Pleomorphic Xanthoastrocytoma Treatment

- Pseudoprogression in Pediatric Spinal Pilocytic Astrocytoma and Myxopapillary Ependymoma after Proton Therapy: A Case Series
- Association of Health Disparities with Glioblastoma Treatment and Outcomes: Insights from a 15-Year National Cohort (2005-2020)
- Targeting Glioblastoma Stem Cells: A40s Aptamer-NIR-Dye Conjugate for Glioblastoma Visualization and Treatment
- MiR 329/449 Suppresses Cell Proliferation, Migration and Synergistically Sensitizes GBM to TMZ by Inhibiting Src/FAK, NF-kB, and Cyclin D1 Activity
- Molecular Biomarkers of Glioma
- Epigenetic Alterations in Glioblastoma Multiforme as Novel Therapeutic Targets: A Scoping Review
- Oxamate, an LDHA Inhibitor, Inhibits Stemness, Including EMT and High DNA Repair Ability, Induces Senescence, and Exhibits Radiosensitizing Effects in Glioblastoma Cells
- The Circadian Rhythm Gene Network Could Distinguish Molecular Profile and Prognosis for Glioblastoma

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Standard Treatment Approach 1. Surgical Resection Goal: Maximal safe resection.

Gross total resection (GTR) is associated with significantly improved prognosis.

In cases where GTR is not feasible (e.g., due to proximity to eloquent brain areas), subtotal resection is performed.

2. Histological and Molecular Analysis Confirm diagnosis with neuropathology.

Test for BRAF V600E mutation – present in ~60-70% of PXAs.

Evaluate for anaplastic features: high mitotic activity, necrosis, and/or high Ki-67 index. These suggest Anaplastic PXA (WHO Grade III), which has a more aggressive course.

3. Adjuvant Therapy (case-dependent) For WHO Grade II PXA after GTR:

Often no additional therapy is needed initially.

Close surveillance with MRI every 3–6 months for the first 2 years, then annually.

For subtotal resection or recurrence:

Radiotherapy may be considered.

Chemotherapy options include temozolomide, especially if there is progression or recurrence.

For Anaplastic PXA or progressive/recurrent disease:

Adjuvant radiotherapy and/or chemotherapy are often recommended.

Targeted therapy with BRAF inhibitors (e.g., dabrafenib, vemurafenib) ± MEK inhibitors (e.g., trametinib) has shown promising results in BRAF-mutant cases.

4. Clinical Trials Consider for recurrent or anaplastic PXA, particularly those that are BRAF wild-type or resistant to standard therapies.

Follow-Up Long-term imaging surveillance is critical due to risk of recurrence.

Neurological exams and neurocognitive assessments may be part of regular follow-up, especially in pediatric cases.

The discovery of BRAF mutations within a substantial percentage of Pleomorphic Xanthoastrocytoma fosters a clearer understanding of the pathophysiology of these tumors with clear prognostic and therapeutic implications. These findings are expected to provide insight into the spectrum of clinical behavior observed in PXA, ranging from cure with surgery to diffuse dissemination throughout the neuraxis [1)].

Surgery

Conflicting reports exist on the importance of extent of resection (EOR)

Radiotherapy

Pleomorphic Xanthoastrocytoma Radiotherapy.

1)
Shaikh N, Brahmbhatt N, Kruser TJ, Kam KL, Appin CL, Wadhwani N, Chandler J, Kumthekar P, Lukas RV. Pleomorphic xanthoastrocytoma: a brief review. CNS Oncol. 2019 Nov 1;8(3):CNS39. doi: 10.2217/cns-2019-0009. Epub 2019 Sep 19. PMID: 31535562; PMCID: PMC6880293.