Pediatric astroblastoma: Case report and review of the literature

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ABSTRACT

Astroblastoma is a rare primary brain neoplasm with uncertain histopathological origin and unpredictable clinical behavior. WHO classified it as neuroepithelial tumor. However, grading for this variant of the tumor is not yet established. Complete excision without any adjuvant radiotherapy is sufficient to treat low-grade astroblastoma. We present a case of a 5-year-old female child presenting with seizures. Her magnetic resonance imaging of the brain showed well-defined cortical-based intra-axial mass lesion in the right frontal lobe. A gross total excision of the tumor was done. Histopathological features were suggestive of astroblastoma.

Key words: Astroblastoma, glial tumor, management

INTRODUCTION

Astroblastoma is a rare neuroepithelial tumor of uncertain origin. Astroblastomas are known but rare neuroglial tumor.[1] Bailey and Cushing initially described these rare tumors. Later, detailed description regarding these tumors was published by Bailey and Bucy.[2] These tumors are easily misdiagnosed as they are rarely seen in clinical practice and share common radiological and histological appearances with other glial neoplasms. We encountered such a case of pediatric astroblastoma in clinical practice. Our aim was to discuss the clinical presentation, pathology, differential diagnosis, and management of this rare entity.

CASE REPORT

A 5-year-old female child presented with two episodes of generalized tonic-clonic seizures followed by loss of consciousness, starring look, and vomiting since 3 days. Neurological examination was within normal limits. Her computed tomography scan showed large hyperdense focal lesion with multiple coarse calcification in right basifrontal region [Figure 1]. Magnetic resonance imaging (MRI) of the brain revealed well-defined cortical-based intra-axial mass in right frontal lobe extending into adjacent white matter without any surrounding edema [Figures 2-5]. Contrast MRI exhibited mild enhancement [Figures 6 and 7]. Magnetic resonance spectroscopy showed increased choline levels [Figure 8]. She underwent gross total excision of the tumor via right frontal craniotomy. Surgical findings visualized grayish colored, soft, suckable, and moderately vascular tumor underlying a thin rim of normal cortex over it, with few areas of calcification. This could be easily differentiated from surrounding normal grey/white matter. Postoperative period was uneventful. The patient was neurologically stable without any fresh deficits. Histopathological diagnosis was consistent with low-grade astroblastoma [Figure 9]. Immunohistochemistry revealed strong glial fibrillary acidic protein (GFAP) labeled tumor cells with perivascular accentuation [Figure 10]. Other findings included 5–6% MIB-1 [Figure 11] labeling, negative p-53 with retained IN1-1 expression.

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DISCUSSION

Around 40 cases of astroblastoma have been reported in the literature after Bailey *et al.* reported this condition for the first time in 1930.[2] Astroblastoma is most commonly seen in children and young adult and are rarely congenital.[3] They usually present as supratentorial tumors in the cerebral hemispheres located peripherally with both solid and cystic components. These tumors

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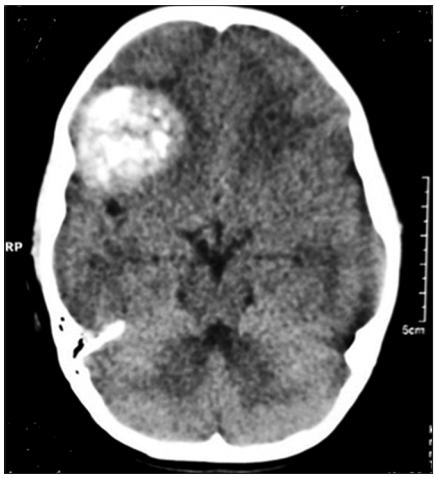

Figure 1: Well-circumscribed lobulated hyperdense mass lesion with multiple coarse calcifications in right frontal lobe without significant mass effect or edema

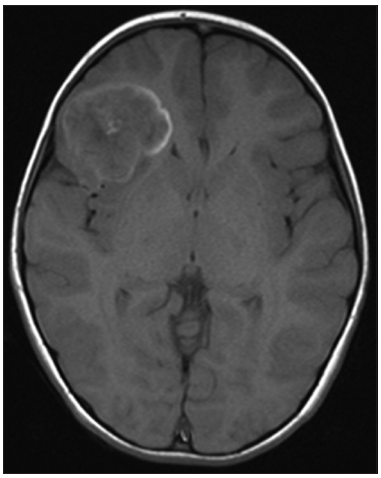

Figure 2: Well-defined lobulated mass lesion predominantly isointense to grey matter in T1-weighted imaging in right frontal lobe, thin rim of peripheral hyperintensities and small hyperintense foci in the matrix

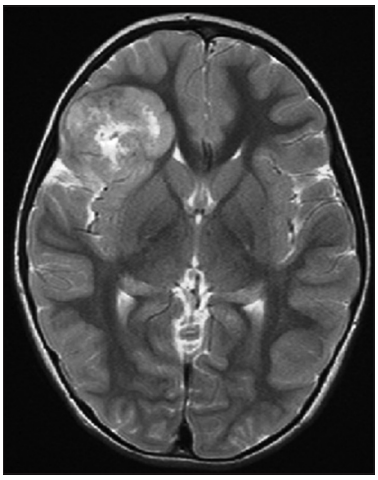

Figure 3: On T2-weighted imaging lesion is predominantly isointense to grey matter. Central matrix shows hyperintense foci without mass effect or edema

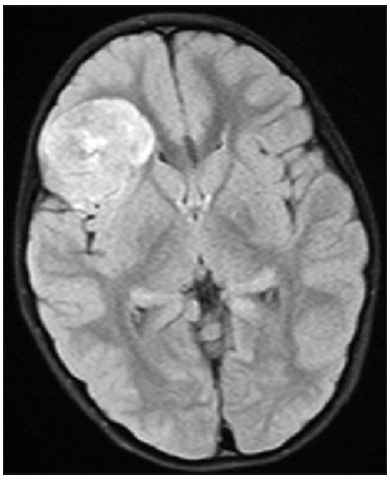

Figure 4: On fluid-attenuated inversion recovery lesion is mild heterogeneously hyperintense signal

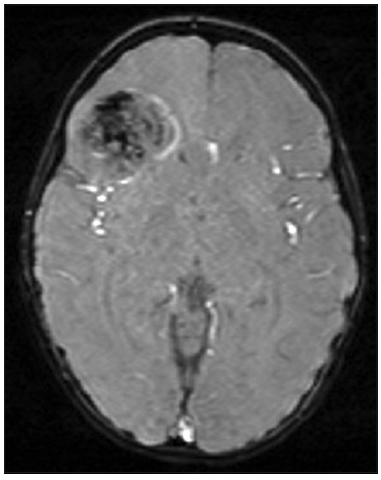

Figure 5: Large areas of blooming/T2 shortening within the matrix of lesion suggestive of calcifications

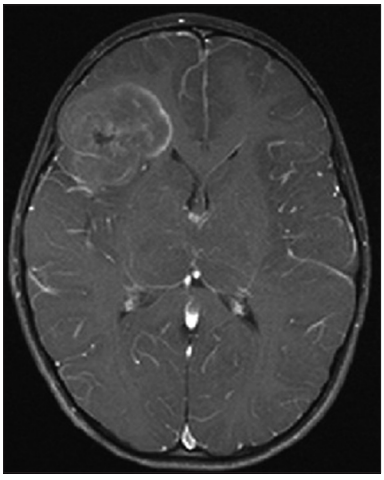

Figure 6: Contrast axial mild heterogeneous patchy enhancement within the lesion

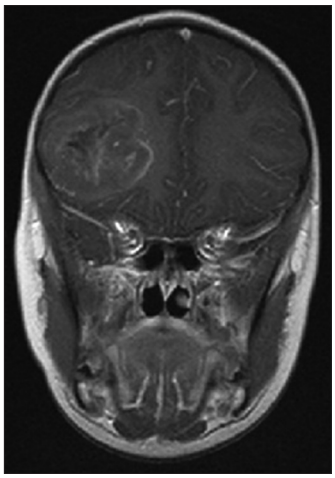

Figure 7: Contrast coronal mild heterogeneous patchy enhancement within the lesion

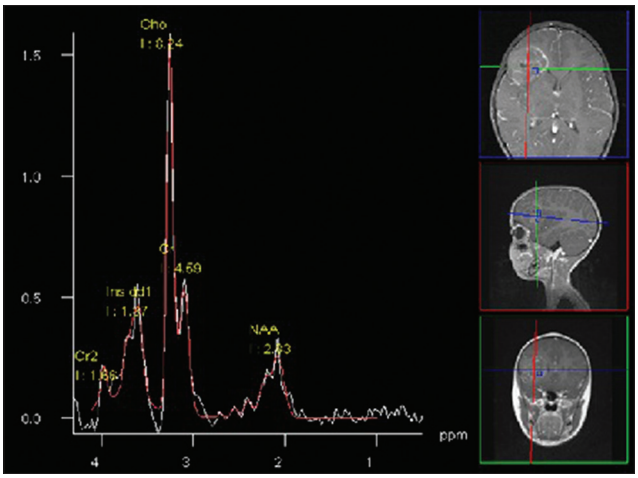

Figure 8: Spectroscopy: Significant elevation of choline peak with decreased N-acetyl aspartate, increased choline/creatinine ratio suggestive of tumor profile

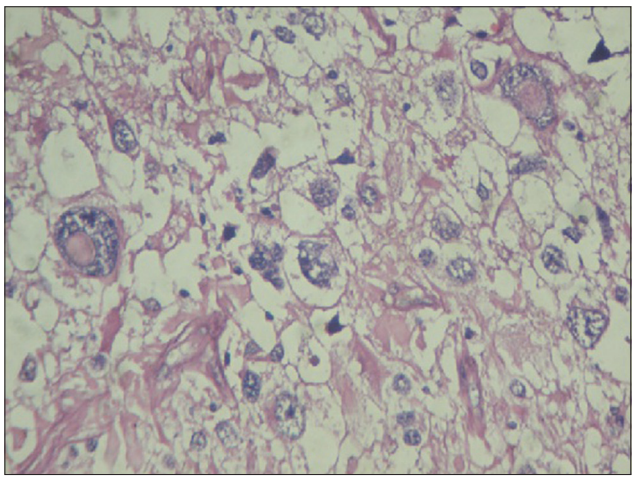

Figure 9: Section shows tumor fragment that are composed of plump cells with glass eosinophilic cytoplasm, eccentric vesicular nuclei. Admixed are bloated cells with vacuolated cytoplasm. Cells demonstrated degenerative atypia with intranuclear cytoplasmic inclusion. Several fragments show striking perivascular arrangement of tumor cells with broad tapering extending to the vessel wall

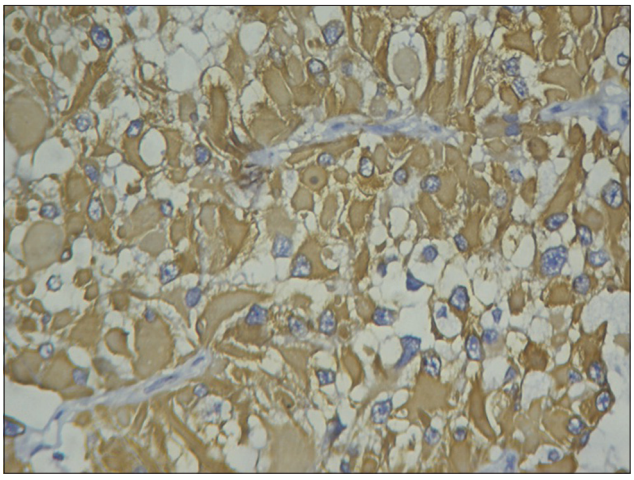

Figure 10: Glial fibrillary acidic protein strongly labels tumor cells with perivascular accentuation

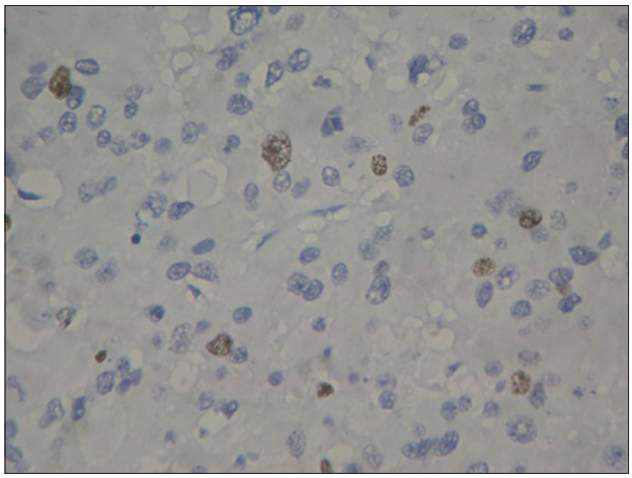

Figure 11: MIB-1 labeling index is 5–6%

also have been reported in other sites such as corpus callosum, cerebellum, brain stem, and optic nerves. Clinical symptoms and signs depend upon the location and size of the tumor with headache and seizures being the most common.[3] No sex predilection[2] or female preponderance[4] was reported in different studies. In our case, it was a female child.

Bell *et al.*[5] reported the largest series of supratentorial astroblastomas exclusively located peripherally and comprising both solid and cystic components. On MRI, solid and cystic components have characteristic bubbly appearance. Calcification is a consistent feature on imaging, and less common findings are intratumoral hemorrhages. Our case was unique with unusual radiological presentation, that is, solid with large nodular or clumped calcification Contrast MRI of brain shows heterogeneous enhancement

with astroblastomas. Usually, these tumors show little peritumoral edema due to lack of infiltration, even though high-grade tumors show infiltration.[6] Differential diagnosis of astroblastomas based on imaging includes high-grade astrocytomas, oligodendrogliomas, primitive neuro-ectodermal tumors, ependymomas, meningiomas, and atypical rhabdoid tumors.

Astroblastomas are histologically defined by the presence of perivascular pseudorosettes and prominent perivascular hyalinization.[7] They may resemble astrocytic tumors, ependymomas and nonneuroepithelial tumors due to their astroblastic aspects. The absence of fibrillar pattern is an essential feature in distinguishing astroblastomas from other glial neoplasms. No definitive cell of origin has been established. Hypothesized to be arising from abnormally persisting group of embryonal precursor cells like tanycytes which is a transitional cell type between astrocytes and ependymal cells during embryogenesis.[8] Astroblastomas are immunoreactive for GFAP, S100 protein, and vimentin. Focally, they show cytoplasmic immunoreactivity for epithelial membrane antigen.[9] Bonnin *et al*. classified astroblastomas into two distinct histological types, a low-grade type with better-differentiated pattern and an expected favorable postoperative prognosis and a high-grade type showing more anaplastic microscopic features with poor prognosis.[10] High-grade lesions show multifocal or unifocal regions of elevated mitotic indices, vascular proliferation, high cellularity, anaplastic proliferation, and necrosis with pseudopalisading nature.

Natural history of astroblastoma is midway between astrocytoma and glioblastoma, and gross total resection is the best way of treating an astroblastoma.[8] Pizer *et al*. have reported radiological evidence of tumor showing positive response to chemotherapy.[3] Bonnin and Rubinstein in their large series of astroblastomas demonstrated that high-grade astroblastomas who did not receive adjuvant therapy had shorter survival rate.[10] Our case was a low-grade astroblastoma, hence was advised to be on regular follow-up and no adjuvant radiotherapy was advised.

CONCLUSION

Reports of such rare entity bring awareness to clinicians to improve the indices of suspicion as radiological features of such tumors are heterogenous. This entity should be considered in the diagnosis of intra-axial, supratentorial tumors solid and cystic components with areas of calcifications. Gross total resection of tumors of low-grade is sufficient and with added adjuvant radiotherapy is reserved for high-grade tumors.

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