Case Report

Intracranial Solitary Fibrous Tumor with Pseudopapillary Architecture: An Uncommon Tumor with Unusual Histopathology

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Solitary fibrous tumor (SFT) is an uncommon soft tissue neoplasm first described in the pleura in 1931 and later recognized in other anatomical locations. Involvement of the central nervous system is rare; there are approximately 250 cases of central nervous system SFTs in the English literature. To the best of our knowledge, this is the first report of an intracranial SFT showing pseudopapillary architecture. The patient was a 73-year-old male who presented with recent onset altered mental status. Imaging studies showed a large left posterior parieto-occipital region intradural extra-axial mass with significant mass effect in the adjacent brain. Neuropathologic examination demonstrated a spindled mesenchymal neoplasm with variable cellularity and prominent collagen deposition. In areas, the tumor was discohesive, imparting a prominent pseudopapillary architecture. Entrapped brain parenchyma was present, indicating brain infiltration. The tumor cells were positive for CD34, CD99, vimentin, BCL-2, and STAT6; EMA was negative. Both the Ki-67 and mitotic indices were low, and anaplastic nuclear features were absent. To the best of our knowledge, this is the first example of an intracranial SFT with pseudopapillary architecture. Occasional extracranial SFTs showing papillary features have been reported. The significance of this architectural pattern is unclear and may be elucidated by future studies.

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Key Words: *solitary fibrous tumor, central nervous system, intracranial, pseudopapillary*

INTRODUCTION

Solitary fibrous tumor (SFT) is a rare soft tissue neoplasm that was first described as a tumor arising from the pleura by Klemperer et al. in 1931.[1] SFTs have subsequently been found in many different locations; 40% of tumors are in subcutaneous tissues and other SFTs are found in the deep soft tissues of extremities or extra-compartmentally in the head and neck region (especially the orbit), thoracic wall, mediastinum, pericardium, retroperitoneum, and abdominal cavity. Other described locations include the meninges, spinal cord, periosteum, salivary glands, lungs, thyroid, liver, gastrointestinal tract, adrenals, urinary bladder, prostate, spermatic cord, and testes.[2] SFTs involving the central nervous system (CNS) is a rare occurrence. In 1996, Carneiro et al. reported the first cases of CNS SFT, describing seven cases of meningeal SFT that could be distinguished from fibrous meningioma based on morphologic and immunohistochemical grounds.[3] Since then, approximately two hundred fifty cases have been reported at various sites within the CNS in the English literature.[4-12]

SFTs in the CNS can affect both cranial and spinal meninges and may involve spinal nerve roots. The tumors are seen primarily in adults, and may show invasion of brain parenchyma or nerve roots as well as the skull base.[11] According to the study by Bisceglia et al., most CNS SFTs are intracranial; just over one-fifth of tumors involve the spine. In decreasing frequency, intracranial tumors involve the supratentorial compartment, infratentorial compartment, pontocerebellar angle, sellar and parasellar regions, and cranial nerves. Intraspinal tumors are mainly located in the thoracic and cervical segments.[9]

Histologically, SFTs present as a patternless proliferation of spindled cells lacking significant atypia in a collagenous stroma, with variably prominent sinusoidal (occasionally staghorn-type) vessels present. The cells express CD34, BCL2, CD99 and STAT6 antigens by immunohistochemistry.[11,13,14] Herein, we report a case of a dural-based solitary fibrous tumor that showed prominent pseudopapillary architecture, an unusual histomorphology that may result in diagnostic confusion.

CASE REPORT

Case History

This patient was a 73-year-old male with a history of hypertension, diabetes and prostatic adenocarcinoma (stage

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T1c, Gleason pattern 3+3), who presented to the Emergency Department with a six-week history of confusion and right sided weakness. At the time of presentation, the patient was unable to recognize his family members. The patient denied fevers, night sweats, or unintentional weight loss. There was no significant history of tobacco or alcohol use.

On physical exam, the patient was noted to be confused, with a right-sided facial droop and facial asymmetry, weakness, and numbness. He was able to communicate and follow commands appropriately. Increased muscle tone was noted in the right upper extremity, with decreased muscle tone in the right lower extremities. The left upper and lower extremities had normal muscle tone. The remainder of the physical examination was non-contributory. A brain magnetic resonance imaging (MRI) with gadolinium demonstrated a single 5-cm mass in the left occipital region and caused significant mass effect **(Figure 1a & 1b)**. The adjacent left cerebral hemisphere showed diffuse vasogenic edema. CT scans of the chest, abdomen and pelvis were unremarkable. The neuroimaging results were interpreted as most likely representing metastatic disease, given the known history of prostate cancer. The serum PSA, however, was only mildly elevated, and the subsequent bone scan showed no evidence of bone metastases.

The patient underwent elective surgical resection of the extra-axial tumors, utilizing bilateral craniotomy and stereotactic microsurgery. During surgery, the large mass was noted to be encapsulated, and firmly adherent to the dura and superior sagittal sinus.

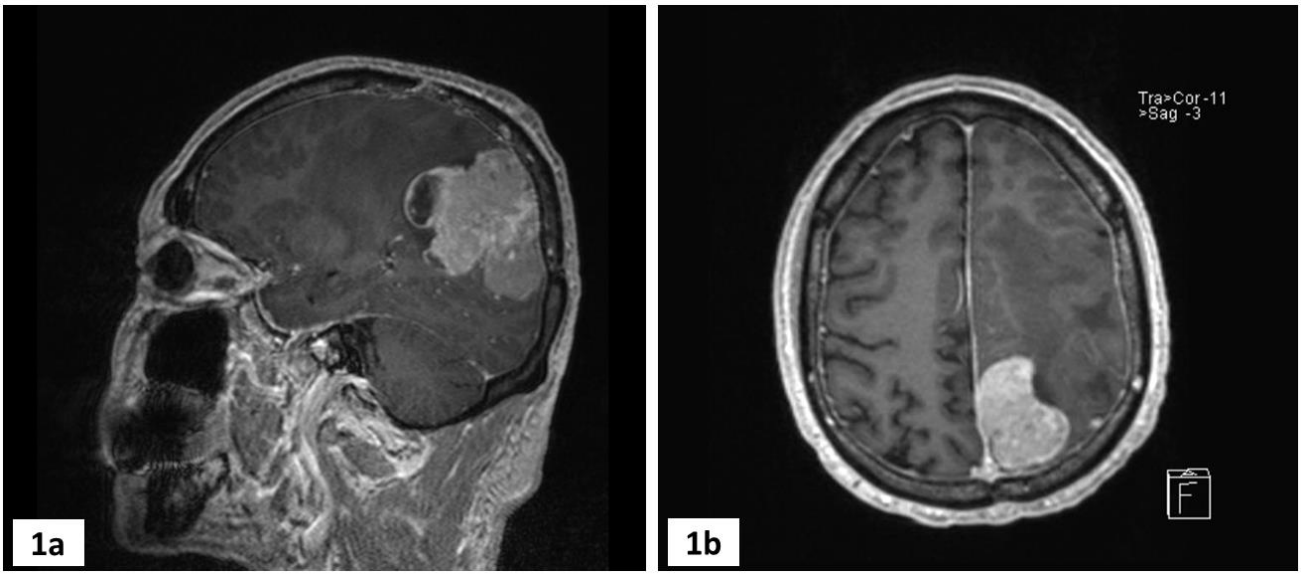

Figure 1. Presurgical MRI T1 images with Gadolinium administration showing a 5 cm mass in the left parieto-occipital region causing significant mass effect (1a sagittal, 1b axial).

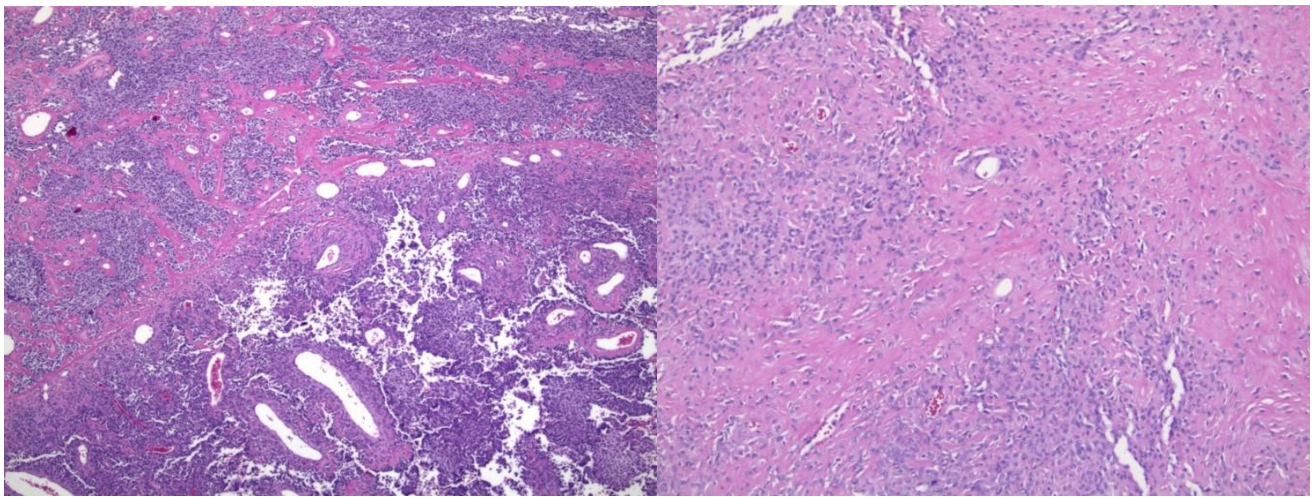

Figure 2. Areas showing classical SFT morphology and pseudopapillary architecture, HE (10X).

Figure 3. Spindled mesenchymal neoplasm with prominent intralesional collagen deposition, HE (10X).

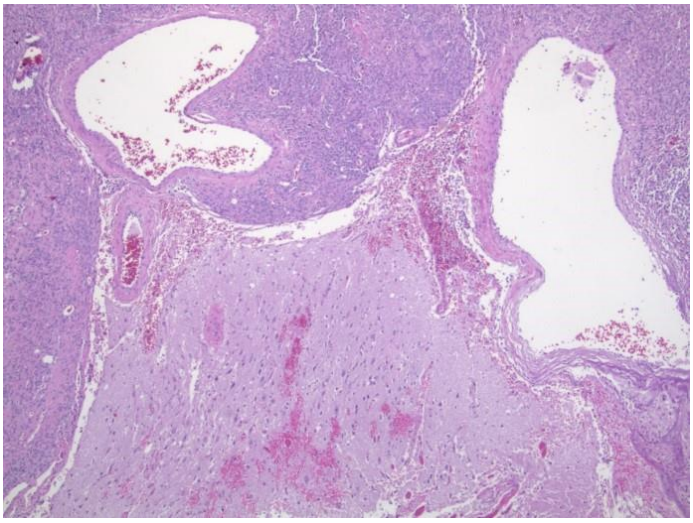

Figure 4. Entrapped brain parenchyma evidencing brain infiltration, HE (10X).

Pathologic Findings

A 5.5 x 4.8 x 3.2 cm aggregate of friable tan-red soft tissue was received in the Surgical Pathology laboratory. After formalin fixation and paraffin-embedding, routine H&E histology and subsequent special staining and immunohistochemical studies were performed. By light microscopy, the tumor comprised a monotonous low-grade pindled mesenchymal neoplasm lacking overt features of malignancy. The tumor showed variable cellularity, prominent intralesional collagen deposition, scattered ectatic sinusoidal vessels, and entrapped brain parenchyma evidencing brain infiltration **(Figure 2-4)**. Moreover, the tumor showed areas with prominent cellular discohesion, resulting in a pseudopapillary architecture **(Figure 5a & 5b)**. The smear preparation demonstrated the relatively monotonous cells and their relationship to the lesional vessels **(Figure 6)**. Typical histologic features of a meningioma (e.g. meningothelial whorls, Psammoma bodies) were not seen.

By immunohistochemistry, the tumor cells were positive for CD34, CD99, vimentin, BCL-2, and STAT6 **(Figure 7-8)**. The EMA, CAM 5.2, GFAP, CD31, SMA, PR and TTF-1 stains were negative. There was focal weak positivity for S-100. Trichrome special staining confirmed the presence of prominent collagen deposition **(Figure 9)**. The reticulin stain showed no significant intercellular reticulin deposition. Both the Ki-67 proliferation index (2.7%, eyepiece graticule) and the mitotic index were low (0-1 mitotic figures per 10 high-power field). Anaplastic nuclear features were absent, and there was no tumoral necrosis.

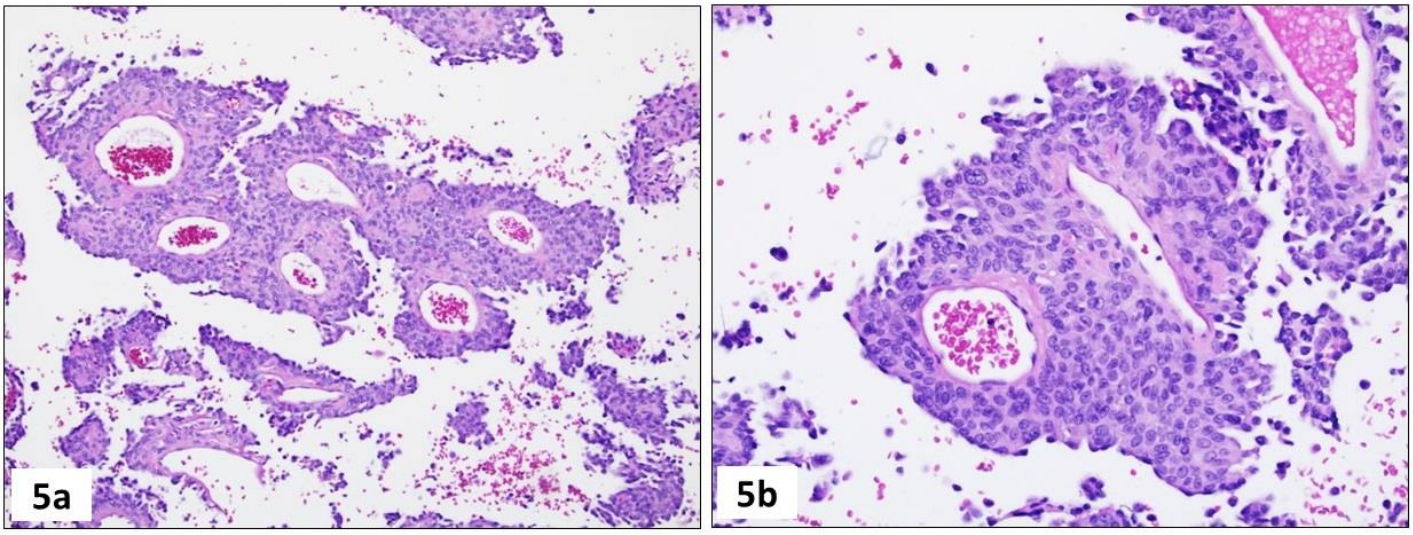

Figure 5. Portion of tumor shows prominent cellular discohesion, resulting in a pseudopapillary architecture, HE (5a10X, 5b 20X).

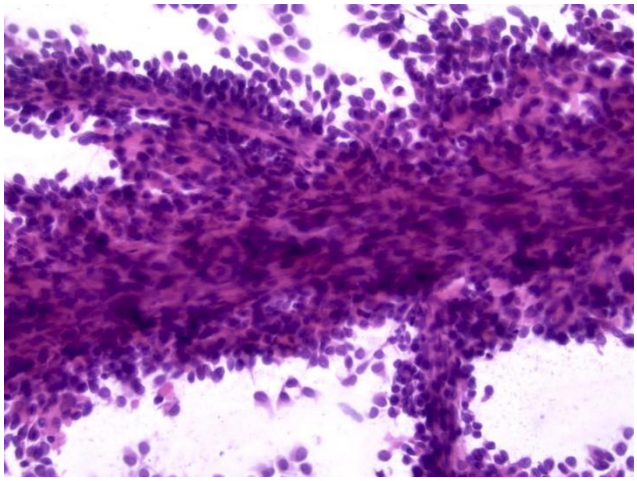

Figure 6. Smear showing vascular structure and relatively monotonous cells, HE (20X).

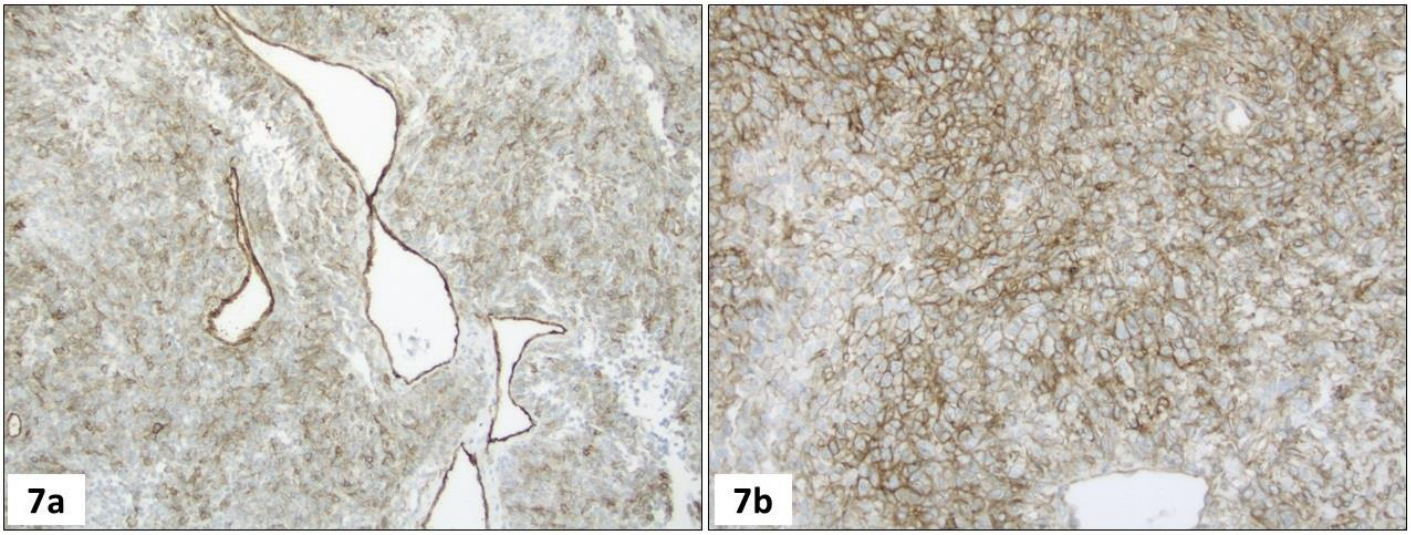

Figure 7. Immunostaining showing strong positivity for CD34 (**7a**) and CD99 (**7b**) (10X).

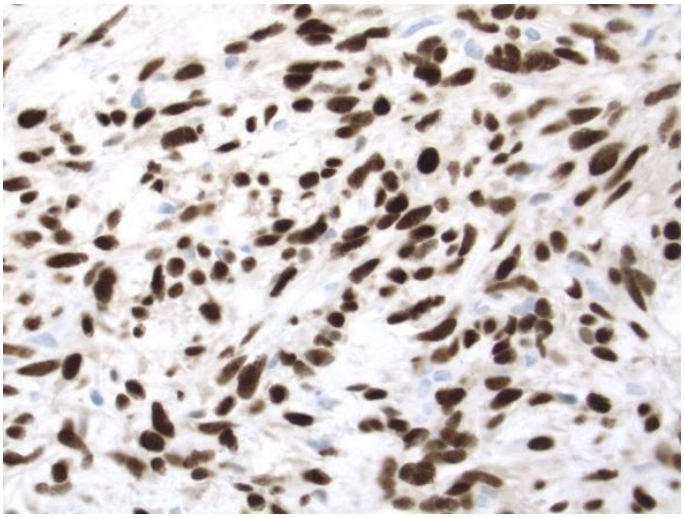

Figure 8. Diffuse and strong positivity for STAT6 in the tumor cells (40X).

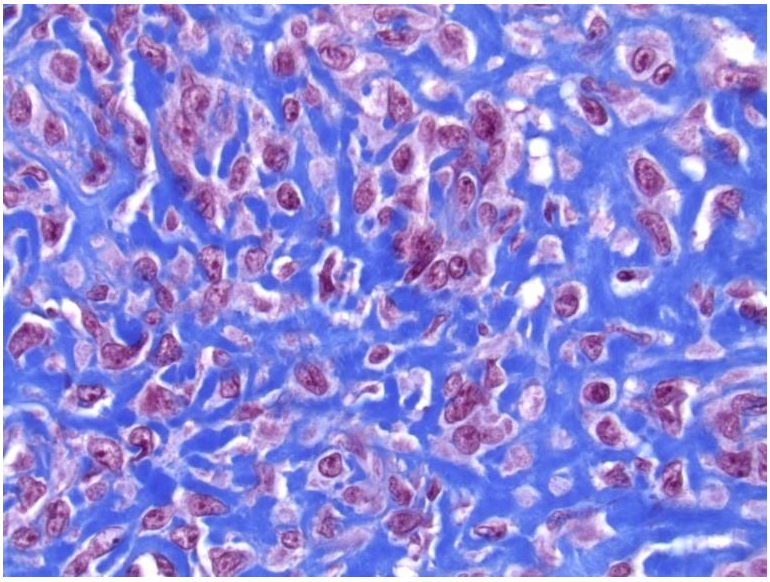

Figure 9. Trichrome staining demonstrated prominent collagen deposition (20X).

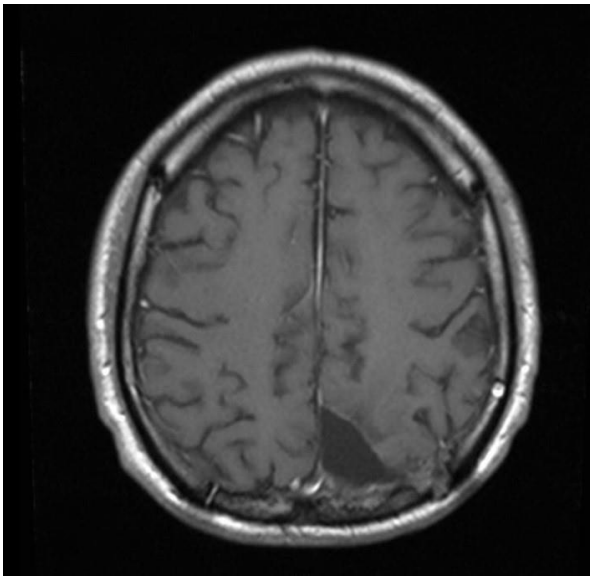

Figure 10. Post-surgical MRI imaging showing no residual tumor (T1, sagittal).

Follow-Up

The patient had an uncomplicated recovery with complete resolution of the neurologic signs and symptoms. On follow-up post-surgical MRI imaging, there was gross total resection of the tumor without residual enhancing tumor **(Figure 10)**. Given the pathologic findings, the patient underwent adjuvant radiotherapy. At one year of follow-up, there was no evidence of tumor recurrence.

DISCUSSION

We present here a case of a dural-based SFT secondarily invading the brain parenchyma, showing both classic histologic features as well as areas of prominent discohesion imparting a pseudopapillary architecture. Although occasional extracranial SFTs showing papillary features have been reported, to the best of our knowledge, this is the first report of an intracranial SFT showing pseudopapillary architecture.

Extrapleural SFTs typically occur in middle-aged adults (20 - 70 years, median age 50 years), with no sex predilection.[2] For CNS SFTs, the age of onset ranges from 11 years to 67 years with a mean age of 47.6 years. Interestingly, CNS SFTs are more common in males, with an M:F ratio of 13:9. The presenting signs and symptoms are dependent on the location of the tumor. The most common symptoms reported are headache, dizziness, unstable gait, and hearing loss.[10] Although most solitary fibrous tumors of the central nervous system are dural-based, a small subset presents as subpial, intraparenchymal, intraventricular, or as tumors involving the nerve rootlets with no dural connection.[9]

The classic histologic features of SFT include "patternless-pattern" of spindle cells, alternating hypo- and hypercellular areas, thick strands of hyalinized collagen, and hemangiopericytoma-like sinusoidal vessels. The round to spindle-shaped tumor cells have scant cytoplasm with indistinct borders and dispersed chromatin. Myxoid change, areas of fibrosis and interstitial mast cells are commonly observed.[2] Rare tumors showing myxoid, lipomatous, and epithelioid variants have also been described.[9] Malignant SFT are usually hypercellular lesions, showing at least focally moderate to marked cytological atypia, necrosis, frequent mitoses, and infiltrative margins.[2] In 2013, Tomek et al. reported extradural retiform and papillary projections within a spinal, extradural SFT.[11] In our case, the presence of areas of pseudopapillary architecture is unusual, widening the initial pathologic differential diagnosis and the subsequent panel of immunohistochemical and special stains. Despite the lack of significant cellular atypia, the presence of brain invasion is worrisome for tumor recurrence.

Pathologically, SFT/HPC most closely resembles a fibrous meningioma. In most fibrous meningiomas, classic meningothelial whorls are focally present, greatly aiding the distinction. Immunoreactivity for EMA, when present, is typical of meningioma. The presence of pseudopapillary architecture, as seen in this case, also raises consideration of other tumors with papillary morphology. A wide variety of metastatic tumor from diverse sites may show papillary morphology, including the ductal variant of prostatic adenocarcinoma. Papillary morphology may also be seen in a limited subset of glial tumors, including choroid plexus tumors, ependymomas, and neuroglial neoplasms. Historically, SFTs and hemangiopericytomas (HPCs) were considered distinct biologic entities. HPCs and SFTs are now considered tumors of the same spectrum, with HPCs representing tumors with higher cellularity, and SFTs representing lower cellularity tumors with intratumoral fibrosis. In 2014, Yoshida et al reported the utility of the

nuclear stain STAT6 as a highly sensitive and specific marker for SFTs and is of particular utility when the diagnosis is inconclusive by conventional methods.[13] In another study, Doyle et al found that STAT6 can be helpful to distinguish SFTs from histologic mimics. [14] In our case, immunohistochemical staining for STAT6 showed diffuse and strong nuclear positivity in the tumor cells.

Surgical resection is the treatment of choice for SFTs, and in most cases, complete surgical resection is curative. Postoperative and long-term follow-up are necessary to monitor for tumoral recurrence.[9] In our case, no residual enhancing tumor was found on post-surgical MRI, and at one-year follow-up, the patient was without evidence of recurrence.

In conclusion, we describe an intracranial dural-based SFT with a distinctive pseudopapillary architecture. The significance of this architectural pattern is uncertain and continued reporting of mesenchymal neoplasms with pseudopapillary architecture may reveal a unique biologic behavior. The presence of the pseudopapillary pattern may result in widening of the pathologic differential diagnosis, as other tumors with papillary features would need to be excluded by appropriate means.

CONFLICT OF INTEREST
None.

DISCLOSURE OF GRANTS OR OTHER FUNDING
None.

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