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(CASE REPORT)

Uncommun presentation and clinicopathological features of Anaplasic pleomorphic Xanthoastrocytoma: A case report

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Abstract

Background: Pleomorphic xanthoastrocytoma (PXA) is a rare primary tumor of the central nervous system. Anaplasia is defined by a mitotic index of 5 per 10 high-power fields and/or the presence of necrosis. Complete surgical excision and radiochemotherapy are recommended for this entity.

Case presentation: This is a 29-year-old woman with a one-month history of headaches and intracranial hypertension. Imaging revealed a large right basifrontal mass with a double fleshy component and a predominantly cystic appearance. Surgical excision was macroscopically complete. Immunohistological examination confirmed a pleomorphic anaplastic xanthoastrocytoma (grade 3; WHO, 2021).

Conclusions: We report this case to improve the understanding of the clinicoradiological, histopathological, and immunohistochemical features of this rarely encountered tumor.

Keywords: Anaplastic pleomorphic xanthoastrocytoma; Macroscopic total resection; Immunohistochemistry

1. Introduction

Pleomorphic xanthoastrocytomas (PXA) are rare, low-grade astrocytic brain tumours that account for less than 1% of astrocytomas and have a relatively favorable prognosis [1,2,3].

First described in 1973 [3], the term PXA was coined by Kepes in 1979 [1,3]. In 1993 [1,3], it was officially included in the WHO classification of central nervous system tumors as a grade 2 tumor [3].

In 1999, "PXA with anaplastic characteristics" was defined as PXA with increased mitotic activity, with a high-power field of 5 in 10 and/or necrosis [1,2], and subsequently recognised as a distinct entity, a grade 3 according to the WHO classification of central nervous system tumors (2016) [2].

Our paper reports a rare case of anaplastic pleomorphic xanthoastrocytoma in a 29-year-old female patient with atypical clinical and radiological features.

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2. Case Presentation

- ***Patient information***: The patient is a 29-year-old woman with no notable medical history, presenting to the emergency department with a one-month history of severe headaches. The condition progressed 15 days prior to admission, marked by vomiting and blurred vision.
- ***Clinical findings*:** Upon admission, the patient was in good general condition. Her Glasgow Coma Scale score was 15/15. Fundoscopy revealed grade 2 papilledema, and no seizures were noted in the context of an afebrile state. Neurological examination was unremarkable.
- ***Diagnostic assessment*:** A CT scan (Figure 1) revealed a double cystic and fleshy cortico-subcortical right basifrontal mass with irregular borders and inhomogeneous contrast enhancement, along with intratumoral hemorrhagic foci.

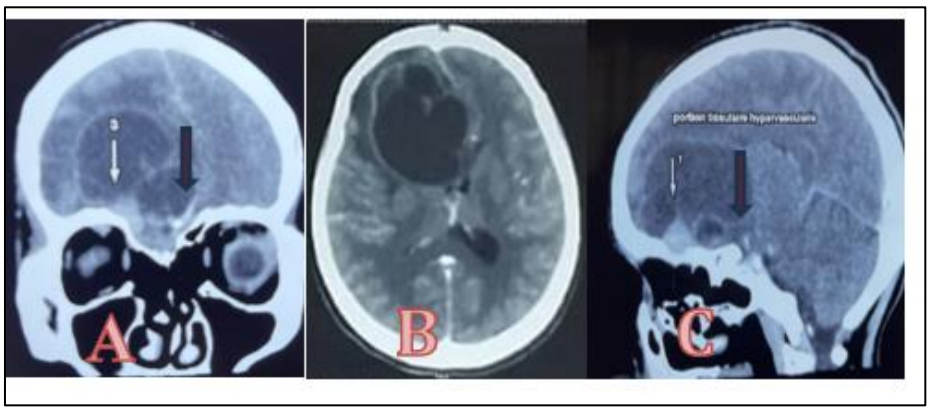

Figure 1 Cerebral CT scan in coronal (A), axial (B) and sagittal (C) sections shows a right basifrontal image with a double component, fleshy (red arrow) and predominantly cystic

On MRI (Figure 2), a well-defined and enhanced right-sided intra-axial frontal and basifrontal cystic mass with a heterogeneous signal was visualized, accompanied by surrounding vasogenic edema. The T1-weighted imaging revealed a double-component tumor: the fleshy part appeared hyperintense, while the cystic part appeared hypointense. A cystic intra-axial lesion was observed, associated with a right basilar frontal tissue component showing contrast enhancement after GADO injection. Septa were noted inside the cyst, with hypersignal on T2-weighted imaging, along with a deviation of the midline and compression of the frontal horn of the right ventricle.

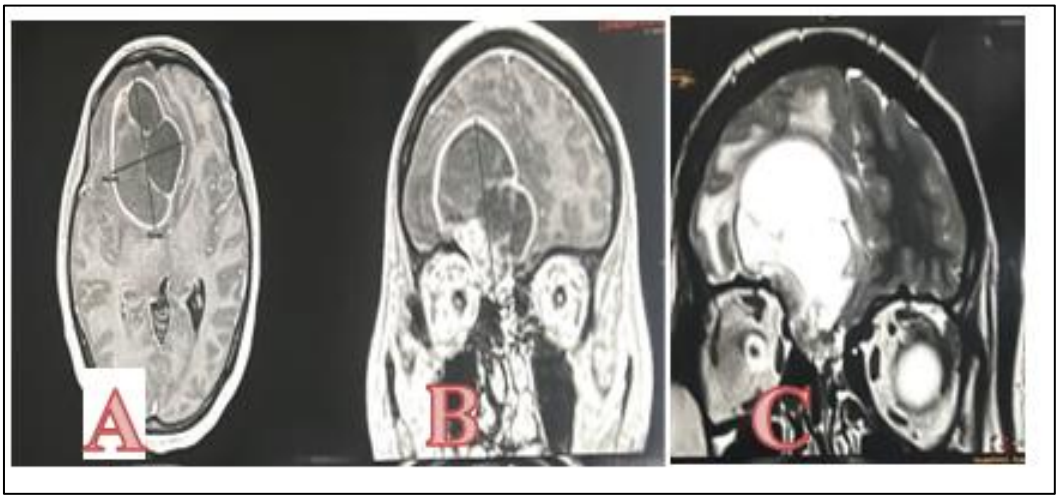

Figure 2 Cerebral Magnetic resonance imaging (T1) axial(A) and coronal(B) with contrast showing the cystic wall with intense enhancement and the cystic zone without enhancement. Coronal (T2) magnetic resonance imaging (C) showing the isointense cystic wall and the homogeneous hyperintense appearance of the cystic zone. Obvious vasogenic edema surrounded the mass

2.1. Therapeutic intervention

The patient underwent surgery, which included a cyst puncture and a macroscopically complete excision of the lesion (Figure 3).

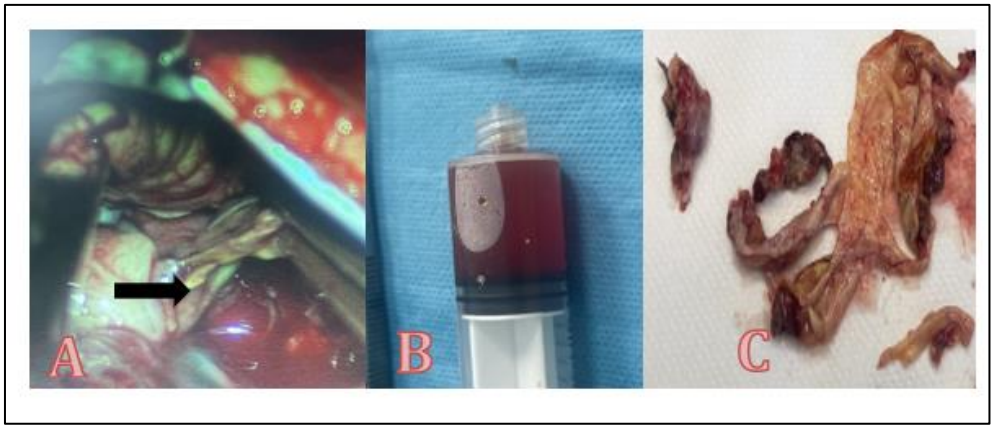

Figure 3 A: Intraoperative discovery of kystique tumor B: hematic appearance of cyst fluid C: macroscopic appearance of the cyst capsule

Microscopic analysis revealed cerebral parenchyma containing a highly cellular tumor proliferation composed of layers of globoid cells, as well as sheets of globoid, rhabdoid, spindle, and epithelioid cells with marked nuclear atypia. The mitotic activity was estimated at 5 mitoses per 10 GX40 fields.

Immunohistochemical staining (Figure 4) demonstrated diffuse positivity of tumor cells for glial fibrillary acidic protein (GFAP) and focal positivity for anti-OLIG2 antibody and PS100. The tumor cells were negative for synaptophysin, anti-IDH1, and CD34. Sequential analysis revealed no mutations in the IDH1 gene or the BRAF V600E mutation. Ki-67 immunostaining showed heterogeneous marking with a proliferation index of 10%.

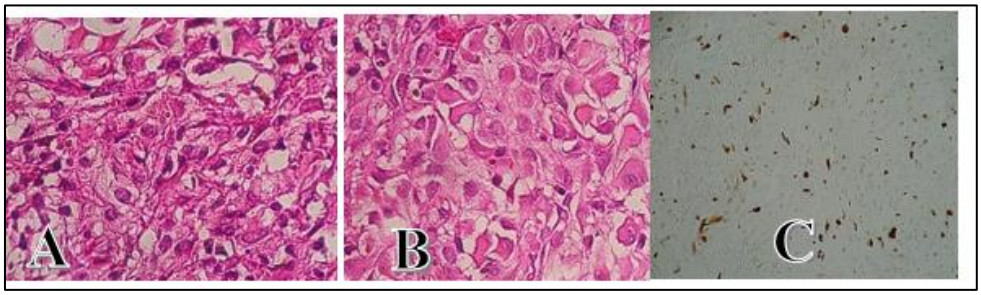

Figure 4 A: Astrocytic glial proliferation of pleomorphic cell sheets showing marked nuclear atypia and mitosis estimated at 5mitosis/10fields HEX20 B: Tumor cells are linked to xanthelasmic foamy histiocytes and harbor hyaline globules. HEX40; C: High mitotic index estimated at 20%

The recognition of microscopic foci of necrosis led to the final diagnosis of anaplastic pleomorphic xanthoastrocytoma (APXA), classified as grade 3 in the WHO classification system for central nervos system tumors.

2.2. Follow-up and Outcome

The surgical follow-up was straightforward. The patient remained conscious and exhibited no sensorimotor deficits. A postoperative CT scan (Figure 5) confirmed postoperative remodeling. She was discharged and referred for adjuvant radiotherapy. Three months later, the patient was doing well and had returned to her usual activities.

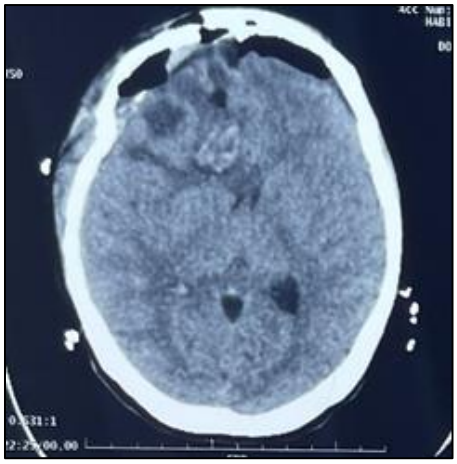

Figure 5 Post operatory CT scan

3. Discussion

Anaplastic pleomorphic xanthoastrocytoma (a PXA) is a rare and highly malignant astrocytoma, accounting for less than 0.5% of astrocytomas and less than 0.05% of primary intracranial tumors [4,3]. This condition is most prevalent in children and young adults, with a markedly lower incidence observed in patients over the age of 40 years [4].

It can be classified as either primary (de novo) or secondary, resulting from anaplastic transformation of recurrent PXA.

The tumour is supratentorial and localised in the superficial cortex, predominantly in the temporal or temporoparietal lobes, followed by the parietal, occipital and frontal lobes. Cases involving the cerebellum, ventricles, thalamus, spinal cord and pineal gland have been documented in the literature [4].

In the present case, the lesion was identified in the right frontal and basifrontal cortex. While these locations are not the most common, they have been documented in previous literature.

The clinical manifestations are not specific, but seizures are the most common presenting symptom. This patient, however, presented with a history of severe headaches and no evidence of epileptic seizures.

These tumours frequently comprise both solid and cystic components. APXA is characterised by a significantly larger tumour size than PXA, with greater tumour enhancement and peri-lesional oedema.

On computed tomography, PXA appears hypodense or isodense and may be well or poorly circumscribed. The solid component and the cyst wall frequently exhibit enhancement, and a minimal degree of edema is observed in the vicinity of the tumor.

On magnetic resonance imaging, the solid component demonstrates an isointense signal on T1-weighted images, with a signal intensity similar to that of the surrounding tissue. On T2-weighted imaging, the tumour is isointense or hyperintense, with small peritumoral oedema present. T1-weighted post-gadolinium imaging reveals heterogeneous contrast enhancement of the fleshy component and the peripheral rim of the cyst.

Perfusion-weighted imaging (PWI) demonstrates an increase in blood flow volume, indicating the presence of microvascular hyperplasia in the anaplastic cells and a substantial blood flow in a PXA. The cystic fluid displays either isointense or hyperintense characteristics in comparison to the CSF, contingent on the imaging sequence [3].

Other desmoplastic neuroepithelial neoplasms, including gliofibroma, gliosarcoma, desmoplastic childhood ganglioglioma, and desmoplastic childhood cerebral astrocytoma, exhibit certain clinical, radiological, and pathological characteristics in common with aPXA. Furthermore, epithelioid glioblastoma can be challenging to differentiate from aPXA due to the presence of similar histopathological characteristics.

The standard treatment is surgical excision, with the objective of achieving total removal of the affected tissue. Given the highly malignant nature of aPXA, there is a significant potential for its volume to increase rapidly over a relatively

short period. Furthermore, the recurrence interval for aPXA following total surgical resection is 14 months, as evidenced in [3].

Immunohistochemical staining demonstrated that the tumor cells exhibited differential positivity for GFAP , indicative of an astrocytic origin for the tumor tissue.

Ki-67 immunostaining exhibited heterogeneous labelling with a proliferation index of 10%. BRAF V600E mutation is prevalent in 65% of aPXA cases [2,3]. In our case, the mutation was not present.

It is standard practice to recommend additional chemotherapy and/or radiation therapy for patients diagnosed with aPXA. All patients in the Rutkowski M Jet series [5] exhibited disease progression despite receiving adjuvant radiation and chemotherapy. The median time to progression was 20 months, with a one-year progression-free survival rate of 57% [4]. Given the rarity of aPXA, there is limited research identifying risk factors for overall survival or progression-free survival. The clinical outcome is influenced by several factors, including the extent of resection and histological features such as the mitotic index, necrosis, and lymphocyte infiltration.

The prognosis for aPXA is poor, as the disease is highly invasive and prone to recurrence. The 5-year survival rate is less than 57.1% [4, 2], with a mortality rate of 15-20% [2].

List of abbreviations

- APXA: Anaplastic pleomorphic xanthoastrocytoma
- CSF: Cerebrospinal fluid
- MRI: Magnetic Resonance Imaging
- PXA: Pleomorphic xanthoastrocytoma
- GFAP: glial fibrillary acidic protein

4. Conclusion

APXA is a particularly rare and highly malignant astrocytoma that requires individualized identification and treatment. In such cases, complete surgical excision combined with adjuvant radio chemotherapy is recommended. An accurate initial diagnosis, often with the assistance of experienced neuropathologists, is a critical first step in implementing aggressive and proactive management strategies. A comprehensive understanding of the disease's molecular characteristics is essential, as these not only inform prognosis but also guide therapeutic management. Regardless of the treatment strategy employed, the presence of anaplastic features is associated with a poor prognosis.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare that they have no competing interests

Statement of informed consent

The authors certify that they have obtained all appropriate patient consent.

Authors contributions

SA contributed to conceptualization,writing,original draft editing. EM H contributed to writing and supervision. MY O contributed to writing.AM contributed to writing.NC performed the histological examination, H EO performed the histological examination and was a major contributor in writing the manuscript. YA contributed to supervision and validation.

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