborn cavernomas, arterio-venous malformations (AVMs) such as perimedullary fistulae, glomerular and juvenile AVMs, and presumably acquired dural arterio-venous fistulae [1]. Cavernous haemangiomas (CHs) are uncommon central nervous system (CNS) vascular disorders, and constitute 9% of CNS vascular malformations [2]. These angiographically occult lesions are well-circumscribed entities, consisting of thin-walled, lobulated vascular channels, without intervening neural tissue [3]. The spinal cord is an uncommon site for CHs. Intramedullary CHs are very rare and only few cases have been described [4].

We present a case of spinal intramedullary CH in a patient with multiple CNS CHs.

Case Report

A 49-year-old male presented to the emergency department with tingling sensation and bilateral lower limb weakness. He also suffered from acute retention of urine. This was the first occurrence of urine retention with relatively long history of back pain and lower limb weakness. He had no prior history of trauma. Physical and neurological examinations showed mild weakness of both lower limbs with no other abnormalities. There were no abnormal relevant laboratory findings. The previous patient's history and his family history were negative for any neurological disease.

Magnetic resonance imaging (MRI) of the thoraco-lumbar spine was requested on emergency basis to rule out spinal cord compression. The MRI study was performed on Siemens Avanto 1.5 Tesla (Siemens Medical System, Germany) in sagittal, axial and coronal planes. The examination demonstrated an intramedullary lesion at Th11 vertebral level (Figure 1A–D). The lesion appeared hyperintense on T1W images and also hyperintense on T2W images with associated low signal intensity peripheral rim, denoting haemosiderin deposit with an adjacent area of bright signal representing oedema pattern within the cord, just opposite Th9 and Th10 vertebrae levels. It showed no contrast enhancement.

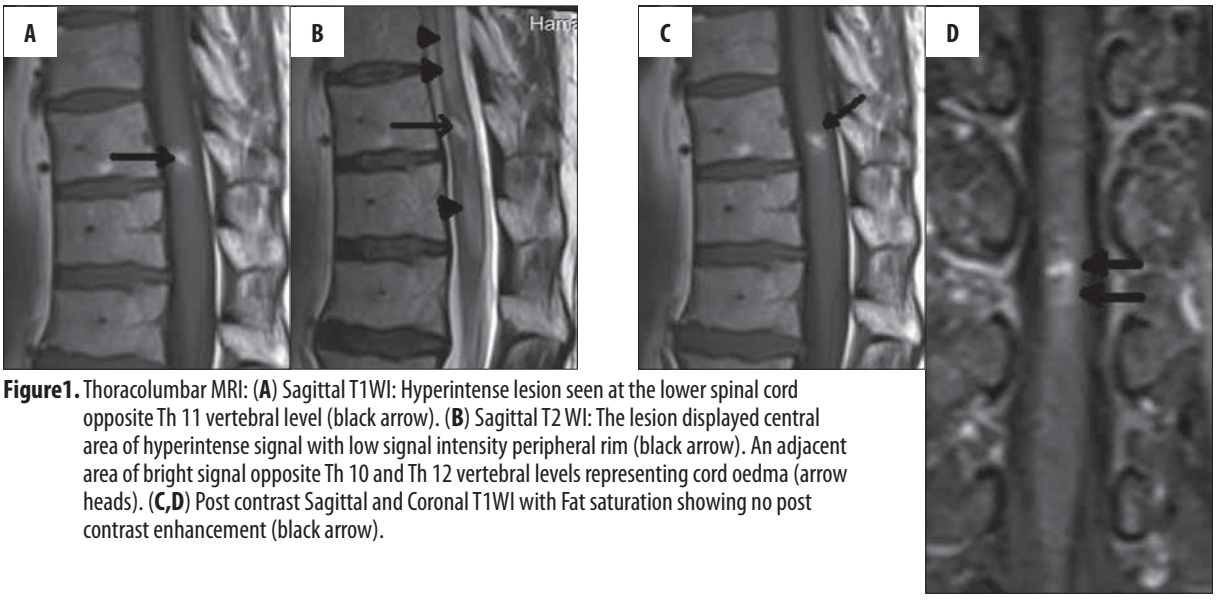

Figure1. Thoracolumbar MRI: (**A**) Sagittal T1WI: Hyperintense lesion seen at the lower spinal cord opposite Th 11 vertebral level (black arrow). (**B**) Sagittal T2 WI: The lesion displayed central area of hyperintense signal with low signal intensity peripheral rim (black arrow). An adjacent area of bright signal opposite Th 10 and Th 12 vertebral levels representing cord oedma (arrow heads). (**C,D**) Post contrast Sagittal and Coronal T1WI with Fat saturation showing no post contrast enhancement (black arrow).

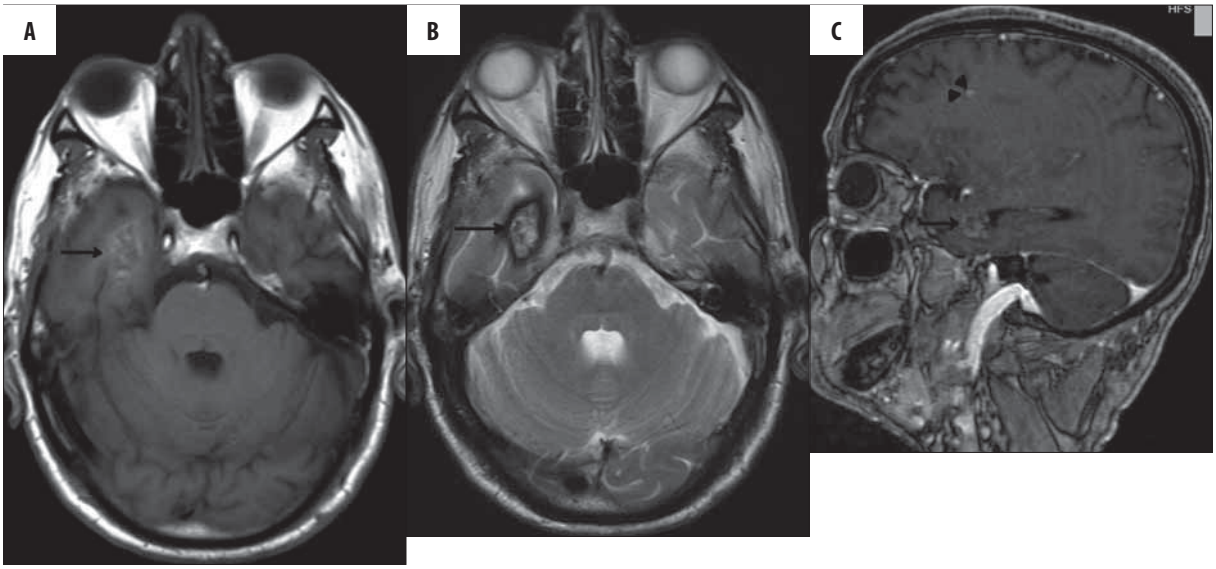

Figure 2. Brain MRI: (**A**) Axial T1WI: Hyperintense lesion seen at medial aspect of the right temporal lobe (black arrow). (**B**) Axial T2 WI: The lesion displayed central area of hyperintense signal with low signal intensity peripheral rim (black arrow). (**C**) Post contrast Sagittal T1WI showing two lesions with no post contrast enhancement (black arrows).

Then MRI of the head was requested as a surveillance study for the neuroaxis to look for possible similar lesions. The MRI was performed in sagittal, axial and coronal planes using T1, T2, Flair and Gradient Echo (GE) pulse sequences. The study showed innumerable lesions, various in size, with typical presentation of CHs. The lesions were found in the cerebral hemispheres, cerebellum, and brain stem (Figure 2). The largest of those lesions was in the right temporal lobe and it measured 32 mm in the biggest diameter. All the lesions displayed low signal intensity on GE images and showed no contrast enhancement (Figure 3).

The patient refused surgery and was discharged following symptomatic improvement on conservative management. Unfortunately, the patient left the country and died soon later. The immediate cause of death is not known and we were not able for the follow up study or genetic screening.

Discussion

Spinal cord CHs can be extradural, intradural extramedullary, or intramedullary [5]. The lesions may be seen at any level from the upper cervical cord to cauda equina, but are most frequently localized at the cervical and thoracic spinal cord [6]. They may be asymptomatic, or they may cause pain, myelopathy with sensory or motor deficit due to hemorrhage and mass effect [7]. Widespread use of MRI has resulted in enhanced sensitivity and specificity of the diagnosis of CH and thus led to an increase in the number of reported cases of spinal cavernous haemangiomas [8].

Intra-spinal CHs account for 5–16% of all spinal vascular abnormalities. Multiple spinal cord CHs are very rare, and only a few cases have been described [9].

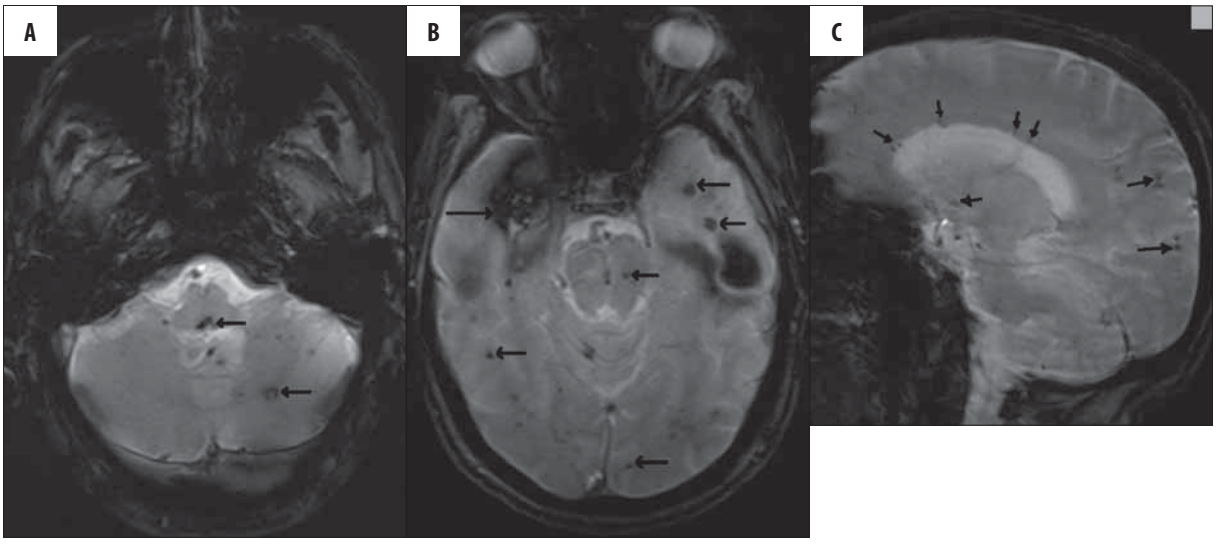

Figure 3. Brain MRI: (**A,B**) Axial and (**C**) Sagittal T2* WI (Gradient Echo images) showing multiple innumerable low signal lesions, variable in size, distributed allover the cerebral hemispheres, cerebellum and brain stem representing the multiple intracranial cavernous haemangiomas (black arrows).

Zevgaridis et al. reviewed 116 patients with intramedullary spinal CHs which had been published between 1903 and 1996. They found only one patient who had two spinal intramedullary CHs [10]. Vishteh et al. reported 17 patients with intramedullary spinal cord CH. Only one patient had a multiple CHs in the spinal cord [11].

Intra-spinal CHs commonly accompany intracranial CHs [11]. Cohen-Gadol et al. reported that as many as 40% of patients with a spinal CM may harbor a similar intracranial lesion [4].

As much as 40% of coexisting spinal and intracranial CHs may have the non-familial (sporadic) form; however coexistence of CHs in the brain and spinal cord typically occurs in patients with the familial form [12]. This condition has an autosomal dominant pattern of inheritance with incomplete penetrance [13]. Genetic analysis has identified foci on chromosomes 7q11-21, 7p13-15, and 3q25, 2-27 [14].

Our patient was considered to have the non-familial, sporadic form of spinal intramedullary CH, located in the thoracic spinal cord, with multiple intracranial involvements, yet we consider that it is necessary to study the family's genetic analysis.

We are of the opinion that our case of spinal intramedullary cavernous haemangioma with multiple intracranial involvements deserves an attention not only due to its rarity but also because it is an important reminder to search the neuroaxis (brain and whole spinal cord) in patients with spinal intramedullary CHs, regardless they are symptomatic or asymptomatic.

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