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Image Report

A cervical solitary fibrous tumor with intramedullary invasion

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ABSTRACT

Solitary fibrous tumor is a tumor originating from the mesenchymal cells, which occurrence in the central nervous system is extremely rare and was described in few patients as to yet. We report on a 53-years old male patient presenting with right upper limb radicular pain and ipsilateral limbs paresis, who was diagnosed with a cervical spinal lesion which, after surgical resection, resulted to be a solitary fibrous tumor (SFT). We discuss imaging, clinical and histopathological findings to allow considering this tumor early in the differential diagnosis.

Keywords: Solitary fibrous tumor, Intramedullary spine tumor, Spinal cord compression

Solitary fibrous tumor is usually a benign neoplasm originating from mesenchymal cells, historically associated with the pleura. Its occurrence in the central nervous system is extremely rare. [8]

Clinical and imaging findings of SFT are not specific and it is considered a great mimicker, [11] thus making misdiagnosis a possible occurrence.

The proper and early diagnosis is crucial due to the higher propensity to both local as well as distant recurrences compared to similar pathologies like meningiomas.

We describe a 53-year-old male patient presenting with the right upper limb radicular pain and ipsilateral limb paresis. MRI of the cervical spine revealed a large spinal mass at the level of the sixth and seventh cervical vertebral bodies. The lesion appeared isointense on T1-weighted sequences and hypointense on T2-weighted images and was homogeneously contrasted by gadolinium [Figures 1a-d].

The initial suspected diagnosis was that of a meningioma, and surgical resection through a laminectomy was planned. Intraoperatively, the lesion appeared gray-whitish and was present within the intramedullary space without a clear cleavage plane with the spinal cord [Figure 2].

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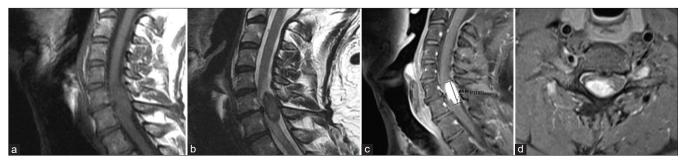


Figure 1: (a-d) Preoperative MRI aspect of the lesion, showing isointensity in T1 (a), hypointensity in T2 (b) and a homogeneous contrast enhancement lesion (c and d).

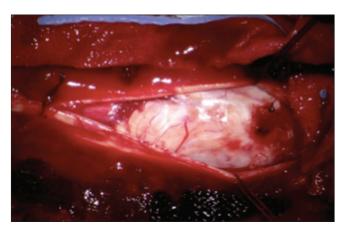


Figure 2: Intraoperative microscopical view after the opening of dura mater and hemostasis, showing the gray-whitish intramedullar lesion that includes radicles.

The surgical procedure was uneventful and complete resection was achieved. Postoperatively, the patient presented with gait ataxia that completely recovered over time with the help of intensive physical rehabilitation.

Histological analysis revealed the main diagnostic characteristics of SFT: presence of cells organized into fascicles and hypocellular areas with fibrous stroma, immunoreactivity for CD34 and bcl-2, evidence of nuclear expression of STAT6, and the absence of mitosis or necrosis [Figures 3a, b]. From a histopathological point of view, the neoplastic cells in our case of SFT resulted furthermore negative for Epithelial Membrane Antigen, which is marker, for example, meningiomas.

MRI alone cannot reliably distinguish SFT from other intradural tumors such as meningioma, schwannoma, astrocytoma, and ependymoma.[11] It has been proposed that SFT can be suspected in the presence of a black and white mixed pattern ("Ying-Yang" sign) on T2-weighted MRI imaging, showing marked heterogeneous contrast enhancement.[12]

During surgery, the mass typically shows a hard consistence and poor vascularization.^[9] Although SFT mostly manifest benign behavior, malignant subtypes and future malignant

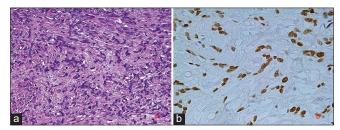


Figure 3: (a and b) SFT composed of spindled monomorphic cells admixed with collagenous stroma and capillaries (Hematoxilin-Eosin, ×2.5) (a), STAT6 immunoistochemistry: nuclear expression in neoplastic cells; endotelial cell are negative (STAT6, ×30) (b).

conversion are possible and must be taken into consideration.

Imaging is not helpful in predicting the grade of malignancy.^[2] Metastasis can be found even years after the total resection of an SFT.[5] Local recurrence and metastases occur in about 10-15% of SFT in general.^[10] A case of extramedullary cervical SFT recurrence 5 years after the first surgery was recently reported by Chen et al.[3] Long-term follow-up is, therefore, recommended, for at least 10 years.[4] The number of reported SFT is increasing over time. This is likely related to recently developed histological characterizations. In the past, this tumor entity may have been mistaken for other tumors such as fibrous meningioma and hemangiopericytoma. Although cervical fibrous tumor is a known entity, its rare and unspecific nature can make it easy to misdiagnose as our case demonstrates. In 4% of cases, hypoglycemia is associated with SFT. We did not encounter this situation in the described case. [6]

Treatment of SFT consists of total surgical excision. Radiotherapy and chemotherapy are suggested in cases of incomplete resection.^[1,7] It is important to consider this rare tumor entity in the early differential diagnosis of any mass in the spinal canal to ensure prompt diagnosis and adequate treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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