Diffuse leptomeningeal glioneuronal tumor

This tumor was described as a distinct entity for the first time in 2010 [1)].

Schniederjan et al. present 9 pediatric cases of such diffuse leptomeningeal neuroepithelial tumors (DLNT), 8 with assessment of 2 common genetic alterations seen in oligodendrogliomas, 1p and 19q chromosomal deletions and isocitrate dehydrogenase-1 (IDH1) R132H mutations. Four patients were male and 5 female, with a mean age at presentation of 4 years (range, 2 to 7 y). All presented with signs of increased intracranial pressure and diffuse contrast enhancement of the leptomeninges by magnetic resonance imaging. Three had a cervical or upper thoracic spinal cord tumor, and another had a small cerebellar lesion. Leptomeningeal biopsies showed a thickened and fibrotic arachnoid infiltrated by monotonous cells with round nuclei and prominent perinuclear clearing. All cases were strongly immunoreactive for S100 protein, and most showed faint granular synaptophysin reactivity. Six of 8 cases showed deletions of chromosome arm 1p by fluorescence in situ hybridization, 2 of which also had loss of 19q. None of the lesions reacted with IDH1-R132H antibodies. Although the clinicopathologic features show overlap of these DLNT lesions with oligodendroglioma and extraventricular neurocytoma, they do not exactly match either one, suggesting that DLNTs are a distinct tumor entity [2)].

Diffuse leptomeningeal glioneuronal tumor (DL-GNT) is a rare brain tumor that presents as a plaque-like subarachnoid tumor, commonly involving the basal cisterns and interhemispheric fissure of children but lacking intraparenchymal tumor.

Histology

Histologically, the tumors are composed of sheets of monotonous rounded cells.

Cho et al. report three cases of DL-GNTs, focusing on clinicopathologic features. Two patients were adult male, but one patient was child. The patients presented with seizures (n = 1) or headaches (n = 2). In all patients, radiography revealed characteristic leptomeningeal thickening and enhancement with minor superficial parenchymal lesions. All three cases were diffusely positive for both GFAP and synaptophysin, and scattered positive for OLIG2 and NeuN, but negative for IDH-1 (H09). Electron microscopic examination showed astrocytic and neuronal differentiation. The patient with the anaplastic tumor died due to aggressive progression of the tumor, but the remaining two patients were stable without tumor recurrence for 23 and 37 months. Thus, these findings suggest that DL-GNT can occur in both children and adult and both supra- and infra-tentorial leptomeninges. It has unique radiological and histopathological features and biological behavior. Further clinicopathological data with molecular genetic study are required for establishing DL-GNT as a unique entity [3)].

Differential diagnosis

The differential is essentially that of leptomeningeal enhancement, with the florid nature of enhancement particularly reminiscent of tuberculous leptomeningitis. Other causes to particularly consider include:

tuberculous leptomeningitis

leptomeningeal carcinomatosis

leptomeningeal seeding from (another) primary CNS tumour

1)
Gardiman MP, Fassan M, Orvieto E, D'Avella D, Denaro L, Calderone M, Severino M, Scarsello G, Viscardi E, Perilongo G. Diffuse leptomeningeal glioneuronal tumors: a new entity? Brain Pathol. 2010 Mar;20(2):361-6. doi: 10.1111/j.1750-3639.2009.00285.x. Epub 2009 May 22. PubMed PMID: 19486008.
2)
Schniederjan MJ, Alghamdi S, Castellano-Sanchez A, Mazewski C, Brahma B, Brat DJ, Brathwaite CD, Janss AJ. Diffuse leptomeningeal neuroepithelial tumor: 9 pediatric cases with chromosome 1p/19q deletion status and IDH1 (R132H) immunohistochemistry. Am J Surg Pathol. 2013 May;37(5):763-71. doi: 10.1097/PAS.0b013e31827bf4cc. PubMed PMID: 23588371.
3)
Cho HJ, Myung JK, Kim H, Park CK, Kim SK, Chung CK, Choi SH, Park SH. Primary diffuse leptomeningeal glioneuronal tumors. Brain Tumor Pathol. 2015 Jan;32(1):49-55. doi: 10.1007/s10014-014-0187-z. Epub 2014 Apr 26. Review. PubMed PMID: 24770606.