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Review

Malignant potential of pleomorphic xanthoastrocytoma

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ABSTRACT

Pleomorphic xanthoastrocytoma (PXA) is a low-grade astrocytic tumour that occasionally progresses to a higher grade. We have extensively reviewed the literature on the potential for malignant transformation of PXA. An illustrative case of a PXA transforming to glioblastoma multiforme is presented.

1. Introduction

Pleomorphic xanthoastrocytoma (PXA) is a rare astrocytic neoplasm that commonly affects children and young adults, usually presenting with seizures.[1] PXA is typically superficial, involving the cortex and meninges. Most occur supratentorially, many in the temporal lobe.[1]

PXA has a characteristic histopathological appearance. Cells are pleomorphic and exhibit cytoplasmic xanthic change. Most express glial fibrillary acidic protein (GFAP). Tumour cells may be surrounded by a reticulin network and eosinophilic granular bodies.[1]

PXA is classified as a World Health Organization (WHO) grade II astrocytoma, and is conventionally regarded as a benign neoplasm that carries a generally favourable prognosis after gross total resection (GTR).[1] However, a subset of PXA denoted "PXA with anaplastic features" (APXA) that corresponds histologically with WHO grade III due to its more malignant histology and behaviour, has also been identified.[2] Further, it is recognised that a proportion of PXA will recur, frequently having accumulated anaplastic features that increase their malignancy to grade III or even IV.[1] The growing literature around PXA increasingly suggests that primary APXA and PXA that has undergone anaplastic or malignant transformation carries a significantly less favourable prognosis.

Here we describe an unusual case of PXA that underwent transformation into glioblastoma multiforme (GBM) within 6 months of initial resection.

We then review the recent PXA literature with particular focus on primary APXA as well as the small number of reported cases of transformation of simple PXA into APXA or high grade glioma on recurrence.

2. Case report

A previously well 50-year-old man presented after two episodes of generalised tonic-clonic seizure. MRI showed a heterogenous 14 × 12 × 13 mm mass in the anterior pole of the left temporal lobe, with peripheral contrast enhancement and perifocal oedema (see Fig. 1). High-grade glioma was suspected.

Gross macroscopic excision was achieved, and histological examination revealed a densely hypercellular glial tumour. Tumour cells exhibited marked pleomorphism with variably shaped, large vesicular nuclei. Occasional mitotic figures were identified. There was no necrosis or endothelial cell hyperplasia. Moderate numbers of tumour cells had foamy, xanthomatous cytoplasm, and tumour cells, including those with xanthomatous features, were immunoreactive for GFAP and nestin. Prominent perivascular cuffing by small lymphocytes was present, and scattered multinucleated tumour giant cells were also noted. The topoisomerase labelling index was approximately 5%. The diagnosis was PXA without anaplastic features (see Fig. 2).

Accordingly, the decision was made not to commence adjuvant therapy, subject to regular outpatient radiological monitoring. The patient made a good post-operative recovery and MRI four months later demonstrated no residual tumour (see Fig. 3a).

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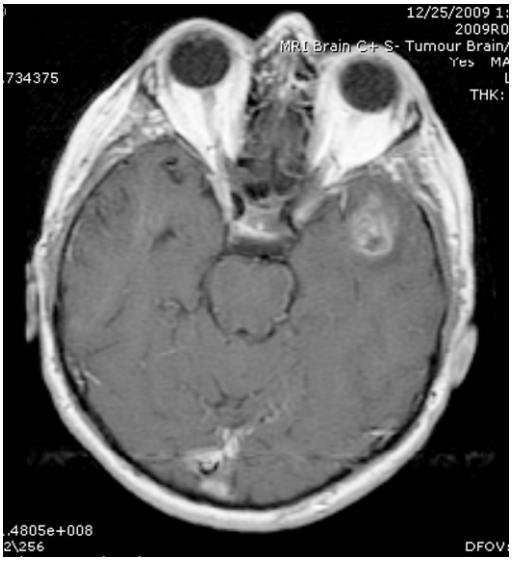

Fig. 1. Axial T1-weighted contrast-enhanced MRI at diagnosis demonstrating a ring-enhancing lesion in the left anterior temporal pole.

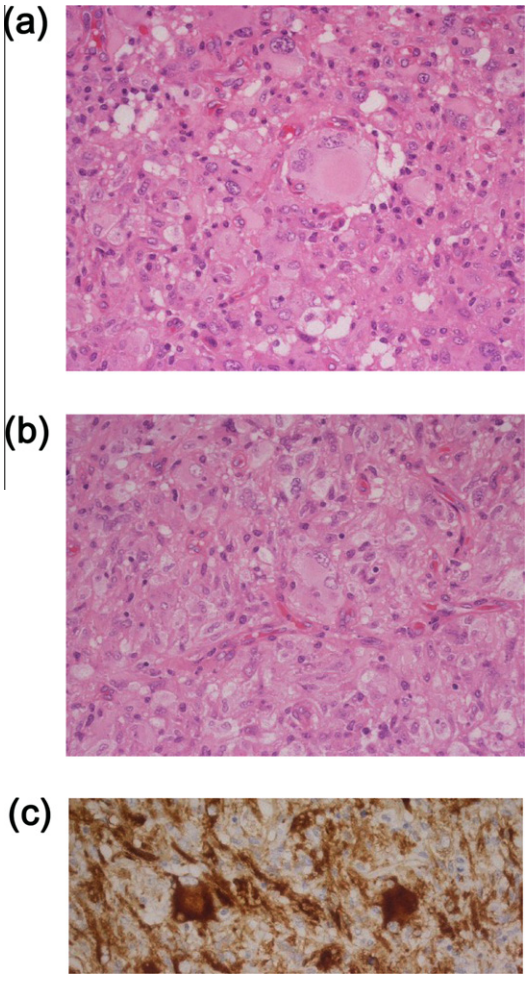

Fig. 2. Histopathological examination of biopsy specimen at diagnosis demonstrating: (a) xanthomatous tumour cells (haematoxylin and eosin [H&E] stain, ×400 magnification); (b) pleomorphic tumour cells (H&E staining, ×200 magnification); and (c) immunostaining for glial fibrillary acidic protein (GFAP) with reactivity in the cytoplasm of the xathomatous cells (×400 magnification). [This figure is available in colour at www.sciencedirect.com.]

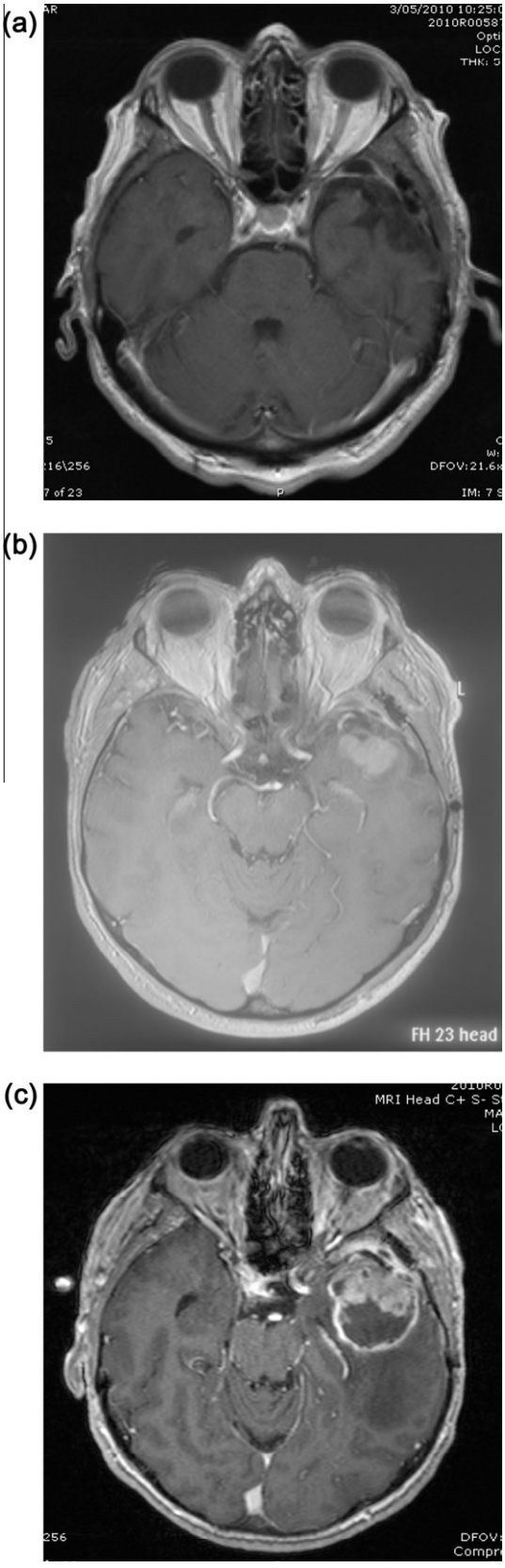

Fig. 3. Axial T1-weighted contrast-enhanced MRI: (a) performed four months after the initial resection and showing no residual tumour; (b) two months later, the MRI demonstrated a 27 × 23 mm lobulated, mixed solid and cystic contrast-enhancing recurrent tumour at the primary site; (c) one month later the tumour had undergone 6-fold enlargement and developed areas of necrosis and significant vasogenic oedema, requiring urgent craniotomy and debulking.