Commentary

Tanycytic ependymoma (TE) originates from the tanycyte (derived from the Greek word “tanus” means “elongated”), a bipolar cell with long processes within the neutrophil, bridging the ependymal lining with the capillary wall; given the peculiar intertwined position, it is thought to function in establishing communication between the cerebrospinal fluid, the brain parenchyma, and the vasculature.[1] TE is a rare form of ependymoma.[2] According to Choi et al.,[3] 34 cases have been reported up to 2016 since the first case was described by Friede and Pollak[4] as a distinct pathological variant of ependymoma. However, Tao et al.[5] reported forty cases of histologically proven spinal TE (from among 4000 spinal tumors operated in a Beijing Hospital); these patients had a mean age of 40 years at diagnosis (range: 22–69 years) with no sex preference. The age of the cases reviewed by Choi et al.[3] ranged from 40 to 59 years, with a male-to-female ratio of 1:3. In view of the absence of a single pediatric case in these two series of TE, the histologically proven TE case in a 12-year-old boy, as reported by D’Souza et al.,[6] gains significance as a very rare presentation of this entity. According to Kleihues and Cavenee,[7] TEs are almost always intramedullary in location; of
the 25 cases reported, 17 involved the spinal cord and 2 were extramedullary. Krisht and Schmidt[2] reviewed 18 TE cases, which included 15 intramedullary, 2 extramedullary, and one of unknown location. Till date, three cases of TE adjacent to the filum terminale have been reported in the literature.[8‑10] In the present TE case reported by D’Souza et al.,[6] the magnetic resonance imaging finding was of an intradural, extramedullary mass at L1‑L3 region, inferior to the conus medullaris and inside the cauda equina, which at laminectomy, was located within the subarachnoid space and attached to the filum terminale and a few nerve roots.

Under the World Health Organization (WHO) classification, the histologic subtypes of ependymoma are of three grades based on the degree of malignancy.[11] Grade II lesions include classic, cellular, papillary, clear cell, and tanycytic subtypes, grouped together for their lack of anaplastic features and similar biologic behavior.[12] According to Friede and Pollak,[4] TE is characterized by the presence of elongated bipolar or unipolar spindle cells possessing the nuclear characteristics of ependymal cells with conspicuous absence of the characteristic perivascular pseudorosettes and ependymal rosettes. Thus, TE is a WHO Grade II tumor with histological characteristics distinct from the typical features of commonly encountered ependymomas.[2]

TE is a challenging diagnosis under light microscope due to its similarity with schwannoma (spindle cell nature) and pilocytic astrocytoma (long cytoplasmic processes). Many authors have described TE as a lesion in which the classic ependymal rosettes and pseudo‑rosettes are replaced by more fibrillary cells, giving rise to resemblance with schwannoma and astrocytoma.[1,2,9] The more oval character of the nuclei and the characteristic “salt and pepper speckled” appearance of chromatin in TE distinguish it from pilocytic astrocytoma which is associated with Rosenthal fibers and eosinophilic granular bodies.[1] In cases of difficulty, immunohistochemistry is of help. TE is glial fibrillary acidic protein (GFAP+) and vimentin+ and rarely stains for S100, but schwannoma is S100+ and GFAP− and pilocytic astrocytoma is GFAP+ and vimentin−.[2] According to Langford and Barré[1] and D’Souza et al.,[6] the spindle cells in TE are immunopositive for GFAP and give rise to variable dot‑like intracytoplasmic immunoreactivity for EMA, whereas schwannoma may only focally express GFAP and are EMA negative. In one of the TE cases reported by Choi et al.,[3] the tumor was composed of spindle cells with a palisading pattern that resembles Verocay bodies of schwannoma and clear cells in some areas; prominent blood vessels, macrophages, and a few giant cells were also noted. However, the tumor was strongly and uniformly positive for GFAP, supporting the diagnosis of TE. Tao et al.,[3] in their immunohistochemical studies of TE cases, observed positive reaction for GFAP in all the 12 (100%) cases of TE tested; EMA was positive in five of ten (50%) cases; S100 was positive in all the five (100%) cases; oligo 2 was positive in one of five (20%) cases; vimentin and nestin were positive in all the three (100%) cases tested, and myelin basic protein was negative in all the three cases tested. Furthermore, Ki‑67 labeling index was performed in nine cases, with negative reaction in three cases, positivity in 1%‑5% cells in four cases, and 5%‑10% positivity in three cases.[3] In ultrastuctural studies, the spindle cells of TE are arranged in bundles with scant extracellular matrix,[5] and the tumor cells have intracytoplasmic intermediate filaments, prominent intercellular junctions, numerous slender surface microvilli, and microvilli‑lined lumina.[1,5]

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