



This article has been reviewed by the NeuroWiki Editorial Board



MRI showing PXA

Tumor Class Astrocytoma

Cell of origin glioneuronal precursor

WHO Grade II (may progress)

Survival >60% at 10 years

Treatment Surgical resection

Histologic Features

Gross Cystic tumor with tumor nodule

Micro giant astrocytes and pleomorphic, bizarre nuclei

Radiographic Features

MRI usually a large cystic tumor with an enhancing mass (frequently in the temporal lobe)

CT cystic lesion with a dense nodule

Contents

- [1 Definition](#)
- [2 Epidemiology](#)
- [3 Localization](#)
- [4 Clinical presentation](#)
- [5 Imaging](#)
- [6 Macroscopy](#)
- [7 Histopathology](#)
- [8 Molecular genetics](#)
- [9 Prognosis](#)

Definition

PXA is a low-grade astrocytic neoplasm of young life and early adulthood, with superficial localization in the cerebral hemispheres and involvement of the meninges. PXA is a WHO Grade II astrocytoma.

Epidemiology

PXA typically develops in children and young adults. It accounts for less than 1% of all astrocytic neoplasms.

Localization

PXA typically occurs in the superficial cerebral hemispheres, most commonly in the temporal lobe. Involvement of the meninges is frequent. There are some reports of PXA in the cerebellum.

Clinical presentation

Most patients present with a long history of seizures.

Imaging

On MRI, PXA are discrete, cystic lesions associated with a mural nodule.

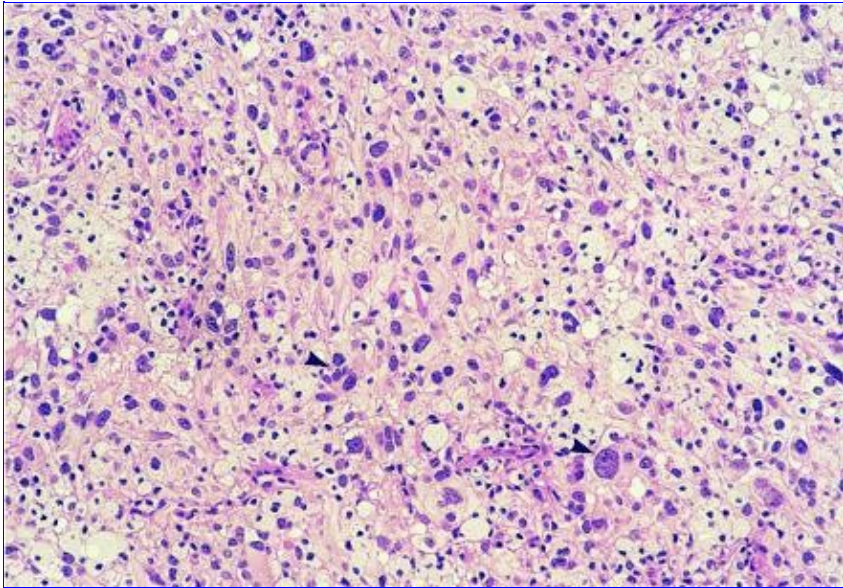
Macroscopy

PXA are discrete lesions, often cystic in nature, with an associated mural nodule. These lesions are often attached to the meninges.

Histopathology

PXA is a pleomorphic tumor composed of fibrillary and giant, often multi-nucleated, neoplastic astrocytes. Diagnostic hallmarks include large, xanthomatous cells expressing GFAP, a dense intercellular reticulin network, and lymphocytic infiltrates. Eosinophilic granular bodies are frequently encountered. Necrosis is a poor prognostic indicator and degeneration to GBM is associated with this feature.

Pleomorphic_xanthoastrocytoma_(PXA)



Photomicrograph of PXA showing moderately cellular glial neoplasm composed of giant astrocytes with pleomorphic nuclei in the area of cellular pleomorphism (arrow heads).

Molecular genetics

The molecular genetics of PXA have not been elucidated, though gains of chromosomes 3, 7 and 1q are not uncommon.

Prognosis

Recurrence-free survival has been reported as 72% at 5 years and 61% at 10 years. Overall survival has been estimated as 81% at 5 years and 70% at 10 years. Extent of resection at initial presentation appears to be the most important factor for both recurrence-free and overall survival.