A cervical solitary fibrous tumor with intramedullary invasion

Andrea Ciuffi1#, Christian Saleh2#, Maria Rosa Terreni3, Phillip Jaszczuk4#, Edvin Zekaj5, Claudia Menghetti5, Andrea Franzini6, Domenico Servello5

1Department of Neurosurgery, Fondazione IRCCS Istituto Neuroligico Carlo Besta, Milan, Lombardia, Italy, 2Clinic for Forensics, University Psychiatric Clinic Basel, Basel, Switzerland, 3Department of Pathology, IRCCS San Raffaele Scientific Institute, Milan, Italy, 4Department of Spinal Surgery and Spinal Cord Surgery, Swiss Paraplegic Centre, Notwill, Switzerland, 5Department of Neurosurgery, IRCCS Istituto Ortopedico Galeazzi, Milan, 6Department of Neurosurgery, IRCCS Humanitas Research Hospital, Rozzano, Lombardia, Italy.

E-mail: *Andrea Ciuffi - andreciuffi@hotmail.it; Christian Saleh - chs12us75010@yahoo.com; Maria Rosa Terreni - terreni.mariarosa@hsr.it; Phillip Jaszczuk - philja.ja@gmail.com; Edvin Zekaj - ezekaj@yahoo.com; Claudia Menghetti - menghetti@gmail.com; Andrea Franzini - andrea.franzini1@hotmail.it; Domenico Servello - servello@libero.it

*These authors have equally contributed to this work.

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. Clinical and imaging findings of SFT are not specific and it is considered a great mimicker, 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].

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, transform, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

©2022 Published by Scientific Scholar on behalf of Surgical Neurology International
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. 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. During surgery, the mass typically shows a hard consistency and poor vascularization. Although SFT mostly manifest benign behavior, malignant subtypes and future malignant conversion are possible and must be taken into consideration. Imaging is not helpful in predicting the grade of malignancy. Metastasis can be found even years after the total resection of an SFT. Local recurrence and metastases occur in about 10–15% of SFT in general. A case of extramedullary cervical SFT recurrence 5 years after the first surgery was recently reported by Chen et al. Long-term follow-up is, therefore, recommended, for at least 10 years. 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.

Treatment of SFT consists of total surgical excision. Radiotherapy and chemotherapy are suggested in cases of incomplete resection. 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.
Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Allen AJ, Labella DA, Richardson KM, Sheehan JP, Kersh CR. Recurrent solitary fibrous tumor (Intracranial hemangiopericytoma) treated with a novel combined-modality radiosurgery technique: A case report and review of the literature. Front Oncol 2022;12:907324.
2. Brigui M, Aldea S, Bernier M, Bennis S, Mireau E, Gaillard S. Two patients with a solitary fibrous tumor of the thoracic spinal cord. J Clin Neurosci 2013;20:317-9.
3. Chen Y, Xu Z, Liu M, Xu H. Recurrent solitary fibrous tumor of the spinal cord: A case report and literature review. Clin Neuropathol 2020;39:86-91.
4. Flores-Justa A, López-García E, García-Allut A, Reyes-Santías R. Tumor fibroso solitario/hemangiopericitoma de la médula espinal. Neurocirugía 2018;29:309-13.
5. Hwang U, Kim SB, Jo DJ, Kim SM. Intramedullary solitary fibrous tumor of cervicothoracic spinal cord. J Korean Neurosurg Soc 2014;56:265-8.
6. Jang JG, Chung JH, Hong KS, Ahn JH, Lee JY, Jo JH, et al. A case of solitary fibrous pleura tumor associated with severe hypoglycemia: Doege-potter syndrome. Tuberc Respir Dis (Seoul) 2015;78:120-4.
7. Koduru MR, Ail S, Shetty J, Shetty V. Solitary fibrous tumour of cervical spinal cord. Ann Afr Med 2020;19:144-6.
8. Louis D, Perry A, Reifenberger G, Von Deimling A, Figarella-Branger D, Cavenee WK, et al. The 2016 world health organization classification of tumors of the central nervous system: A summary. Acta Neuropathol 2016;131:803-20.
9. Mariniello G, Napoli M, Russo C, Briganti F, Giumundo A, Maiuri F, et al. MRI features of spinal solitary fibrous tumors. Neuroradiol J 2012;25:610-6.
10. Nagano A, Ohno T, Nishimoto Y, Oshima K, Shimizu K. Malignant solitary fibrous tumor of the lumbar spinal root mimicking schwannoma: A case report. Spine J 2014;14:e17-20.
11. Sebaaly A, Raffoul L, Moussa R. Solitary fibrous tumor of the lumbar spine: The great mimicker report of the fifth case. Case Rep Orthop 2014;2014:852830.
12. Weon YC, Kim EY, Kim HJ, Byun HS, Park K, Kim JH. Intracranial solitary fibrous tumors: Imaging findings in 6 consecutive patients. AJNR Am J Neuroradiol 2007;28:1466-9.

How to cite this article: Ciuffi A, Saleh C, Terreni MR, Jaszczuk P, Zekaj E, Menghetti C, et al. A cervical solitary fibrous tumor with intramedullary invasion. Surg Neurol Int 2022;13:343.