Solitary fibrous tumors (SFTs) are rare mesenchymal neoplasms of fibroblastic origin. They commonly arise from visceral pleura, but also arise from nonserosal sites such as meninges, central nervous system parenchyma, and spinal cord. In the spinal cord, SFTs commonly arise from the thoracic spinal cord, followed by cervical spinal cord, lumbar spinal cord, and sacrum. Histologically, SFTs can be similar to hemangiopericytoma, schwannoma, fibrous meningioma, fibroma, gliofibroma, and ependymoma. Immunohistochemistry (IHC) plays an important role in differentiating SFTs from other identical tumors. Here, we report a rare case of SFT of the cervical spinal cord, which was initially reported as hemangiopericytoma, and the diagnosis of SFT was confirmed by IHC.

**Keywords:** Cervical spine, immunohistochemistry, solitary fibrous tumors

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### Résumé

Les tumeurs fibreuses solitaires (SFT) sont des néoplasmes mésenchymateux rares d’origine fibroblastique. Ils proviennent généralement de la plèvre viscérale, mais aussi proviennent de sites non séreux tels que les méninges, le parenchyme du système nerveux central et la moelle épinière. Dans la moelle épière, les SFT proviennent de la moelle épière thoracique, suivie de la moelle épière cervicale, de la moelle épière lombaire et du sacrum. Histologiquement, les SFT peuvent être similaires à hémangiopéricyctome, schwannome, méningiome fibreux, fibrome, gliofibrome et épendymome. L’immunohistochimie (IHC) joue un rôle important dans la différenciation des SFT des autres tumeurs identiques. Ici, nous rapportons un rare cas de SFT de la moelle épière cervicale, qui a été initialement signalé comme hémangiopéricyctome, et le diagnostic de SFT a été confirmé par IHC.

**Mots-clés:** rachis cervical, immunohistochimie, tumeurs fibreuses solitaires

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### Introduction

Solitary fibrous tumors (SFT) are ubiquitous rare spindle cell neoplasms, commonly arising from visceral pleura.[1] These tumors can also arise from nonserosal sites such as meninges, central nervous system parenchyma,[2] and spinal cord and spinal nerve involvement have been reported.[2,3] They were first described by Klemperer and Rabin in 1931, and they proposed a submesothelial origin.[4] It is now believed that SFTs are mesenchymal neoplasms of fibroblastic origin. Initially, SFTs were grouped together with hemangiopericytomas, but with the advent of immunohistochemistry (IHC), they were separated from hemangiopericytomas. Most of these lesions are benign, but local recurrence and metastases have been reported in the number of cases.[5] Pleural SFTs are more likely to be malignant when compared with extrapleural SFTs.[6] Spinal cord SFTs are a rare entity commonly occurring in the thoracic spinal cord, followed by the cervical and lumbosacral spinal cord. Here, we report a case of spinal SFT arising from the cervical spinal cord. The biopsy was suggestive of hemangiopericytoma, but the final diagnosis of SFT was confirmed after IHC.

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A 16-year-old female patient presented with progressive weakness of the right upper and lower limbs for 3 months. She also had radiating pain over her right upper limb. Her general physical examination was within the normal limits. Neurological examination revealed a power of 4/5 for the right upper limb and 3/5 for the right lower limb. There was absence of flexion of the right upper limb phalanges, and the foot drop was observed on the right lower limb. Magnetic resonance imaging (MRI) of the cervical spine revealed a well-defined intense uniform enhancing T2 hyperintense and T1 hypointense intradural and extramedullary mass lesion at C5-C7 level.

A C5-C7 laminectomy was performed. Tumor was found on the right side of the spinal cord. It was greenish white in color and moderately vascular. The lesion was released from the dura and then from adjacent arteries; venous connections of lesion were completely resected.

Histological examination of the tumor showed short spindle to epithelioid cells arranged in diffuse sheets having scant cytoplasm and indistinct cell borders. Nuclei are round to oval with dense chromatin and inconspicuous nucleoli. Many staghorn type blood vessels lined by the endothelium are noted. Mitosis is brisk, and focal fibrinous necrosis is also made out. With the above histological findings, a diagnosis of hemangiopericytoma was given.

IHC of the tumor showed a strong positive staining for CD34 and Vimentin and negative staining for CD31 and smooth muscle actin. Ki-67 proliferation index was 10%. With the above immunohistological findings, the final diagnosis of SFT was given.

The postoperative period was uneventful, and the patient was asked to follow-up after 1 month for neurological examination and check MRI.

**DISCUSSION**

SFT is a rare neoplasm of mesenchymal origin commonly arising from an intrathoracic location. However, SFTs in extrathoracic locations such as spinal cord, head and neck, extremities, abdomen, pelvis, and retroperitoneum have been reported.[7-9] As pleura was the most common site of involvement, SFT was initially thought to be mesothelial in origin. Further, characterization with immunohistochemical stains and its ubiquitous presence determined the mesenchymal origin.[1]

Muñoz et al. reported that the mean age at diagnosis of spinal SFT was 46.5, and male-to-female ratio was 1.4:1.[10] Common symptoms of spinal cord SFT include pain, myelopathy, radiculopathy, and motor or sensory deficit. Predominant location of spinal cord SFTs was thoracic spine (53.3%), followed by the cervical spine (33.4%), lumbar spine (6.7%), and sacrum (3.3%).[11] Most of the spinal cord SFTs are intramedullary or intradural and extramedullary. In our case, the age of the patient (16 years) is far less than the mean age at diagnosis of spinal SFTs, but the symptoms are in line with relevant literature.

Radiologically, Fargen et al. reported that two-thirds of lesions were isointense on T1-weighted MRI, and others were either heterogeneous or hypointense. Two-thirds of cases were hypointense on T2-weighted MRI, and 17% cases were hyperintense. Over 78% of cases demonstrated diffuse or homogeneous contrast enhancement with gadolinium, and 21% of cases demonstrated only partial or heterogeneous enhancement.[12]

Histologically, SFTs can be similar to hemangiopericytoma, schwannoma, fibrous meningioma, fibroma, gliofibroma, and ependymoma.[13] IHC is important to differentiate SFT from the above-mentioned tumors. SFTs are strongly positive for CD 34 and Vimentin and negative for epithelial membrane antigen and S-100 protein.[14] Hemangiopericytoma may be positive for CD 34 antigen, but are typically hypercellular with mitosis and
necrosis. SFT normally exhibits strong and diffuse reactivity for CD 34, whereas hemangiopericytoma shows patchy and focal reactivity for CD 34. Correct differentiation between SFT and hemangiopericytoma is very important, as the latter is more aggressive and requires radiotherapy and chemotherapy postoperatively.

SFTs are usually benign, but malignant cases have been reported. Malignancy or high risk of recurrence is associated with hypercellularity, marked nuclear atypia, high mitotic activity, necrosis, high Ki-67 proliferation index, and subtotal resection. However, according to Muñoz et al., histological findings of SFT are not reliable for predicting the behavior, as they presented a case of malignant SFT with multiple metastases but had benign histological features.

The primary treatment of SFT is complete surgical excision. Recurrences after total resection are uncommon. In cases where resection is incomplete, adjuvant treatment with radiotherapy or chemotherapy is suggested.

In our case, complete resection of tumor was done, and as her Ki-67 was 10%, she was advised follow-up at regular intervals for a long duration.

**Conclusion**

SFT is a rare spindle cell tumor, which is difficult to diagnose radiologically and histologically. IHC plays an important role in the diagnosis of SFT and in differentiating other spindle cell tumors. In spite of SFTs being benign with indolent course, cases with malignant transformation and late recurrences have been reported. Hence, the regular follow-up is advised for a long duration.

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**Conflicts of interest**

There are no conflicts of interest.

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