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Intramedullary cavernoma with extralesional haemorrhage

Naureen Farhan Ahmed, Fatima Mubarak, Nida Sajjad

Abstract

Intramedullary cavernomas are rare lesions constituting 5 to 12% of all intramedullary tumours. They are more recognized after introduction of magnetic resonance imaging and detection rates have improved by advance techniques. They may be solitary or multiple or may have associated cranial lesions. They may present with gradual neurological decline or with acute loss of spinal function. In addition neurological deficit depends on the location of the lesion within the spinal cord. We are reporting the case of a 45 year old male who presented with neck pain and progressive right arm weakness with numbness. MRI cervical spine with contrast showed intramedullary lesion with peripheral T2 hypointense rim and extralesional haemorrhage consistent with cavernoma. Patient underwent successful surgery and his symptoms markedly improved with mild residual grip weakness in right hand.

Keywords: Intramedullary cavernoma, Magnetic resonance imaging, T2 hypointense rim, Extralesional hemorrhage.

Introduction

Intramedullary cavernomas represent approximately 5% to 12 % of intramedullary lesions in adults and 1% of intramedullary lesions in children.¹ It is a lesion composed of tiny vessels without intervening neural tissue.² They usually present with neurological deficit that could be mild to para or quadriplegia depending upon the location. As these are surgically curable cause of myelopathy, early imaging is important in cases presenting with the symptoms to allow appropriate diagnoses, treatment and better outcome.¹

Case Report

A 45 year old male presented with neck pain for six months and progressive right arm weakness and numbness for one week.

MRI cervical spine with contrast showed a small eccentric intramedullary lesion at C3-C4 level having typical reticular or pop corn appearance with peripheral rim of T2

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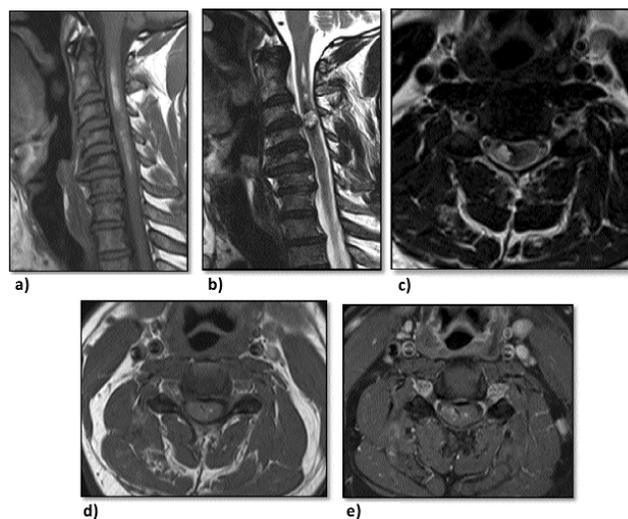


Figure: a) SagT2WI, b) SagT1WI, c) axial T2WI show intramedullary eccentric lesion with typical reticular pattern and peripheral rim of siderosis. Note significant extralesional haemorrhage. d) axial T1 non contrast, e) axial T1 post contrast image show patchy enhancement.

hypointensity representing siderosis. Significant perilesional haemorrhage was noted appearing hyperintense on T1 and T2WI representing sub acute stage. There is mild cord expansion and minimal extramedullary extension towards right lateral aspect. The lesion showed patchy post contrast enhancement.

Discussion

Cavernous angiomas or haemangiomas account for 5 to 12% of spinal vascular malformations.³ These are present at three sites, intraosseous, intraspinal extramedullary and intramedullary. Vertebral location is most common involving bodies and less likely posterior elements, and may have extradural extension. Intramedullary cavernoma represent 3% of cases and extramedullary cavernomas are extremely rare.⁴

There is a wide spectrum of clinical manifestations determined by the location of the lesion. Progressive focal sensorimotor deficits are most frequent presentations. There may be associated intense radicular or central pain. Patients may present with progressive myelopathy secondary to micro-haemorrhages and surrounding gliosis. Patients with extralesional haemorrhage show

significant neurological decline.^{5,6}

The mean age of presentation of intramedullary cavernoma is 37 years (range 12-88 years.) with a peak occurrence between the third and fourth decades.⁷ It has been reported that intramedullary cavernomas are two times more common in women than in men⁸ but other studies do not support this.

Histologically intramedullary cavernoma are same as that of cranial lesions. These are most frequently found at the thoracic level in 77% of the cases.⁹ Cervical location is less frequent, occurring in less than 23% of patients. Intramedullary cavernoma in the lumbar region is rare, (4%) as quoted in the literature.

Most common location of intramedullary cavernomas on axial axis is central, occurring in 34% of cases. Then posterior and lateral locations are common occurring in 23% and 17% of cases, respectively. Anterior location is rare occurring in 9% of cases.

Advanced neuroimaging techniques have increased the detection of intramedullary cavernomas. MRI is the most reliable diagnostic tool. Typical feature of intramedullary cavernoma on MRI is "popcorn appearance" which is a webbed core, composed of blood and blood products in various stages of evolution. It has variable signal intensity and enhancement related to evolution of haemorrhage. It shows typical hypointense rim on both T1 and T2 weighted images representing siderosis. Small cavernous malformations may represent as focal area of decreased signal density, "black dots". Susceptibility weighted imaging is valuable in diagnosis and should be performed for evaluation in suspected cases. It has been reported that they may have an associated large draining vein however this is not a characteristic feature of intramedullary cavernoma. If perilesional oedema is present other pathologies like cryptic AVMs and haemorrhagic neoplasm should be excluded.⁸

There is good association of intramedullary cavernomas and concomitant cranial cavernomas. In a series of 14 intramedullary cavernomas, three patients (21%) were reported with multiple intracranial lesions.¹ Another series reported that 47% of 17 patients with intramedullary cavernoma had multiple intracranial cavernomas.¹⁰ Four of these patients had a family history of cavernomas. Therefore, it is recommended to do MRI of the brain in young patients having intramedullary

cavernoma, as they may have cranial lesions too.⁵

Intramedullary cavernomas are easily resectable tumours from surrounding haemosidrin bed and have a very good surgical outcome. Due to the aggressive clinical course, surgical resection is valuable in the management.¹ Patients with longer duration of preoperative symptoms are associated with less successful outcome after surgery secondary to chronic myelopathy.⁶ For this reason early detection is important. Surgery of deep and anteriorly locating lesions carries greater risks because of excessive manipulation and dissection of the spinal cord.

Conclusion

Intramedullary cavernomas are rare lesions that may be found as an incidental finding in asymptomatic patients but can present with mild to severe gradual or acute neurological deficit secondary to intra and extra lesional haemorrhage and resulting gliosis. They are easily resectable tumours and immediate diagnosis and surgical intervention would result in better outcome. Although rare, it has typical imaging features on MRI that is imaging modality of choice favouring diagnosis.

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