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ASTROBLASTOMA IN AN 8-YEAR-OLD FEMALE: A RARE PEDIATRIC HIGH-GRADE GLIOMA

Paediatrics

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ABSTRACT

Astroblastoma is a rare and aggressive glial neoplasm predominantly affecting children and young adults. We present the case of an 8-year-old female who exhibited neurological symptoms including progressive hemiparesis, headache, vomiting, and fever. MRI identified a large heterogeneous mass in the posterior frontal lobe. Histological analysis revealed classical features of a high-grade glial tumor, including perivascular pseudo-rosettes, microvascular proliferation, and necrosis. Immunohistochemistry confirmed the diagnosis with strong positivity for GFAP, S100, vimentin, and focal EMA expression. The tumor was negative for synaptophysin, ruling out ependymoma. This case highlights the critical role of histopathological and immunohistochemical evaluation in diagnosing rare CNS tumors.

KEYWORDS

Astroblastoma, Perivascular Pseudo-rosettes, GFAP, Pediatric Glioma, Ependymoma Differential

INTRODUCTION

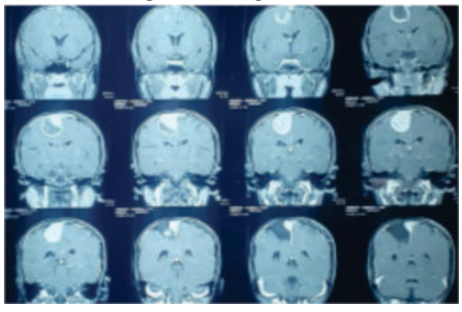
Astroblastoma is a rare glial neoplasm that typically affects children and young adults. We report a case of an 8-year-old female presenting with progressive left-sided weakness, headache, vomiting, and fever. Imaging revealed a high-grade lesion in the right posterior frontal lobe. Histopathology showed hypercellularity, pseudo-palisading necrosis, perivascular pseudo rosettes, and vascular proliferation. Immuno-histochemistry demonstrated positivity for GFAP, S100, vimentin, and focal EMA, confirming the diagnosis of astroblastoma. The differential diagnosis included ependymoma, which was excluded based on distinct cytological and IHC differences. Early diagnosis and classification are essential due to the tumor's aggressive behavior and implications for treatment Astroblastoma is a rare neuroepithelial tumor, accounting for <3% of primary brain gliomas. It is characterized by perivascular pseudo-rosettes and often presents in children and adolescents. The case discussed herein exemplifies the clinical, radiological, and histopathological features of this entity, and highlights the role of immunohistochemistry in distinguishing it from histologically similar tumors like ependymoma.

Case Study

An 8-year-old girl presented with: Progressive left-sided weakness (2 months), Headache (2 days), Vomiting and fever (1 day). She was initially diagnosed in 2019 with desmoplastic infantile ganglioglioma, for which surgery and partial chemotherapy were administered.

Radiological Findings

MRI (24/05/2023) revealed a large (67 × 56 × 46 mm), irregular, heterogeneously enhancing mass in the right posterior frontal lobe with cystic and solid components, and perilesional edema.



Gross Pathology

Received multiple soft, friable, grey-brown tissue fragments aggregating to 6 × 6 cm. The largest measured 2 × 3 cm; the smallest 1 × 1.5 cm.

Microscopy: Histology revealed: Hypercellularity, Vascular proliferation, Pseudo-palisading necrosis, Perivascular pseudo-rosettes with broad cytoplasmic processes, True ependymal rosettes also noted. These findings were indicative of a high-grade glial tumor.

Differential Diagnosis

Astroblastoma: Compact growth with perivascular pseudo-rosettes, Broad, non-tapering processes, High mitotic activity (>5/10 HPF), WHO classification: Circumscribed astrocytic gliomas. Ependymoma: True rosettes and perivascular pseudo-rosettes, Fibrillar areas and infiltrative margins, Smaller, less pleomorphic nuclei.

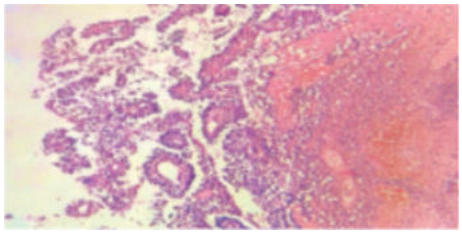


Fig:1. 4x Showing Areas of Vascular Proliferation Necrosis and Perivascular Pseudo-rosettes Orosste

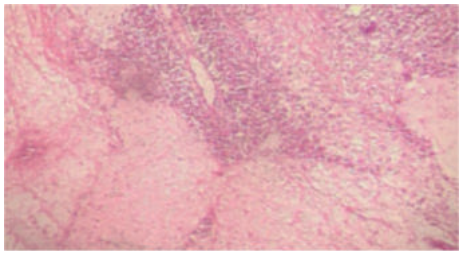


Fig:2. 20x Showing Pseudo-palisading Necrosis

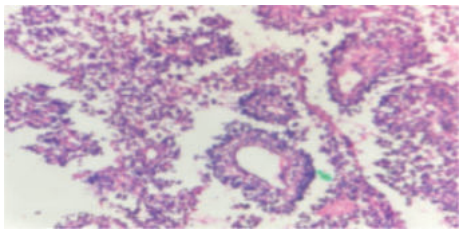


Fig:3. 20x Showing Vascular Proliferation and Perivascular Pseudo-rosettes

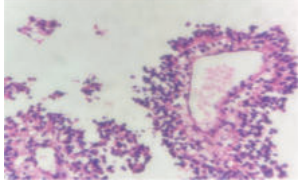


Fig:4. 40x Showing Perivascular Pseudo-rosettes

DISCUSSION

Astroblastoma is a diagnostically challenging tumor due to its rarity and overlapping morphological features with other gliomas, notably ependymoma and high-grade astrocytoma. The differential diagnosis becomes even more critical when the histopathological features show both perivascular pseudo-rosettes and high mitotic activity. The current WHO classification places astroblastoma under circumscribed astrocytic gliomas, reflecting its partially defined histogenesis and biological behavior. Although previously considered a low-grade tumor, newer literature supports the recognition of both low- and high-grade variants based on mitotic activity, necrosis, and endothelial proliferation—elements assessed by the 'AMEN' criteria (Atypia, Mitosis, Endothelial proliferation, Necrosis).

This case exemplifies a high-grade astroblastoma with more than five mitoses per 10 HPF, vascular proliferation, and necrosis—criteria supporting a WHO Grade III or IV designation. Immuno-histochemistry played a pivotal role in ruling out mimickers. Astroblastomas are strongly positive for GFAP, vimentin, and S100, whereas synaptophysin negativity helps exclude ependymoma. The focal EMA positivity in astroblastoma also contrasts with the classic perinuclear dot-like EMA pattern in ependymomas.

Clinically, astroblastomas often present with signs of increased intracranial pressure, focal neurological deficits, and seizures. Radiologically, they appear as well-circumscribed masses with both solid and cystic components, and often mimic other pediatric brain tumors. Treatment typically involves maximal safe surgical resection followed by adjuvant radiotherapy, particularly for high-grade tumors. The role of chemotherapy remains unclear due to limited data.

Given the paucity of cases and evolving understanding of the tumor's molecular and clinical behavior, accurate diagnosis through integration of histopathology, immunohistochemistry, and emerging molecular markers is essential. Further multicentric studies and case series are needed to develop standardized therapeutic protocols and to better understand prognosis and recurrence risks.

CONCLUSION

In conclusion, this case report underscores the importance of a multidisciplinary diagnostic approach to pediatric brain tumors, particularly those with overlapping histopathological features. Astroblastoma, though rare, should be considered in the differential diagnosis of glial tumors with perivascular pseudo-rosettes. Histological grading and immunohistochemical profiling are indispensable for distinguishing it from ependymomas and other CNS neoplasms.

Accurate grading influences therapeutic decisions and helps anticipate tumor behavior. High-grade variants require prompt surgical intervention and postoperative adjuvant therapy to reduce the risk of recurrence. Increased awareness, early diagnosis, and reporting of such rare tumors are crucial for enhancing our understanding and management of astroblastoma in pediatric patients.

Immunohistochemistry (IHC)		
Marker	Astroblastoma	Ependymoma
GFAP	Strong Positive	Positive
S100	Positive	Positive
Vimentin	Positive	Positive
EMA	Focal positivity	Perinuclear dot-like positivity
Synaptophysin	Negative	Positive
CD56, CD99	Variable	Positive (luminal and membranous)

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