

Case Report

A case of hematomyelia caused by intramedullary cavernous haemangioma

E. AMOROSO, A. GUARRACINO

Neurosurgical Unit, "Umberto I Hospital", Salerno, Italy

SUMMARY: Here we report the case of a 35-year-old man with a history of cutaneous cavernomas presenting with sudden onset low back pain and right leg numbness followed by rapidly increasing weakness in both legs. One week after onset, the patient showed paraplegia (Frankel-scale B), bilateral impairment of all sensory perception below the L2 level, and no sphincter control. Spinal magnetic resonance imaging revealed hematomyelia with perifocal oedema at the D11 vertebra. The patient initially refused surgical treatment, and was therefore given steroids and physiotherapy, but clinical improvement was very modest. After two months the patient therefore agreed to surgery, namely laminectomy, posterior midline myelotomy, and total surgical excision of the hematomyelia, its capsule and the accompanying cryptic vascular malformation. Microscopic examination of the excised tissue revealed cavernous haemangioma. Following the operation, the patient gradually recovered mobility (Frankel-scale D), sensory perception and sphincter control. Total surgical resection can be considered a procedure of choice for the management of symptomatic hematomyelia caused by intramedullary cavernous haemangioma, and delaying the operation does not seem to affect its success.

KEY WORDS: Haemangioma, Hematomyelia, Intramedullary spinal cord lesion.

INTRODUCTION

Spontaneous hematomyelia caused by sudden haemorrhage of a cryptic intramedullary cavernous haemangioma is a rare event^(21,24), and best-practice for management of this lesion is not clear. Although surgical removal of both the clots and the haemangioma is recommended, the roles of both surgical timing and preoperative neurological impairment on the prognosis require further definition^(2,14-18).

A case of hematomyelia with spinal cord cavernoma, treated via complete surgical excision 2 months after its presentation is reported.

CASE REPORT

Two months before admission to hospital, a 35-year-old male experienced sudden-onset low back pain with numbness of the right leg, and subsequent rapidly increasing weakness in both legs. One week after the onset of pain, the patient developed paraplegia (Frankel Scale grade B), bilateral impairment of all sensory perception below the L2 level, and loss of sphincter control. Thirteen days after this event, spinal MRI showed a longitudinal area of T1 and T2 hyperintensity at the D11 level, consistent with hematomyelia at the stage of deoxyhaemoglobin conversion to methaemoglobin. Initially the patient refused the proposed surgical treatment, and was therefore given a

Correspondence: Dr. Ettore Amoroso, Neurosurgical Unit, "Umberto I" Hospital, via Iodice, 84014 Nocera Inferiore (Salerno), Italy, ph. +39-(0)81-9213307, fax ++39-(0)81-9213474, e-mail: ettoreamoroso@alice.it
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LIST OF ACRONYMS AND ABBREVIATIONS: ImCH = intramedullary Cavernous haemangioma; MRC = Medical Research Council; MRI = Magnetic Resonance Imaging; T1WI = T1 weighted images; T2WI = T2 weighted images.

course of steroids and physiotherapy, but with very modest clinical benefit. After 2 months of this treatment, therefore, the patient consented to the operation. Pre-operative neurological examination showed muscle strength grade 2/5 in the right leg and 1/5 in the left leg on the MRC scale. Deep tendon reflexes were brisk. Left plantar response was extensor, while the right was equivocal. Sensory impairment to light touch, pin prick and vibration was present in the right and, to a lesser extent, left leg. Neither previous treatment nor systemic factors were contributory, but the patient had a history of multiple cutaneous capillary cavernous haemangiomas. Routine haemogram and biochemical parameters were normal. X-rays of both chest and spine were normal.

Subsequent neuraxis MRI confirmed a spindle-shaped area of T1 and T2 hyperintensity, indicating the formation of hematomyelia (Figure 1).

The patient underwent a T10-T11 laminectomy and complete excision of the malformation. The margins were insolated by ultrasonography, and the dura mater was opened. A myelotomy was centred over the hematomyelia. After incision of the hematoma capsule and drainage of its haematic contents, total excision of the mass (capsule and dark-blue mulberry-shaped lesion) was performed, dissecting it from the spinal cord in the gliotic plane.

Histopathological examination of the mass revealed the presence of large blood spaces lined with endothelial cells, characteristic of cavernous haemangioma. There was no interspersed neural tissue.

After the operation, muscle strength in both legs, sensory perception and urological function progressively improved. After 15 months of follow-up, the patient was able to walk with assistance (Frankel Scale grade D), and only a small residual defect was visible under MRI (Figure 2).

□ DISCUSSION AND CONCLUSION

Cavernous haemangiomas, or cavernomas, are vascular hamartomas that can occur throughout the central nervous system, but are most commonly located in the supratentorial compartment⁽²³⁾. Although intramedullary cavernous haemangiomas are relatively rare, they have been diagnosed more frequently since magnetic reso-

nance imaging has become widely available. Most recent estimates indicate that spinal cord haemangiomas constitute 5% of all intramedullary lesions⁽⁹⁾. Most clinical series demonstrate a higher bleeding risk for ImCHs with respect to their cerebral counterparts; the annual bleeding risk is believed to be roughly 0.7-1.3% in cerebral cavernomas^(4,19,22), 2.7% in brainstem cavernomas⁽²⁰⁾, and 1.6-4.5% in ImCHs⁽²⁴⁾. ImCHs are most commonly found in the thoracic spinal cord, followed by the cervical cord. More rarely - only around 3% of all intramedullary cavernomas - they are found in the conus medullaris^(7,13).

Patients with symptomatic ImCH usually present a wide range of neurological symptoms, from acute onset to gradual neurological decline. Four major clinical patterns have been identified by Ogilvy et al., namely:

- *type 1*: acute episodes of stepwise deterioration with small but repeated haemorrhages or with thrombosis of the malformed vessels;
- *type 2*: slow progression due to progressive enlargement of the cavernoma with subsequent/with or without thickening of the sinusoid vessels and gradual thrombosis;
- *type 3*: acute onset with rapid deterioration due to intraparenchymal haemorrhages, and
- *type 4*: acute onset with gradual decline attributable to altered microcirculation due to intraparenchymal haemorrhage⁽²¹⁾.

In the Ogilvy series, type 3 accounted for only 22% of patients with spinal cord ImCHs, thereby explaining the relatively poor knowledge of the diagnostic and therapeutic aspects of hematomyelia complicating an intramedullary haemangioma.

The typical MR characteristics of cavernomas are reticulated mixed signal areas on both T1WI and T2WI, surrounded by an area of low signal intensity, which is predominant in T2WI. The enhancement is modest, and cord swelling is occasional^(3,10). However, these findings are not so common in the presence of hematomyelia, and an ImCH may only be suspected, particularly in the presence of one or more of the following factors:

- (I) a personal and/or family history of cavernous haemangiomas;
- (II) typical MRI appearances of mixed acute, subacute and chronic haemorrhage;
- (III) a tendency for signal intensity to decrease on follow-up;

(IV) normal spinal angiography; and
(V) associated brain lesions⁽²⁸⁾.

As patients with spinal cavernous malformations may harbour an intracranial lesion⁽⁶⁾, complete neuraxis imaging is therefore recommended⁽¹⁵⁾.

Embryological findings support the role of a personal history of cavernous haemangiomas in the diagnosis of cryptic ImCH. As the primordial vascular plexus, with its solitary layer of endothelium, is indistinguishable from that of the walls of cavernous malformations, it can be postulated that the former may lose the capacity to differentiate, resulting in the latter. Cavernous malformations of the dermis and muscle in the same dermatome and myotome, occurring as vertebral body and spinal cord lesions, are explained by segmentation of a blood vessel with each somite. Cavernous malformations can also be produced in many unrelated organ systems and dermatomes if the loss of the ability of blood vessels to differentiate is more diffuse⁽¹²⁾.

Total surgical resection under high magnification using microsurgical techniques is a procedure of choice for the management of symptomatic ImCHs, with or without hematomyelia^(1,2,5,9,17,18,27). A study involving 117 patients reported that surgical outcomes are encouraging, leaving 66% improved, 28% unchanged, and only 6% worsened⁽²⁹⁾.

A general improvement in neurological function is reported after surgery^(2,11,17,18,24,26,27), with myelopathy secondary to acute presentation appearing to be more reversible than in both stepwise deterioration and slow progression patterns of presentation⁽¹⁵⁾. As surgical morbidity and functional outcomes appear to be closely related to the preoperative functional status of all patients with ImCH, surgery should be performed early, before the patients have developed substantial deficits^(8, 21, 25). Nonetheless, early surgery is not mandatory in patients with acute haemorrhage, and a waiting period of several weeks (3-4 weeks) may facilitate resection, as a gliotic plane develops between the lesion and the spinal cord, enabling relatively safe removal^(15,18). The liquefaction of blood clots, which facilitate the identification of the different tissues (white matter of medulla, capsule of the hematoma, cavernoma), is also thought to be a benefit of delaying surgery⁽⁸⁾.

To our knowledge, this is the first report of a significant neurological improvement owing to delayed surgery on hematomyelia due to ImCH.

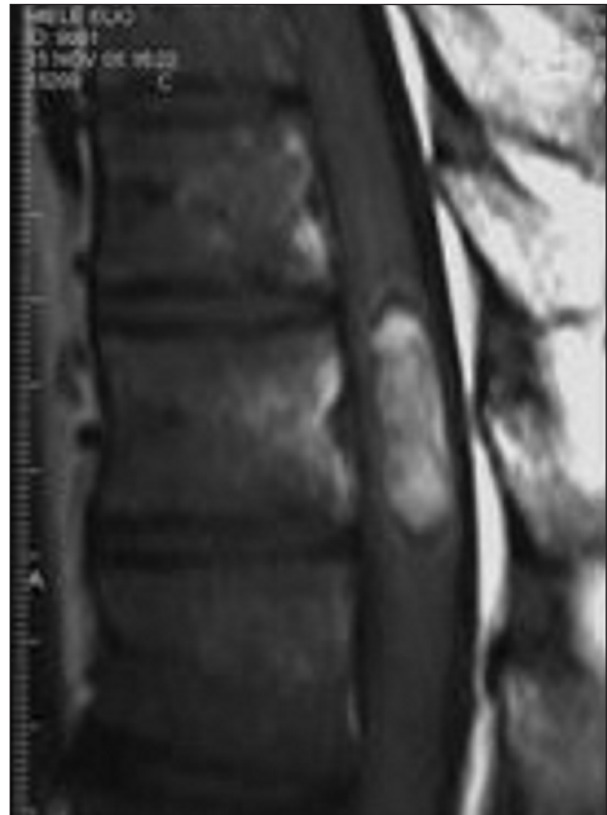


Figure 1. Pre-operative MR images of hematomyelia at the D11 level. Sagittal T1-weighted image.



Figure 2. Post-operative MRI.

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