Commentary

The 2016 "WHO Classification of Tumours of the Central Nervous System" has defined nine molecular groups of ependymoma and their key characteristics. This is likely to have a great bearing on stratification of patients for adjuvant therapy. Certain histological variants, namely papillary, clear cell, and tanycytic ependymoma (TE), however, do not figure among these nine groups and await further characterization. TE, a Grade II ependymoma, is most commonly found in the intramedullary cervical or thoracic spinal region and was originally described by Friede and Pollak in 1978.[1] Tanycytic cells (Greek word "*tanus"* means elongated) are implicated in two types of CNS tumors, namely TE and astroblastoma. They are elongated spindly bipolar cells that generally present in the circumventricular organs, particularly in the third ventricle and central canal of the spinal cord. It is common for TE to be misdiagnosed as any nervous system tumor with spindly/fascicular appearance. The definitive diagnosis of TE requires pathological analysis, including immunohistochemical (IHC) characteristics. Electron microscopy may also contribute to diagnosis.[2]

The "fascicular" histology of TEs makes it a diagnostic challenge. Ependymal pattern is inconspicuous, rosettes are rarely seen, and pseudorosettes are subtle. Distinction from other "fascicular" appearing tumors such as schwannomas and astrocytomas (most commonly pilocytic) is mandatory.

D'Souza *et al*., in their "Letter to the Editor" published in this issue, reported a case of TE in a 12-year-old child, located in the intradural, extramedullary mass at L1–L3 region, inferior to the conus medullaris, and inside the cauda equina.[3] As mentioned by the authors, TEs if present in children are more likely to be found in the brain's intraventricular regions or the intramedullary cervico/cervicothoracic region of the spinal cord with only one case of TE in the extramedullary cauda equina. Uncommon variants at uncommon locations require being aware of such diagnostic possibilities and a workup plan. The authors have effectively used a combination of radiology and IHC panel to reach the diagnosis.

Certain clues on radiology can be looked for when facing a dilemma. Radiologically, ependymoma usually enhances on T1-weighted imaging after administration of contrast agent and is often associated with a syrinx or hematoma. Whereas ependymoma is almost always intramedullary, schwannoma is an extramedullary lesion that also avidly enhances on T1-weighted imaging with gadolinium contrast enhancement. Pilocytic astrocytomas rarely enhance.[4] Homogeneous enhancement and mass effect were also observed on the cauda equina roots in the case reported by D'Souza *et al*.[3]

Intraoperative histological diagnosis is not without pitfalls. The smears mimic a tumor with spindle cells and fibrillary background. The appearance can easily be mistaken for a schwannoma or pilocytic astrocytoma. Careful attention to the radiological findings, the surgeon's impression, and the intraoperative smear preparation details should prompt one to include this uncommon entity in the differential diagnosis of spinal neoplasms.[5]

Pathologists should vary not to offer a diagnosis based only on hematoxylin and eosin appearance, more so, if radiology and surgical findings are not

supportive. A relevant IHC panel should always be applied, as has been illustrated by D'Souza *et al*.[3] Glial fibrillary acidic protein will effectively delineate glial neoplasms from schwannomas. The latter will demonstrate S100 immunopositivity. Among the glial neoplasms, ependymomas show epithelial membrane antigen immunopositivity as dot-like perinuclear or ring-like cytoplasmic structures. Differentiation from schwannomas may be challenging since ependymomas can also express S100 positivity. It is advisable to expand the IHC panel in such cases. SOX 10 and Olig2 have been suggested as negative markers for the diagnosis of ependymomas[6] whereas SOX10 will serve as a more sensitive and specific positive marker for the diagnosis of schwannian tumors.[7] In more recent studies too, high levels of SOX10 mRNA and protein in pilocytic astrocytomas but not ependymomas.[8]

Ultrastructurally, ependymomas are known to retain the characteristic cilia with a 9 + 2 microtubular pattern, blepharoplasts, and microvilli located at the luminal surface. Cases where genetic profile of spinal TEs has been looked into, association with neurofibromatosis type 2 has been seen. This is similar to other spinal ependymomas. No specific genetic abnormality has been reported in TEs.

The letter by D'Souza *et al*.[3] highlights a less encountered variant of ependymoma and the need for a comprehensive workup even when a very familiar "fascicular" pattern is seen under the microscope.

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