# Pleomorphic Xanthoastrocytoma: An Atypical Astrocytoma

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## Abstract

Pleomorphic xanthoastrocytoma (PXA) is rare primary neoplasm of brain. Despite its pleomorphic appearance, it has a relatively good prognosis. We report a case of biopsy proven pleomorphic xanthoastrocytoma in a young male who presented with visual and sensory symptoms, classical neuroimaging findings and showed remarkable recovery, post surgery. We have also reviewed recent literature focusing on neuroimaging, histopathology and prognostic markers of the tumour.

**Keywords:** Pleomorphic xanthoastrocytoma, Mural nodule, Supratentorial, GFAP.

## Introduction

Astrocytomas are the most common glial tumours of the nervous system amongst our paediatric population. Most childhood tumours arise infratentorially (astrocytomas, ependymomas and medulloblastomas) or in the midline (germ cell tumours or craniopharyngiomas). Supratentorial astrocytomas are more likely to be high grade compared with their infratentorial counterpart.[1] Pleomorphic xanthoastrocytoma (PXA) is a rare neoplasm that accounts for 1% of all astrocytic tumours. The correct diagnosis of this uncommon supratentorial, cortical tumour is crucial as in contrast to other pleomorphic and supratentorial neoplasms, these tumours usually have a favourable prognosis.[2]

## Case Report:

A 13-year-old boy presented in our outpatient clinic with mild diffuse headaches and numbness of right half of face for last 2 months. He also noticed difficulty in chasing a ball while playing cricket since in the same period so he had to localize it with its sound. He had bilateral decreased vision for one year corrected partially with refractive lenses. A student of class 7, he had normal cognition and behaviour and was good in studies. Past history and family history was insignificant. On examination he was a young male of average height and built, alert and co-operative. Neurological examination revealed higher normal mental functions, decreased visual acuity in both eyes, left homonymous hemianopsia, bilateral pale discs with normal pupillary reflexes and extraocular movements. Apart from a small area of hypoaesthesia around right angle of mouth, rest of motor, cerebellar and sensory exam was normal. Routine laboratories were unremarkable. On MRI brain (plain), there was a large(size) well defined cystic mass in right temporo-parieto-occipital region with mild vasogenic oedema and mass effects causing midline shift, compression of ipsilateral lateral and third ventricle and cerebral peduncle (A1-2). Post-contrast studies showed rim like enhancement of the cystic mass, with enhancing mural nodule (A3-4). Neurosurgery consult was obtained and complete excision of tumour was performed. Histopathology revealed fascicular arrangement of spindle shape cells admixed with tumour giant cells. The tumour giant cells exhibited marked nuclear enlargement and pleomorphism. The cytoplasm had a fine granular

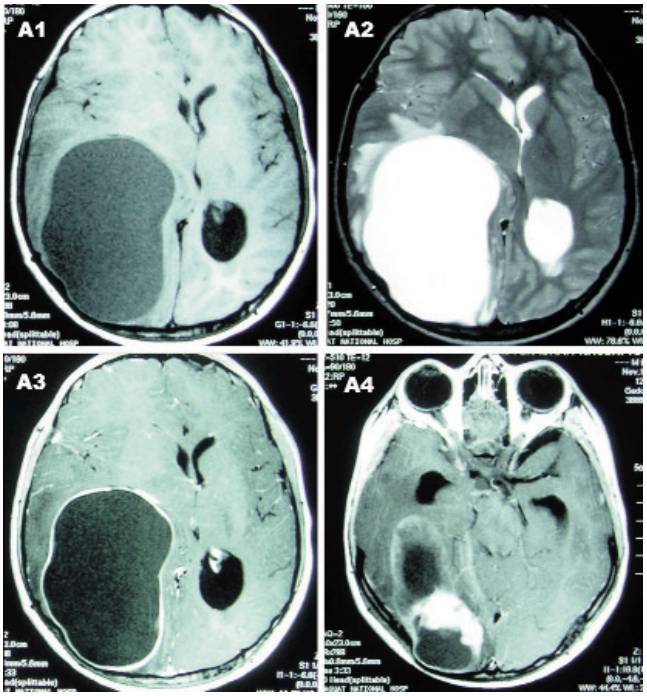


Figure A: (A1, A2) T1 and T2 axial MRI sequence showed a huge well defined cystic mass in right temporo-parieto-occipital region, with mild vasogenic oedema, but severe mass effects. (A3, A4) Post contrast study demonstrated enhancement of the cyst wall with enhancing mural nodule attached to meningeal covering.

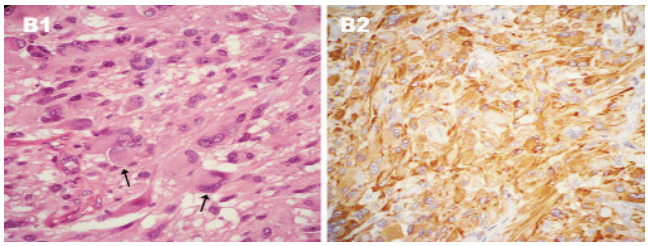


Figure B: (B1) H & E stain × 400 revealed tumour giant cells (arrows). (B2) Diffuse GFAP immune labeling of neoplastic cells.

appearance. No zones of necrosis were observed and mitosis was rare (B1-2). Immunohistochemistry revealed positive results for GFAP and CD 68 (cluster of differentiation 68) and negative for vimentin.

He showed remarkable improvement with resolution of left sided visual defect and a normal visual acuity and peri-oral sensations on 3rd postoperative day. On his last follow-up, 10 months after initial presentation he had had an uneventful course.

## Discussion

In 1993, the World Health Organization recognized PXA as a distinct tumour in classification of grade II tumours.[2]

It belongs to the well-circumscribed variety of astrocytic glial neoplasms and is considered to have a neuroectodermal origin as the cytoplasm of tumour cells shows the presence of both glial fibrillary acid protein (GFAP) and S-100 protein. The peak age of onset is 20 years and 90% of the reported cases are below 30 years of age. It affects males and females in equal numbers.[3]

The typical clinical presentation is protracted seizures and headache.[3] Our patient had an atypical presentation, with reduced visual acuity, peri-oral hypoaesthesia and no history of seizures.

The vast majority of PXA are supratentorial location. The most common location of the mass is the temporal lobe; next, in order of frequency, are the parietal, occipital, and frontal lobes.[4] Location outside the supratentorial compartment has been reported in cerebellum and spinal cord.[5] They frequently have cystic appearance and are often superficially-located, cortical tumours without significant mass effect. These tumours involve the leptomeninges but not the dura mater. They appear as a hypodense or intense mass with distinct borders on CT scan or T1 weighted and hyperintense on T2 weighted MR images. An eccentric mural nodule attached to the meninges is seen. Usually, the cyst wall does not enhance in contrast to intense enhancement of the peripheral nodule.[3,4] Interestingly, in the reported patient, there was intense enhancement of cyst wall on post-contrast studies.

Grossly, a yellowish cyst with a mural nodule is common, with the solid portion contacting the leptomeninges.[4] The characteristic histopathological features of PXA include: large pleomorphic cells, prominent eosinophilic granular bodies, lipidized astrocytes, focal perivascular lymphocytes, abundant reticulin network, absent or scant mitoses, and absent necrosis.[2]

Giannini et al reported immunostained, 40 cases of PXA. Conventional PXAs demonstrated immunoreactivity for glial fibrillary acidic protein (100% of cases), S-100 protein (100%), class III [beta]-tubulin (73%), synaptophysin (38%), NF (nuclear factor) proteins (18 and 8%), and MAP2 (microtubule associated protein 2) (8%).[6]

Other neoplasms with similar pleomorphic morphology are glioblastomas and malignant fibrous histiocytomas.[2] The correct diagnosis is crucial as in contrast to these malignant pleomorphic tumours, PXAs are characterized by a relatively slow clinical course, a favorable prognosis and does not require an aggressive postoperative therapy. The overall prognosis is good, with only 30% of PXA recurring and 20% undergoing anaplastic transformation.[7]

Increased mitotic activity, high MIB-1 and proliferating cell nuclear antigen labeling indices and necrosis are poor prognostic factors, whereas, abundant lymphocytic infiltration is associated with more benign biological behaviour.[7] Other factors influencing clinical outcome include extent of resection, old age, peritumour oedema, and increased FDG uptake on PET. A close follow-up is needed in order to detect any recurrence with malignant transformation.[4,7,8]

Surgery is the treatment of choice; gross total resection achieves better disease control then near or subtotal resection.[6] The role of radiotherapy as adjuvant therapy at diagnosis or recurrence remains controversial,[9] but may be considered in the setting of residual disease or with features such as necrosis or high mitotic index.[2] Few data are available in the literature to support adjuvant chemotherapy in PXAs.[10] Further surgery with added irradiation and/or chemotherapy is recommended in the case of recurrent disease.

## Conclusion

Pleomorphic Xanthoastrocytoma is a rare slowly growing tumour of children and young adults. It can present with unusual, sensory manifestations without any motor symptoms. Classical neuroimaging and histopathological characteristics are useful aids in diagnosis. Treatment with complete surgical excision, if carried out early, usually provides gratifying results. A close follow-up is nevertheless needed, as both recurrence and anaplastic transformation has been reported.

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