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Article

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Intradural spinal, extramedullary, T4-T5 cavernous hemangioma - case report

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Abstract: A very rare, purely intradural, spinal, extramedullary cavernous hemangioma was fortunately discovered in a 56 years old woman, presenting with bilateral brachial paresthesia. Using conventional spin-echo T1 proton density,T2-weighted magnetic resonance and gadolinium images an intradural spinal T4-T5, an extramedullary cavernous hemangioma was discovered. The patient underwent surgery, with laminectomy and microsurgical resection followed by an uneventful postoperative clinical course. Similar as in cerebral locations a mixed signal intensity in all sequences on magnetic resonance images might be indicative of cavernous hemangioma, rendering a presumptive preoperative diagnosis of the lesion and surgical planning for a good microsurgical resection.

Key word: intradural spinal, extramedullary, cavernous haemangioma

Introduction

Cavernous hemangioma, also known as: cavernous angioma, cavernoma, cavernous malformation or hemangioma is a developmental vascular anomaly of dilated blood vessels collection; represent about 7% of all vascular malformations, it can occur anywhere in the central nervous system, with approximately 5% of the cases located in the spine (1-7). Either capillary or cavernous, intradural, spinal, extramedullary hemangiomas can be solitary or multiple; they are well-defined lesions composed of blood-filled endothelial spaces lined by thickened, hyalinised walls without elastic fibers or smooth muscle; they are also very rare (8-16). We report an intra dural spinal extramedullary, T4-T5 cavernoma in a 56-years-old woman, who underwent microsurgical resection.

Case Report

This 56-years-old woman complained of bilateral brahial paresthesia, without neurologic deficits or sphincterian disturbances. The patient underwent cranial and spinal MR examinations and

postoperative MR imaging in a 1,5-Tesla MR imager (General Electric). Sagittal T1 and T2 - weighted images and axial T2 – weighted MR images of the cervico-thoracal spine revealed cervical C4-C6 disc protrusions. Incidentally it was also discovered an intradural, extramedullary lesion at T4-T5, of 6/4 mm – a well-defined postero-lateral lesion, with discreet compression on the spinal cord, which is displaced to anterior and lateral on the right, with heterogeneous signal abnormality in both T1 and T2 – weighted images, representing blood products of various ages, without bone involvement. The lesion was surrounded by a low signal intensity, representing iron storage products and medullary perilesional oedema, occupying most of the thoracic spinal cord. The lesion suggests the diagnosis of an extramedullary thoracal cavernoma (figures 1-3).

Figure 1 - Sagital T2 MRI: a cervical spine examination for bilateral brahial paresthesia - incidental discovery of a lesion in the thoracal spinal canal – an extramedular, intradural lesion, T7 vertebral hemangioma, B & C sagital T2, D sagital T1 after Gadolinium injection – with heterogenous signal in T2w and E sagital T1 w (calcifications possible) + poor enhancement of the lesion

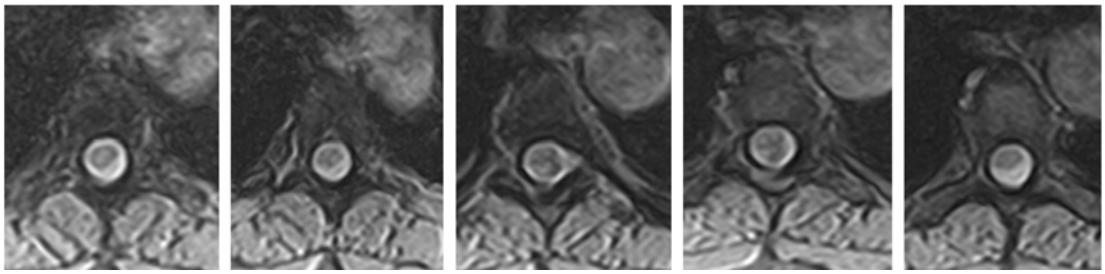


Figure 2 - Axial T2 MRI - emphasize intradural lesion, in postero-lateral left position, with discreet compression over the spinal cord, displaced anteriorly and laterally and to the right

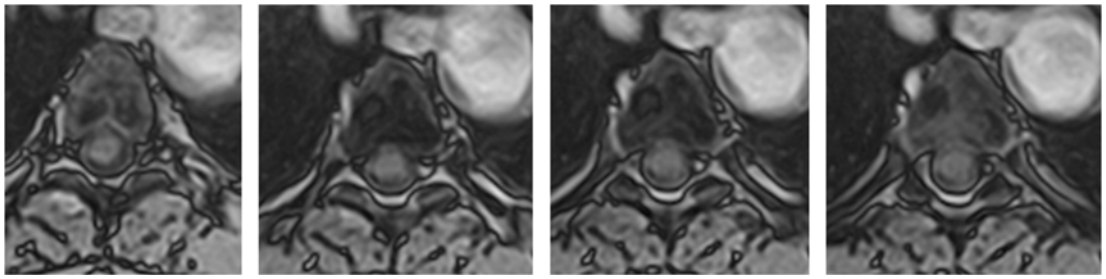
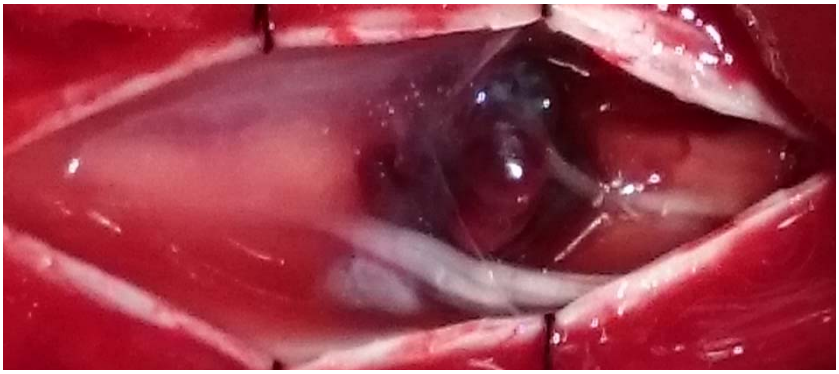


Figure 3 - Axial 3D T1 fatsat postGd – poor enhancement of the lesion

The patient underwent laminectomy from T4 to T5 using a microsurgical approach. When the dura and arachnoid were opened, an extradural polilobulated, violet-dark red mass, of 6/4/3 mm, located dorsally, was identified. The tumor exerted discreet compression over the spinal cord, displacing it anteriorly and laterally to the right. Clearly demarcated from the adjacent spinal cord tissue, with a good cleavage from the dura, the tumor was completely excised and gently extracted from the the hemosiderin-stained bed of the spinal cord (figure 4). After the lesion has been completely resected, no extra-spinal extension or bony involvement was found.

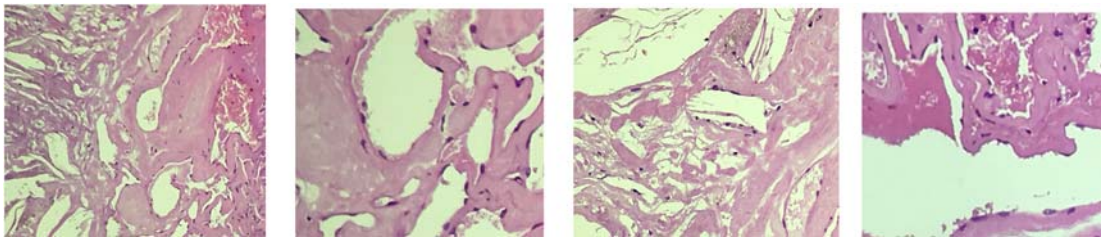
The histological aspect was characteristic of a cavernous hemangioma: multiple, closely spaced - lined by a single layer of benign endothelial cells, dilated vascular channels containing blood, without nuclear pleomorphism and mitotic figures (figures 5-7).



A

B

Figure 4 - Pre- and postoperatory view



A B C D

Figure 5 - Histologic cavernous hemangioma features col HE 10×10 A-D: Dilated vascular channels filled with red blood cells, lined of endothelial cells; without intervening neural tissue and vascular wall muscle fibers

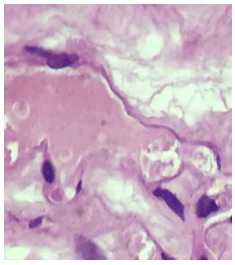


Figure 6 - Cavernous hemangioma, HE coloration 10×40, Detail image: dilated and narrowed vascular channels with red blood cells

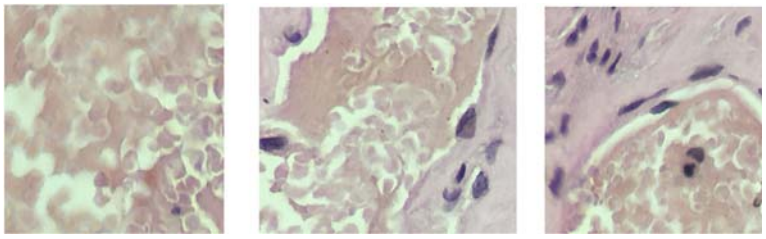


Figure 7 - Cavernous hemangioma, van Gieson coloration 10×40, Detail images, dilated vascular channels with red blood cells

Postoperatively evolution of the patient was uneventful. No adjuvant therapy was given. Six months after surgery, a follow-up MRI scan was performed, showing no evidence of secondary lesion or recurrence.

Discussion

Spinal cavernous and capillary hemangiomas can be found in two forms: sporadic 90% and familial 10% (17); they can be solitary or multiple, and they may be associated with similar lesions in other organs of the body in up to 18.7% of cases (18).

Spinal cavernous hemangioma (1-22) can be found in several topographic locations: confined to the vertebrae, extended epidurally from the vertebral lesions, entirely extradural lesions with no bone involvement, and also purely intradural, extramedullary from the inner surface of the dura or the pial surface of the spinal cord; very rarely, intramedullary in about 3% of cases (21)(24), adherent to the blood vessels of the nerve roots in the cauda equina (16). Intradural spinal, extramedullary, cavernous hemangiomas are most frequently found in the adult population (only 4 pediatric cases cited in literature), mostly in women (23), in the thoracic spine - 80% of cases (9)(12) or lower thoraco-lumbar region (13)(20), with posterior location within the spinal canal in 93% of cases; also with possible lateral recesses extensions; less common in cervical location (7)(8) and mostly adherent to the nerve root or spinal cord (8)(24). Capillary hemangiomas are found in the skin and soft tissues in younger people, and rarely occur in the central nervous system; more frequent they are located around or attached to nerve roots of cauda equina and conus medullaris (25), extremely rarely intradurally (16). Intradural extramedullary capillary hemangiomas tend to present in the fourth or fifth decade of life (mean age: 49 years), with male predominance, especially in the thoracic spine (between T4 and T11 vertebrae) and in the lumbar or conus medullaris region. (1, 2, 4, 5, 9, 13, 16, 25)

Clinical symptomatology at presentation is variable, depending on the size and topography, with the more frequent symptoms beeing: acute spinal pain, radiculopathy and/or myelopathy, progressive gait disturbance, slowly progressive paraparesis, or even asymptomatic – very rare, as in our case (5, 18, 26-28). Cavernous hemangiomas may present in four major clinical patterns: acute episodes of step wise deterioration, slow progression, acute onset with rapid deterioration, and acute onset with gradual decline (10, 12, 14, 15, 18). Acute clinical deterioration is present in cavernous hemangiomas associated with subarachnoid hemorrhage induced by intralesional hemorrhage, lesion growth, thrombotic venous occlusion; bleedind can occur due to the thin-walled vessels, stasis of blood flow in the lesion, estrogen mediated neoangiogenesis in the lesion or drainer compression by a gravid uterus (22)(23)(29). Intradural extramedullary lower thoracic spinal cord capillary hemangiomas can have a variable onset of presentation: low back pain, radiculopathy or cauda equine syndrome (13)(25)

Cavernous hemangioma are diagnosed by MRI - the investigation of choice; these lesions have no communication with the spinal arterial circulation and are angiographically occult (30-32). Computed tomography scan may show hyperdense or calcified lesions on plain studies. There may be minimal or no enhancement with contrast (18)(32).

The MRI signal intensity and character of the spinal epidural cavernous hemangioma reflects its histopathology:

- both T1- and T2-weighted images lack of a low-signal hemosiderin ring relate to the more rapid removal of blood degradation products outside the blood-brain barrier (31-32)

- low or intermediate signal intensity on T1-weighted, and high-signal intensity on proton density and T2-weighted images, with strong homogeneous enhancement after contrast medium injection (14)

- marked hyperintense T2 - weighted signal reflects the high water content and the intense flow enhancement into the numerous vascular channels of the lesion - sinusoidal vascular structure (36).

- heterogenous signal on all pulse sequences, with ring enhancement on post-contrast images – rare, due to intralesional hemorrhage and related degeneration (37)

- hyperintense signal has been reported on both T1 and T2 - weighted images, related to the presence of hemorrhage in the subacute phase (14)(38)

- profoundly hypointense signal areas in the T2 and gradient echo sequences due to acute hemorrhage containing deoxyhemoglobin or as a result of hemosiderin deposition related to recurrent hemorrhages (39).

- complete lack of enhancement in case of sclero-hyaline degeneration

Features of capillary hemangioma on MR images are: isointense lesion on T1-weighted images, hyperintense relative to the spinal cord on T2-weighted images, and homogenous, strong enhancement on contrast-enhanced T1-weighted images (9, 25, 40, 41), and also the presence of enlarged draining perimedullary veins (9).

Capillary and cavernous hemangiomas arise from the same cell type, showing distinct demarcation from the surrounding parenchyma. They can be differentiated on histopathology by vessel size: capillary hemangioma, uncommon in the spinal canal, is composed of small, capillary networks surrounded by collagenous stroma (2), without hemosiderin deposition, while the cavernous hemangioma is composed of irregular, dilated sinusoidal vascular channels lined by a monolayer of benign endothelium, with the large sinusoidal lumina almost adjacent to one another, without intervening mother tissue (6)(8)(11)(12). Moderate stromal chronic inflammation is often present. These are well-circumscribed lesions, with discrete borders, dark to purple in color, and can be multilobulated. Associated calcification and thrombosis may occur. The blood flow in these lesions is slow and, therefore, it is not visualized in standard angiography. Hemorrhages are common in the intra-axial lesions, but less frequent in extra-axial ones.

The usual differential diagnosis of intradural, extramedullary, epidural cavernous hemangioma (8)(11)(14) occurring at the thoracic level is made with other spinal epidural tissue masses:

- schwannoma: frequently seen in the middle age, with equal sex predilection; the signal intensity usually is hypo-intense, less frequently iso-intense, on T1-weighted images, hyper-intense on T2-weighted images, with cystic changes or necrosis on MRI

- meningioma: it is seen the 50–70 years age group, with M:F=1:5; the mass usually shows iso-intensity or slight hypo-intensity on T1-weighted images, and iso-intensity or slight hyper-intensity on T2-weighted images; it has a characteristic broad-based dural attachment, with dural tail sign on the contrast-enhanced study. Sometimes it is difficult to differentiate the intradural extramedullary capillary hemangioma from the other common intradural tumors by MRI.

- nerve sheath tumors are linear, ovoid or characteristically dumbbell-shaped in configuration, of uniform hyper-intense signal in T2

- disk prolapse - ruled out for its lack of anatomic connection with the neighboring intervertebral disk or the exiting nerve root, after Gadolinium injection – with heterogeneous signal in T2 (20)

- paraganglioma rarely occurs epidurally, with heterogeneous salt and pepper appearance due to vascular signal voids on MR imaging mimicking capillary hemangioma (6)(42)

- epidural lymphoma shows characteristic low T2 signal

- drop metastasis usually appears as multiple small nodular and/or linear enhancing lesions within the thecal sac (10)

- filum terminale ependymoma may present intratumoral cystic changes and a less strong degree of contrast enhancement

- round cell tumor

- eosinophilic granuloma, ruled out for the absence of any bony changes

- sarcoidosis (43), lymphoma with a discrete solitary mass, with a multifocal patchy or linear enhancing lesion

- histiocytosis, angiolipoma - representing less than 6% of all spinal neoplasms

- tuberculosis is uncommon, mostly epidural, associated with other systemic signs and symptoms as well as with mediastinal lymphadenopathy, with craniocaudal extension along to more than one vertebral body.

Complete surgical resection is the treatment of choice for intradural extramedullary hemangioma, with no recurrence (3)(6-9)(12)(14)(15)(24)(25)(37). Microsurgical excision of such lesions, in accessible locations, especially for lesions located posteriorly, is the treatment of choice. Because of sudden intralesional hemorrhage, which can lead to spinal cord compression and subsequent severe disability, cavernous hemangioma should be included in the differential diagnosis of purely extradural soft tissue lesions of the spine.

Conclusion

Intradural spinal, extramedullary, cavernous hemangioma are very rare; early recognition by accurate diagnosis using MRI, followed by complete excision of the lesion is important, because of the risk of enlargement or hemorrhage, which may produce sudden neurologic deterioration. Because resection is in general possible without causing morbidity, and because the outcome depends on the severity of preoperative neurological dysfunction, precise diagnosis and timely treatment are mandatory.

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