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Primary anaplastic pleomorphic xanthoastrocytoma in adults. Case report and review of literature

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ABSTRACT

BACKGROUND: Pleomorphic xanthoastrocytoma (PXA) classified as a low Grade (WHO II) astrocytic neoplasm. It is known for its relatively favorable prognosis. It most commonly occurs in young adults. Malignant progression in PXA has been frequently reported since its first description in 1979; however, the presentation of a primary anaplastic PXA tumor with an aggressive clinical course in adults is rare especially in the later age group.

CASE DESCRIPTION: We present a case of primary anaplastic PXA in a 53 year old male that manifested with an early recurrence pattern at 9 weeks. Treatment performed was surgical excision and external beam radiotherapy. The aforementioned tumor followed an aggressive clinical course. Tumor cells exhibited the characteristic expression of GFAP (Glial fibrillary acidic protein), higher proliferative index (8–10%) on Ki-67 staining along with the presence of increased mitoses (>5/10hpf). A review of previously reported primary anaplastic pleomorphic xanthoastrocytoma cases in adults with histological features was also done.

CONCLUSION: Our review of all reported cases of APXA in adults concludes that the clinical behavior of this tumor varies considerably from its benign variant. Early disease recurrence in anaplastic pleomorphic xanthoastrocytomas is associated with fatal outcomes. As per our review of literature it is seen that anaplastic variant of PXA shows histological characteristics as well as clinical course comparable with Grade III astrocytomas.

We recommend further evaluation of PXA with anaplastic features regarding their genetic characteristics to understand the origin as well as behavior of this tumor.

1. Introduction

Pleomorphic xanthoastrocytoma (PXA) is an astrocytic neoplasm with a relatively favorable prognosis [8]. According to WHO classification for astrocytic neoplasms, it has been classified histologically as a grade II (benign) neoplasm [6]. The first case was reported in 1979 [11]. It is often superficially located in the cerebral cortex with leptomennigeal involvement. Morphologically it shows a pleomorphic histological appearance that includes lipidized, GFAP-expressing tumor cells with cytoplasmic xanthic change surrounded by a reticulin network [9]. It has been frequently seen that tumors initially diagnosed as PXA have later shown malignant progression to high grade astrocytomas (grade III or IV). In these cases the initial histological findings corresponded to a grade II neoplasm; however over the recurrences it was found to be malignant [3]. To the best of our knowledge, only a few cases have been reported in the literature, which demonstrate a PXA tumor presenting with anaplastic features at initial presentation. These cases have been reported mostly in children and young adults ranging between 7–25 years [16]. Here we present a case of a primary anaplastic PXA tumor in the later age group with an unusual early recurrence pattern. We then review the literature of previously reported cases of primary anaplastic PXA tumors in adults.

2. Case description

55 years old male presented with a history of sudden onset headaches and two episodes of generalized tonic clonic seizures in 3 months. Neurological examination did not show any focal motor or sensory deficits. MRI brain showed a 2.2 × 1.3 × 1.1 cm nodular

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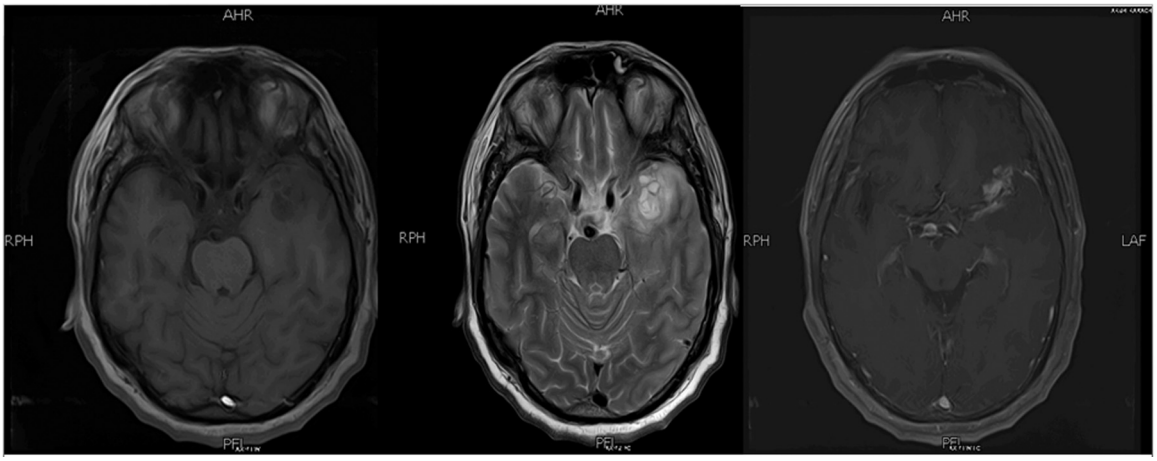

Fig. 1. (A) MRI Brain T1 weighted showing hypointense nodular thickening in the left temporal lobe and para sylvian fissure. (B) T2 weighted image showing hyperintense lesion in the left temporal lobe. (C) T1 contrast image showing patchy enhancement in the left temporal lobe.

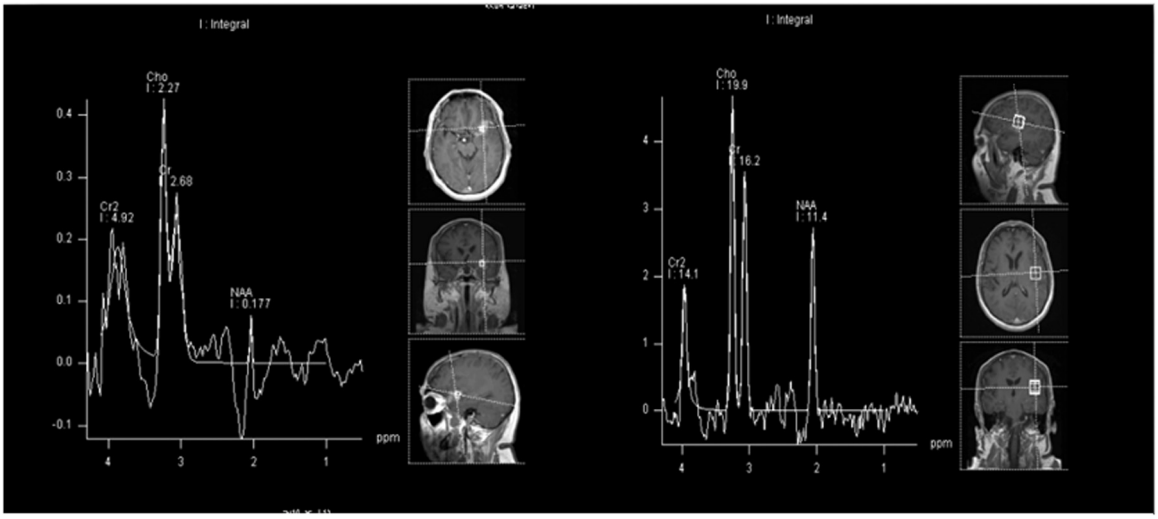

Fig. 2. MR spectroscopy showing high Choline/Creatine and high Choline/NAA ratios in the enhancing areas and persistent lactate peak in all enhancing areas favoring neoplastic lesion.

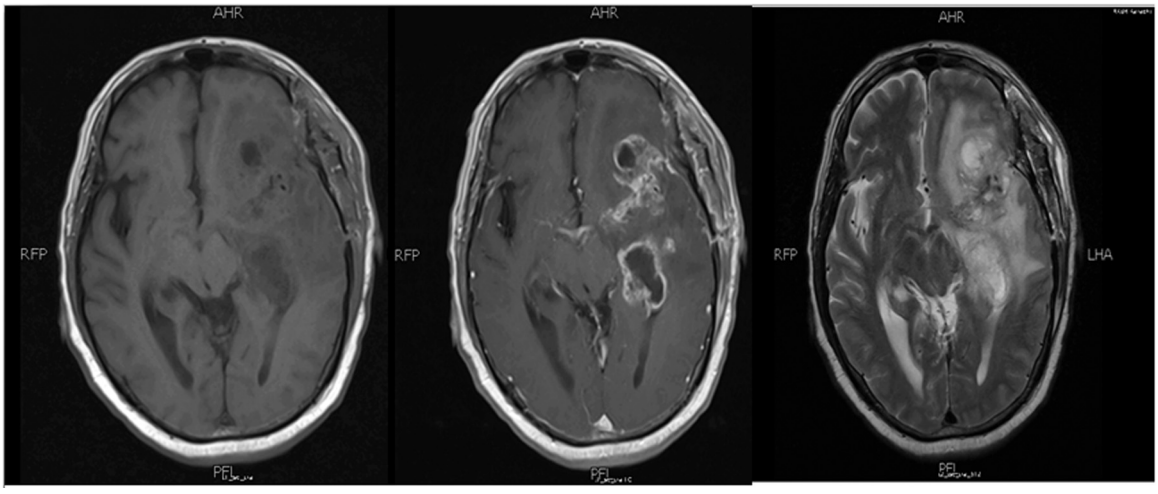

Fig. 3. MRI Brain showing significant overall increase in the size of tumor involving the left frontal, temporal and parietal lobes with perilesional edema and post surgical changes in T1, T1 post contrast and T2 weighted images respectively.