CORE DIAGNOSTICS

The macroscopic appearance of these tumors depends upon their differentiation and similarity to the corresponding extracranial soft tissue tumors. Slowly growing SS tend to be sharply circumscribed, round, or multilobular, with a smooth glistening pseudocapsule, while rapidly growing tumors are poorly circumscribed and exhibit a variegated, friable, or shaggy appearance, with hemorrhage, necrosis, and cyst formation. Meningeal sarcomas are known to invade the brain parenchyma and usually have a firm texture. Histologically SS are divided classically into two major categories: biphasic and monophasic, based on the presence of epithelial and/or spindle cell components. Monophasic fibrous type is the most common type. Lin Y.J and co-workers have reported a case of primary intracranial poorly differentiated round cell SS arranged in a pericytomatous pattern, representing the tumor progression that can occur in either monophasic or biphasic tumors.⁹

Considering the site and morphology of the tumor, the present case had a number of histopathologic differentials, which included meningioma, SFT, including its malignant form, and other sarcomas. Absence of morphologic architectural patterns of meningioma meningothelial and negative staining for EMA and PgR argues against a meningioma.¹⁰ Eman Abdelzaher in his study of 87 meningiomas showed a 100% expression of EMA, concluding the diagnostic efficiency of this marker in diagnosing meningioma.¹¹ Negativity for PgR also strengthened the results as nuclear positivity of PgR is seen in 86% to 90% cases of meningioma.¹² The characteristic appearance of SFT was investigated carefully, but in the sections submitted the spindle cells were not disposed in wavy fascicles between prominent eosinophilic bands of collagen.¹³ The "staghorn" vascular pattern separating the tumor into small lobules was the key morphologic feature in the present case. Hence sarcomas with this feature were sought for. SSs are known to exhibit hemangiopericytoma-like pattern in approximately 10% to 20% cases. Multiple sections were then studied to look for any epithelial differentiation including gland formation. Finally, immunohistochemistry was performed to reach a conclusive diagnosis. PanCK in the index case helped in delineating focal epithelial differentiation in the tumor, which was not discernable on morphology. CD34 was positive only in the intra-tumoral blood vessels and the tumor cells were negative for CD34, hence SFT was excluded. Furthermore, Foo and coworkers studied the diagnostic efficiency of TLE-1 in distinguishing synovial sarcoma from various histologic mimics. They showed that 4 of 49 SFTs showed a weak staining for TLE-1. This highlighted the higher sensitivity and specificity of TLE-1 for synovial sarcomas, particularly when the staining is moderate and strong.¹⁴ Our case had a Ki-67 proliferation index of 10%. SS with low proliferation index are well documented in literature and are known to come under low risk category if the size of the tumor is less than 5cm.¹⁵ However this is the first documented case of primary dural synovial sarcoma with a low proliferation index. In conclusion, primary meningeal SS should be kept as a differential diagnosis in dural based tumors in view of their relative chemosensitivity and future prospect of a molecular target based therapy for this neoplasm. The present case highlights the importance of an extensive morphologic and immunophenotypic analysis of mesenchymal lesions of the meninges to offer the correct and definitive diagnoses to the patient.