CASE OF INTRAMEDULLARY CERVICAL SPINAL CORD CAVERNOUS ANGIOMA

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ABSTRACT

A 23 years old male patient presented with complain of pain in neck and both upper limbs more so on right side for 3 months. Patient came for MRI cervical spine with and without contrast which revealed an intramedullary abnormal signal intensity spindle shaped mass at C5-C6 level.

Introduction

Cavernous malformations/cavernous angiomas or cavernomas are uncommon vascular malformations of central nervous system. They consist of abnormally dilated endothelial lined blood vessels without intervening normal neural tissue.

The incidence of cavernoma in central nervous system including both brain and spinal cord is around 1.9 cases / 100,000 person per year, with 3-5% of lesions located in the spinal cord.[1,2] Spinal cavernomas account for 5-12% of spinal cord vascular lesions. Most of the spinal cavernomas originate in the vertebrae and occasionally extend into the extradural space. Intramedullary cavernomas are very rare.[3] Females are more commonly affected than males and usually manifest clinically during third and fourth decades of life.[3] Patients usually present with a wide variety of symptoms. The most common presentation being slowly progressive myelopathy, but sub arachnoid haemorrhage[4] and haematomyelia[5] can also be present.

Since the advent of MRI, cases of spinal cord cavernoma are diagnosed more frequently. They are usually found incidentally, however may present with symptoms.

We describe here a case of intramedullary cervical spinal cord cavernomain a 23 year old male patient emphasizing its neuroradiological features.

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Case Report

A 23 years old male patient presented with complain of pain in neck and both upper limbs more so on right side for 3 months. Patient also complained of weakness in right upper limb. Clinical and neurological examination showed no significant abnormality.

Patient came for MRI cervical spine with and without contrast which revealed an intramedullary abnormal signal intensity spindle shaped mass at C5-C6 level. It was seen in the right half of spinal cord resulting in mild cord expansion. It showed heterogenous signals on T1W, T2W and fatsat sequences (Fig. 1, 2 & 3). It also showed hyperintense signals on all pulse sequences, these representing haemorrhage of varying duration. It measured 0.8 x 0.7 cm. There was alow signal intensity peripheral hemosiderin rim also noted on all sequences more obvious on T2W and fat-sat images. (Fig. 2 & 3) No significant post contrast enhancement noted (Fig. 4), this excludes the possibility of intramedullary tumor. The lesion demonstrated hypointense blooming on gradient echo sequence represnting depostion of degraded blood products (Fig. 5). These are classical imaging features of intramedullary cavernoma.

Patient is managed conservatively with analgesics. No surgery has been performed yet.

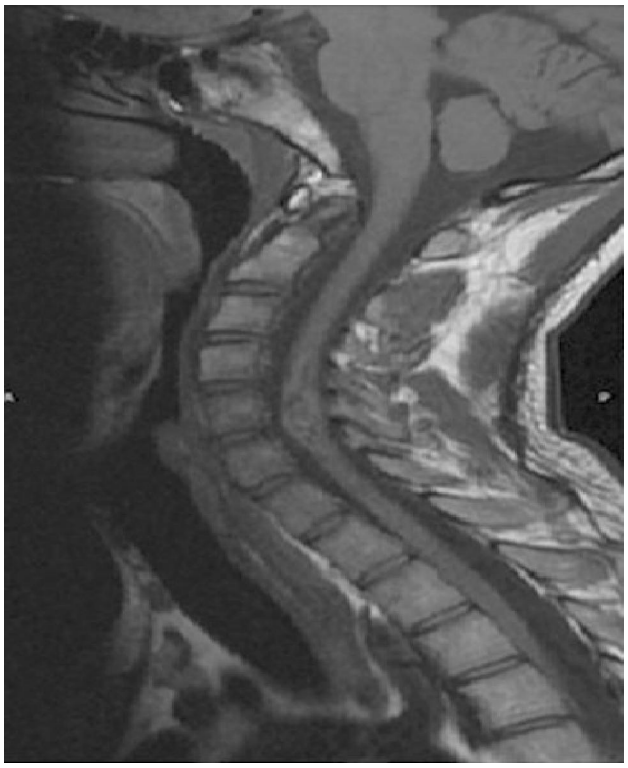

Figure 1: T1-weighted sagital MR image of cervical spine showing intramedullary hetergenous signal intensity mass with internal hyperintense signals representing hemorrhage at C5-C6 level.

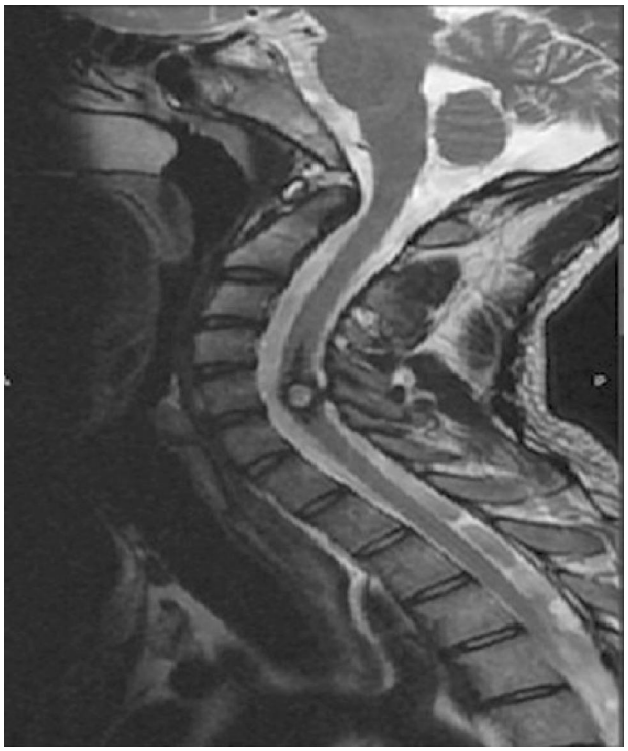

Figure 2: T2-weighted sagital MR image shows heterogenous signals of mass with peripheral hypointense rim of hemosiderin

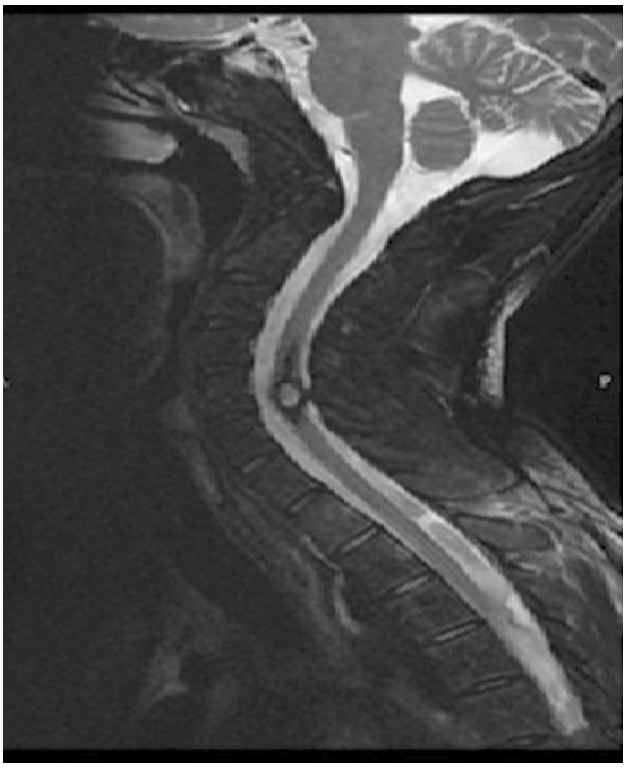

Figure 3: T2-weighted fatsat sagital image: The mass shows peripheral low signal intensity rim of hemosiderin

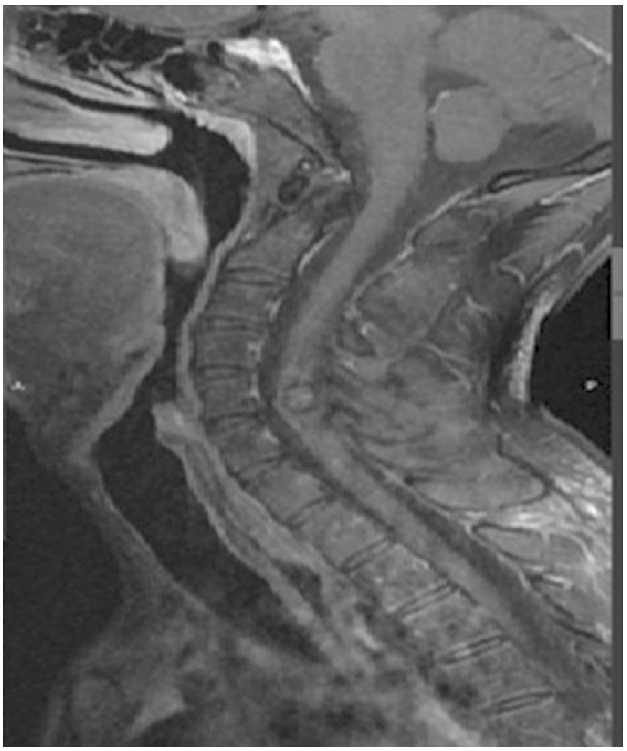

Figure 4: Post contrast T1-weighted image of cervical spine shows no significant enhancement of intramedullary mass

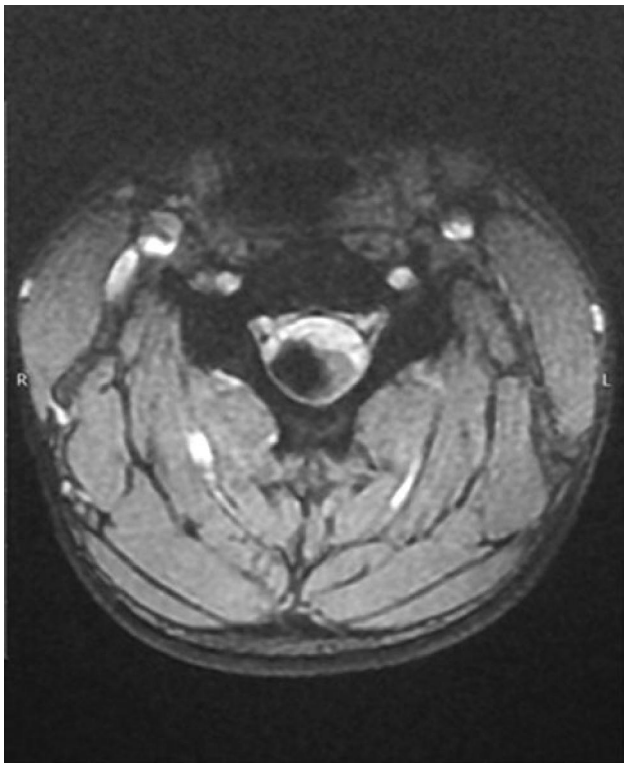

Figure 5: Axial GRE image shows hypointense blooming representing hemorrhage

Discussion

Cavernous angiomas are benign vascular malformations of the central nervous system. They can occurany where in the central nervous system, but are most commonly present in supratentorial compartment. Intramedullary cavernomas are very rare and thoracic spine is found to be the commonest site.[6] The distribution of intramedullary cavernoma reported in the literature is 55 % for the thoracic cord, 40 % for the cervical cord and 5 % for the lumbar conus medullaris.[6]

They are more common in females with peak presentation in fourth decade of life. Patients may present with slowly progressive myelopathy or acute neurological deficit caused by hemorrhage. Histologically, they are characterized by abnormally dilated thin-walled sinusoidal spaces, lined by a single endothelial layer and lack normal intervening nervous tissue.

On imaging, they are often occult on CT scan and angiography.[7] Angiograms cannot visualize cavernous malformations because blood flows through these types of lesions slowly.

Magnetic resonance imaging (MRI), with and without contrast and with gradient echo sequences remains the best means of diagnosing cavernous malformations. They appear as well circumscribed rounded regions of heterogeneous signal intensity on T1 and T2 weighted images due to blood products of varying ages giving typical popcorn ball like appearance. The lesion is surrounded by a complete rim of hypointensity on T2 weighted sequence due to hemosiderin deposition secondary to haemorrhage. The degraded blood product depositions appear as a halo around the lesion and show hypointense blooming on gradient echo sequence. T1 weighted post contrast images show minimal or no enhancement. Absence of significant enhancement rules out possibility of intratumoral hemorrhage. Minimal cord expansion or oedema is present, however 70% of the cases can be complicated by acute massive hemorrhage resulting in significant cord expansion and mass effect.[8]

Repeat MRI scans may be needed to analyze a change in the size of a cavernous malformation, recent bleeding, or the appearance of new lesions.

Total surgical resection should be considered for all symptomatic patients as it has good outcomeand no reported recurrences.[9,10] However studies have also shown long-term morbidity free survival in conservatively treated patients.[11,12] A follow-up MRI examination should be performed to confirm complete removal of the cavernous angioma.

Our patient had all radiological characteristic features of cavernous malformation. This case of intramedullary cavernomais unique not only because it is a rare entity but also due to its unusual presentation in the region of cervical spinal cord in young male patient as most intramedullary cavernoma occur in female patients in forth decade with thoracic spine being the commonest site involved.

Conclusion

Intramedullary cavernous angiomas are rare lesions having characteristic radiological features. MRI plays crucial role in diagnosis and follow-up of these cases. Popcorn like central hyperintensity and peripheral low signal hemosiderin rim are specific imaging features for intramedullary cavernomas. Early diagnosis and proper treatment can prevent bleeding and dangerous enlargement of the lesion.

References

1. El-Koussy M, Stepper F, Spreng A, Lukes A, Gralla J, Brekenfeld C, Sturzenegger M, Schroth G, El-Koussy M. Incidence, clinical presentation and imaging findings of cavernous malformations of the CNS. SwissMedWkly. 2011; **141:** w13172.

2. Mitha AP, Turner JD, Abla AA, Vishteh AG, Spetzler RF: Outcomes following resection of intramedullary spinal cord cavernous malformations: a 25-year experience. J Neurosurg Spine 2011; **14:** 605-11.

3. Ogilvy CS, Louis DN, Ojemann RG. Intramedullary cavernous angiomas of the spinal cord: clinical presentation, pathological features, and surgical management. Neurosurgery. 1992 Aug 1; **31(2):** 219-30.

4. Marconi F, Parenti G, Giorgetti V, Puglioli M. Spinal cavernous angioma producing subarachnoid hemorrhage. Case report. Journal of neurosurgical sciences. Mar 1995; **39(1):** 75-80.

5. Miyoshi Y, Yasuhara T, Omori M, Date I. Infantile Cervical Intramedullary Cavernous Angioma Manifesting as Hematomyelia-Case Report. Neurologia medico-chirurgica. 2010; **50(8):** 677-82.

6. Zevgaridis D, Medele RJ, Hamburger C, Steiger HJ, Reulen HJ. Cavernous haemangiomas of the spinal cord. A review of 117 cases. Actaneurochirurgica. Mar 1999; **141(3):** 237-45.

7. Kharkar S, Shuck J, Conway J, Rigamonti D. The natural history of conservatively managed symptomatic intramedullary spinal cord cavernomas. Neurosurgery. May 2007; **60(5):** 865-72.

8. Maslehaty H, Barth H, Petridis AK, Doukas A, Mehdorn HM. Symptomatic spinal cavernous malformations: indication for microsurgical treatment and outcome. European Spine Journal. Oct 2011; **20(10):** 1765-70.

9. Kivelev J, Niemelä M, Hernesniemi J. Outcome after microsurgery in 14 patients with spinal cavernomas and review of the literature: Clinical article. Journal of Neurosurgery: Spine. Oct 2010; **13(4):** 524-34.

10. Labauge P, Bouly S, Parker F, Gallas S, Emery E, Loiseau H, Lejeune JP, Lonjon M, Proust F, Boetto S, Coulbois S. Outcome in 53 patients with spinal cord cavernomas. Surgical neurology. Aug 2008; **70(2):** 176-81.

11. Steiger HJ, Turowski B, Hänggi D. Prognostic factors for the outcome of surgical and conservative treatment of symptomatic spinal cord cavernous malformations: a review of a series of 20 patients. Neurosurgical focus. Sep 2010; **29(3):** E13.

12. Kharkar S, Shuck J, Conway J, Rigamonti D. The natural history of conservatively managed symptomatic intramedullary spinal cord cavernomas. Neurosurgery. May 2007; **60(5):** 865-72.