Pleomorphic Xanthoastrocytoma differential diagnosis

1. imaging: meningioma is also superficial with dural tail, may also resemble low-grade fibrillary astrocytoma

2. pathology: may be confused with anaplastic astrocytoma

Main differential diagnosis is that of other cortical tumours, with helpful distinguishing features including:

Ganglioglioma can look very similar contrast enhancement often less prominent calcification in ~50% of cases no dural tail sign.

The analysis of the methylation profile suggested the diagnosis of an anaplastic pleomorphic xanthoastrocytoma and as a differential diagnosis an anaplastic ganglioglioma [1)].

Dysembryoplastic neuroepithelial tumors (DNET) contrast enhancement uncommon 'bubbly appearance' common

Oligodendroglioma calcifications common

Desmoplastic infantile ganglioglioma young children dural involvement prominent large often multiple lesions

Cystic meningioma

Fibrillary astrocytoma

Anaplastic astrocytoma.

Glioblastoma

Epithelioid glioblastoma (eGlioblastoma) and pleomorphic xanthoastrocytoma (PXA) with anaplastically transformed foci (ePXA) show overlapping features. Eleven eGlioblastomas and 5 ePXAs were reviewed and studied immunohistochemically. Fluorescence in situ hybridization for EGFR amplification, PTEN deletion and ODZ3 deletion was also performed, with Ilumina 450 methylome analysis obtained in five cases. The average age for eGlioblastoma was 30.9 (range 2-79) years, including five pediatric cases and a M : F ratio of 4.5. The ePXA patients had a M : F ratio of 4 and averaged 21.2 (range 10-38) years in age, including two pediatric cases. Six eGlioblastomas and two ePXAs recurred (median recurrence interval of 12 and 3.3 months, respectively). All tumors were composed of solid sheets of loosely cohesive, “melanoma-like” cells with only limited infiltration. ePXAs showed lower grade foci with classic features of PXA. Both tumor types showed focal expression of epithelial and glial markers, retained INI1 and BRG1 expression, occasional CD34 positivity, and lack of mutant IDH1 (R132H) immunoreactivity. BRAF V600E mutation was present in four eGlioblastomas and four ePXAs. ODZ3 deletion was detected in seven eGlioblastomas and two ePXAs. EGFR amplification was absent. Methylome analysis showed that one ePXA and one eGlioblastoma clustered with PXAs, one eGlioblastoma clustered with low-grade gliomas, and two

eGlioblastomas clustered with pediatric-type glioblastomas. Common histologic, immunohistochemical, molecular and clinical features found in eGlioblastoma and ePXA suggest that they are closely related or the same entity. If the latter is true, the nomenclature and WHO grading remains to be resolved [2].

Addition to the histological and immunohistochemical evaluation, investigation of MGMT promoter methylation and in particular BRAF V600E mutations represent reliable additional tools to sustain differentiation of gcGlioblastoma from PXA on a molecular basis. Based on these data specific BRAF kinase inhibitors could represent a promising agent in the therapy of PXA and their use should be emphasized [3].

1)
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2)
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