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Letter to the Editor: New Observation

Cervical Intramedullary Solitary Fibrous Tumor

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Solitary fibrous tumors (SFTs) are rare spindle-cell tumors of fibroblastic origin most commonly found in the pleura.[1] They can also present in the central nervous system (CNS),[1] either intracranially or in the spinal cord. It is estimated that for every 10 intracranial SFTs, there is one spinal SFT.[2] They are largely considered benign, but recurrence has been reported especially when the tumor is not completely excised, which is a likely outcome in intramedullary tumors.[3]

A 40-year-old previously healthy female presented with a 3-month history of progressive sensory disturbance in her lower extremities and right hand with associated gait disturbance. Physical exam demonstrated intrinsic hand weakness, bilateral patellar hyperreflexia, and an inability to dorsiflex the left foot. Posterior column function of the lower extremities was impaired.

Magnetic resonance imaging (MRI) demonstrated an intramedullary lesion with an exophytic component causing significant cord compression with associated signal change representing edema from C2-T3 (Figure 1).

The patient's symptoms started to worsen, and she underwent an urgent C5-C7 cervical laminectomy with subtotal resection of the tumor. Because the lesion was intimately related to the substance of the spinal cord, a subtotal resection was conducted rather than gross total resection (GTR) in order to avoid the risk of causing significant neurological dysfunction. The lesion had an exophytic component with the consistency of a firm, fibrous tumor.

Upon initial pathological investigation, the specimen demonstrated fascicles of tightly packed, spindle shaped cells that alternated with areas of dense collagen and decreased cellularity suggestive of a palisading pattern (Figure 2A). Reticulin staining showed wrapping of individual cells (Figure 2C). Neither mitotic activity nor necrosis was present.

Immunohistochemistry was performed which revealed nuclear STAT6 expression. Cells also displayed cytoplasmic CD34 and vimentin expression (Figure 2C-F) in the absence of EMA and S-100 expression. Ki-67 proliferation index was 2%. The diagnosis was therefore a WHO Grade 1 SFT.

Following her subtotal resection, the patient was sent for adjuvant radiotherapy consisting of 54 Gy in 30 fractions. Subsequent follow-up imaging showed residual tumor but no further growth or metastases. Her neurological status greatly improved and has become stable at 13 years post-op with no signs of recurrence.

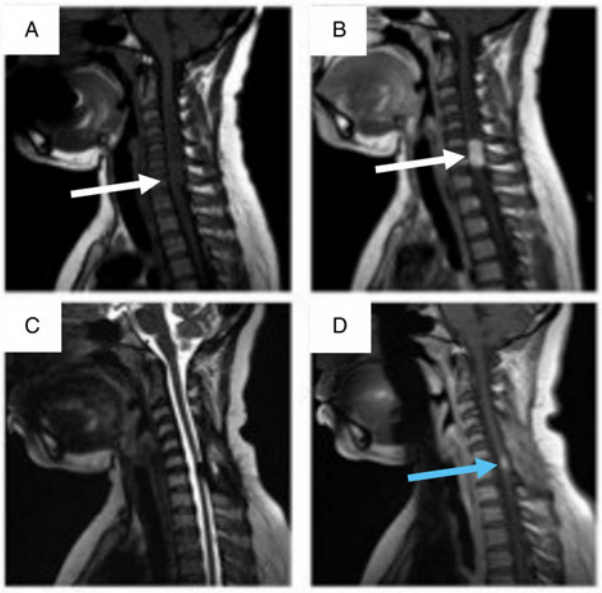

Figure 1: T1-weighted preoperative precontrast (A) and postcontrast (B) sagittal MRI demonstrates marked enhancement of this lesion at the level of C5-C7. Sagittal T2 (C) and T1 postcontrast (D) MRI images demonstrate minimal residual tumor 1 year postoperatively.

SFTs of the CNS are estimated to account for 0.09% of all meningeal tumors and can mimic other tumors such as meningioma or schwannoma.[4] To our knowledge, there have been only 41 known cases reported in the spinal cord, 22 of which were intramedullary.[2,3] The most common location of spinal SFTs is in the thoracic spine, followed by the cervical, lumbar, and finally sacral spine.[2]

Histologically, these tumors are composed of spindle-shaped cells arranged in short wavy fascicles against a collagenous background stoma, with alternating areas of hypo- and hypercellularity,[4] as illustrated in our patient's case in Figure 2.

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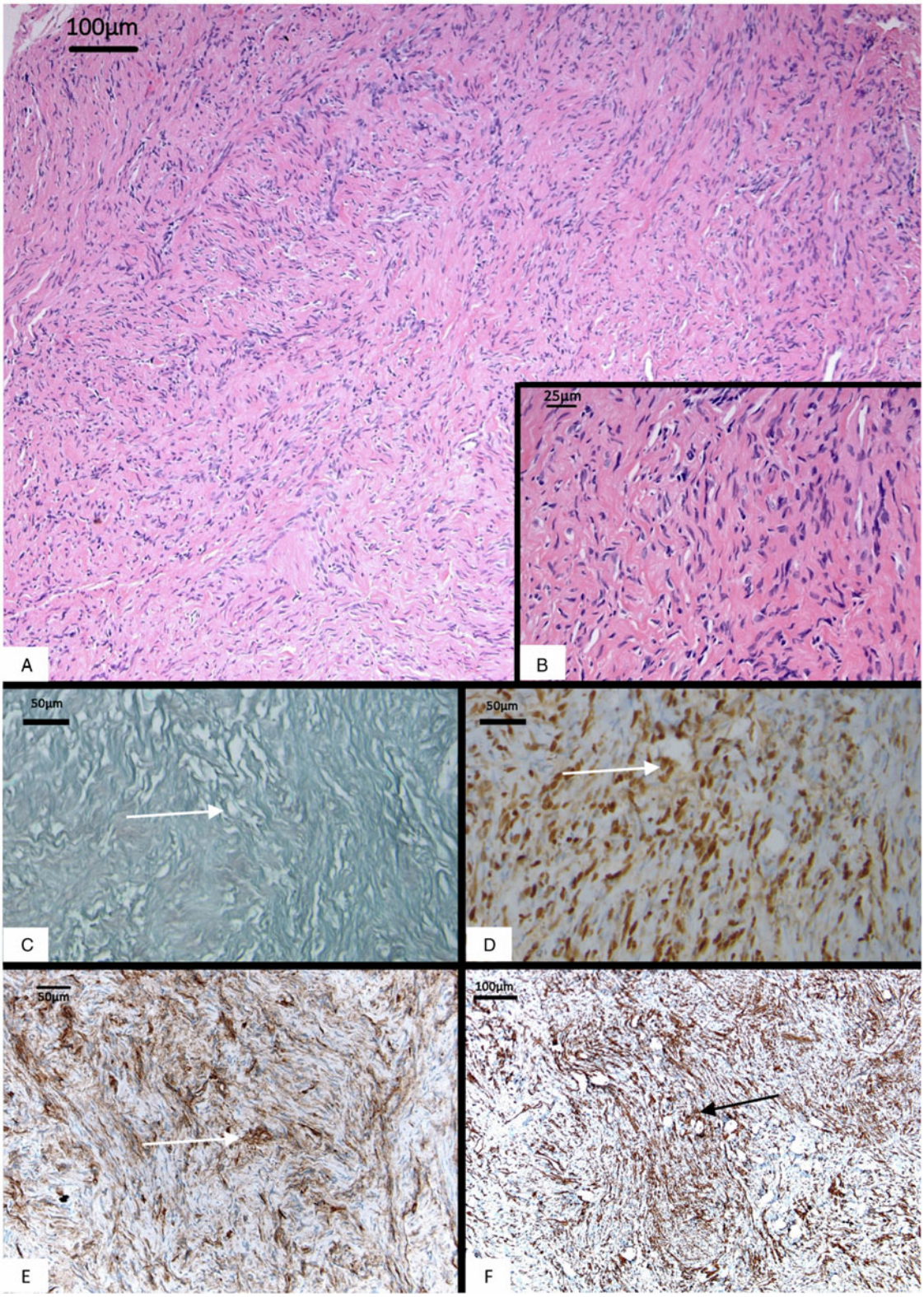

Figure 2: (A) Hematoxylin and eosin (H&E) stain of the tumor illustrates the pseudopalisading nature of the lesion with interspersed areas of pauci-cellular dense connective tissue [×10]. (B) The spindle-shaped cells [×40]. (C) Reticulin stain shows wrapping of individual cells (white arrow) [×20]. (D) STAT6 nuclear expression (white arrow) [×20]. (E) and (F) Cytoplasm stained for CD34 [×20] and vimentin [×10], respectively.

To definitively diagnose SFTs of the CNS. The most sensitive and specific marker for SFTs is Signal Transducer and Activator of Transcription 6 (STAT6).[5] The nuclear relocation of STAT6 that is detected with IHC is now known to point to the presence of a NAB2-STAT6 gene fusion.[6] Furthermore, meningeal hemangiopericytomas have also been characterized by the same NAB2-STAT6 gene fusion. As a result of this finding, the updated 2021 World Health Organization (WHO) classification of CNS tumors

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has entirely retired the term "hemangiopericytoma," with the tumor now only referred to as SFT and with STAT6 overexpression being deemed mandatory for definitive diagnosis.[6,7] Interestingly, certain variants of this gene fusion have been linked to increased rates of recurrence in meningeal SFTs.[2] No further studies have been done to investigate whether testing for variants could be used as a prognostic factor, but future research may be able to elucidate this. SFTs also consistently stain positive for CD34 and vimentin, as seen in our patient (Figure 2E,F).[5]

Once a diagnosis is reached, the next crucial step is to determine the grade in order to risk-stratify the tumor. SFTs have been graded by WHO into three grades, and studies have used this grading system as a prognostic indicator for patients with CNS SFTs.[1,7] This grading system includes factors such as mitotic count and areas of increased cellularity. A large study composed of 132 patients concluded that mitotic count should not be the sole prognostic indicator and that additional histological criteria, including necrosis, can better predict tumor behavior and metastasis.[7] In terms of risk of recurrence, prognostic factors differ slightly. Studies have suggested that higher-grade SFTs were significantly associated with an increased likelihood of recurrence. They also demonstrated that GTR and adjuvant radiotherapy reduced recurrence and that GTR was the single-most effective method to improve overall survival.[1,8] Our patient had a WHO Grade 1 CNS SFT, thus conferring a more favorable prognosis. From a treatment point of view, however, she only received a subtotal resection due to the surgically difficult intramedullary nature of the lesion. The intimate relationship with the spinal cord likely warrants GTR only with the aid of intraoperative neurophysiology, which may not always be available or appropriate. Most of the 22 reported intramedullary spinal SFT cases underwent GTR[3] which results in a sparsity of evidence in relation to those with intramedullary lesions who received subtotal resection, such as our case. Patients who undergo subtotal resections have an estimated 5-year survival of 75% and an estimated 40% recurrence rate,[2] thus emphasizing the need for effective adjuvant therapies that could reduce the risk of recurrence in those unable to undergo GTR. This case therefore presents a patient who underwent a subtotal resection followed by radiotherapy and has been stable 13 years postoperatively. This suggests that despite subtotal resection, radiotherapy may play a role in preventing recurrence and can therefore be used as an effective method of adjuvant treatment for future cases.

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Conflicts of interest. The authors would like to disclose that Dr Sharma has received personal consulting fees from Moderna Pharmaceuticals, Dr Munoz receives payment for expert testimony from Linden and Associates and consulting fees from Cortexyme.

Statement of authorship. EA, SS, and JWC wrote the initial manuscript, JS was the primary neurosurgeon looking after the patient, DGM performed the neuropathological analysis and provided the histology images, JWC was the reporting radiologist and provided the radiology images, and DGM reviewed and edited the manuscript. All authors have read and agreed to publish the final version of this paper.

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