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.[3] 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 Barrė[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*.,[5] 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.[5] 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]

Dilip Kumar Das[1,2]

[1]Department of Pathology, Faculty of Medicine, Kuwait University, Safat and Cytology Unit, [2]Department of Laboratory Medicine, Mubarak Al-Kabeer Hospital, Jabriya, Kuwait

Address for correspondence: Dr. Dilip Kumar Das, Department of Pathology, Faculty of Medicine, Kuwait University, P.O. Box 24923, Safat 13110, Kuwait.
E-mail: dilip76@hotmail.com

References

1. Langford LA, Barré GM. Ependymoma. Ultrastruct Pathol 1997;21:135-42.
2. Krisht KM, Schmidt MH. Tanycytic ependymoma: A challenging histological diagnosis. Case Rep Neurol Med 2013;2013:170791.
3. Choi JY, Kim KN, Shin DA, Yoon H, Yoon DH, Yi S. Spinal tanycytic ependymoma in four Korean patients: Case series. Nerve 2018;4:92-6.
4. Friede RL, Pollak A. The cytogenetic basis for classifying ependymomas. J Neuropathol Exp Neurol 1978;37:103-18.
5. Tao X, Hou Z, Hao S, Zhang Q, Wu Z, Zhang J, *et al.* The clinical features and surgical outcomes of spinal cord tanycytic ependymomas: A report of 40 cases. World Neurosurg 2017;106:60-73.
6. D'Souza P, Martin WE, Bodhireddy S, Belirgen M. Extramedullary tanycytic ependymoma in a 12-year-old boy. J Neurosci Rural Pract 2019;10:384-6.
7. Kleihues P, Cavenee W. Pathology and genetics of tumors of the nervous system. In: World Health Organization Classification of Tumors. Lyon, France: IARC Press; 2000.
8. Mohindra S, Bal A, Singla N. Pediatric tanycytic ependymoma of the cauda equina: Case report and review of the literature. J Child Neurol 2008;23:451-4.
9. Shintaku M, Sakamoto T. Tanycytic ependymoma of the filum

terminale with pleomorphic giant cells. Brain Tumor Pathol 2009;26:79-82.
10. Radhakrishnan N, Nair NS, Hingwala DR, Kapilamoorthy TR, Radhakrishnan VV. Tanycytic ependymoma of filum terminale: A case report. Clin Neurol Neurosurg 2012; 114:169– 171.
11. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, Jouvet A, *et al.* The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 2007;114:97-109.
12. Perry A, Prayson RA. Glial and glioneuronal tumors. In: Prayson RA, editor. Neuropathology. 2nd ed. Philadelphia: Elsevier Inc.; 2012. p. 461-512.

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