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Case Report

Cerebral astroblastoma: case report and literature review

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Astroblastomas are uncommon neuroepithelial tumors of young adults and children. Their diagnosis is made delicate by their rarity, the absence of specific imagery and the histopathologic appearance resembling other glial neoplasms. A 10-year-old female was complaining of progressive headache and diplopia. On examination, she had abilateral 6th nerve palsyand papilledema. Brain magnetic resonance imaging (MRI) depicted awell-demarcated, peripherally enhancing mixed solid and cystic mass in the left frontal lobe. She underwent gross total resection of the lesion. The histopathologic diagnosis was suggestive of high-grade astroblastoma. She had post operative radiotherapy. There was no tumor recurrence after 14 months of follow-up. We review literature for the clinical and radiologic aspects of this tumor, and to highlight the diagnostic tools of this particular tumor.

Keywords: High grade, astroblastoma

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INTRODUCTION

Astroblastomas are uncommon neuroepithelial tumors of uncertain origin. They occur predominantly in the cerebral hemisphere of young adults and children (Rosenblum, 1996; Port et al., 2002). Incidence of astroblastoma is only 0.45–2.8% of all neuroglial tumors. They can be easily misdiagnosed as they are rarely encountered in clinical practice and share common radiological and histopathologic appearances with other glial neoplasms. We report a rare case of astroblastoma. The clinical presentation, pathology, differential diagnosis, and treatment of this rare entity are discussed.

Case Report

A 10-year-oldhealthy girl presented to our hospital with progressive headache and diplopia since 3 months. The neurological examination found bilateral 6th nerve palsy with papilledema. Brain magnetic resonance imaging (MRI) revealed a well-demarcated mass of 5 cm in diameter in the left posterior frontal lobe with large, lobulated, well defined, peripheral, supra-tentorial, solid and cystic components enhanced by Gadolinium. The lesion is surrounded by moderate edema.

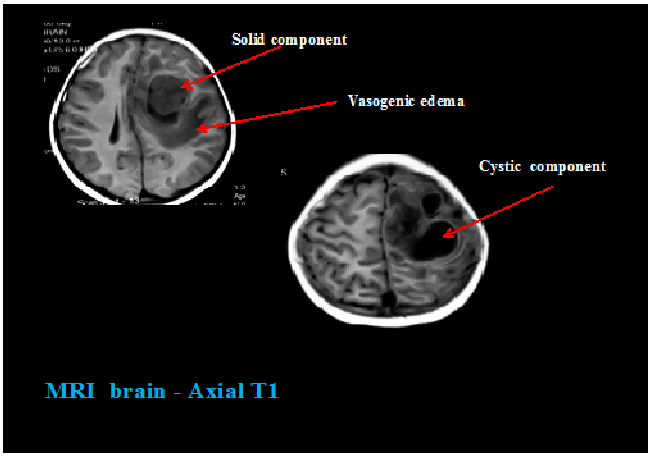

Figure 1. Axial T1 WI that demonstrates a well demarcated mass with solid and large cystic component located supratentorially in the left frontal lobe with mild vasogenic edema.

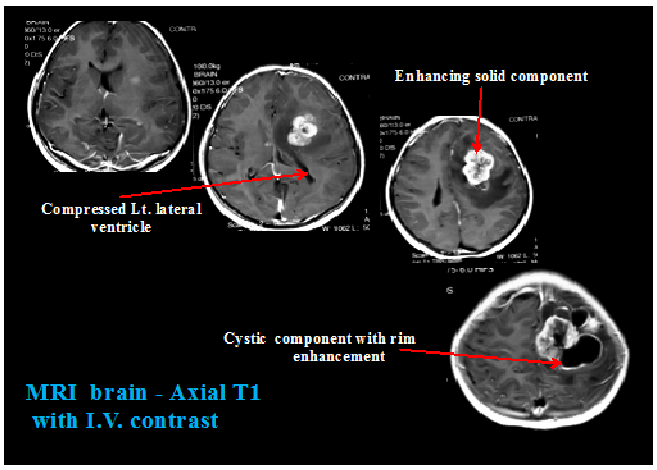

Figure 2. Axial and coronal T1WI post IV contrast that shows enhancing solid mass with peripherally (rim enhancing) cystic component.

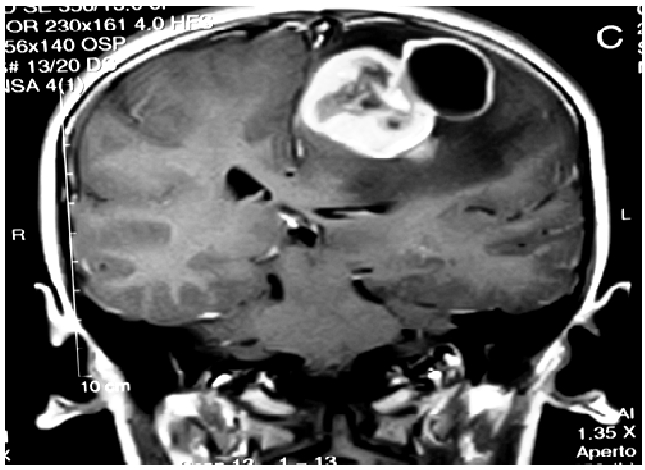

Figure 3. T1W1post IV contrastthat showing solid with cystic componant.

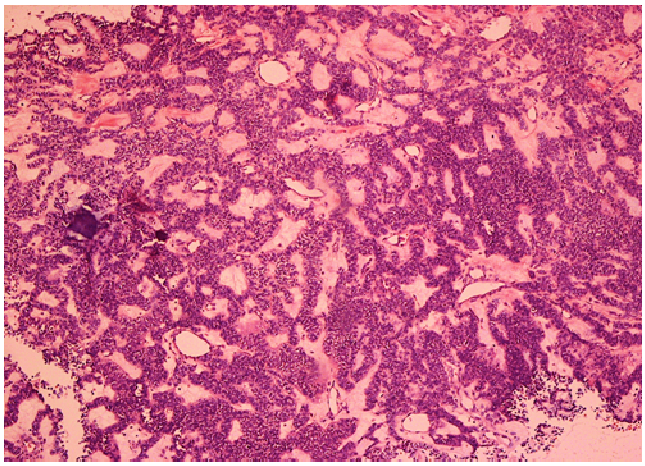

Figure 4. Low-power field slide – Papillary architecture and pseudorosettes around central hyalinized thickened blood vessels

It extend from peripheral cortex to the peri-ventricular region (notice that the lesion does not reach the ventricular wall) (Figures 1, 2 and 3).

The patient underwent gross total resection of the lesion through left posterior frontal craniotomy. The excision went through the superficial cystic lesion after evacuation of its xanthochromic content. The cystic wall was resected along with the solid mass till we reached the normal aspect of the brain. The tumor was rubbery, vascular, and well demarcated from the surrounding brain.

The postoperative course was uneventful. There was no residual tumor on the follow brain CT. The histopathologic diagnosis was high-grade astroblastoma. Microscopically, the tumor showed papillary architecture and pseudorosettes around the central hyalinized thickened blood vessel throughout the tumor (figures 4, 5, 6 and 7) with large area of necrosis. Individual cells were polygonal to spindled, showing moderate eosinophilic cytoplasm and eccentrically placed nuclei. The nucleus was round with coarse chromatin. Mitotic figures were frequently observed. Blood vessels were mostly of capillaries without smooth muscle layers. There was no glial fibrillarity in the fibrovascular stalk. The macrophages were frequently infiltrated in fibrovascular stalks. With presense of feature suggestive of high-grade lesion, as focal area of high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation, and necrosis, Immunohistochemical showscytoplasmic processes of tumor cells composing perivascularpseudorosettes showed strong positive reaction for glial fibrillary acidic protein (GFAP) . Tumor cells in discohesive areas showed GFAP-positive short cytoplasmic processes. The tumor cells showed diffuse strong positivity for S-100 protein, vimentin and neuron specific enolase (NSE), and focal positivity for epithelial membrane antigen (EMA) and CAM 5.2 (Figure 8). The tumor cells were negative for synaptophysin,

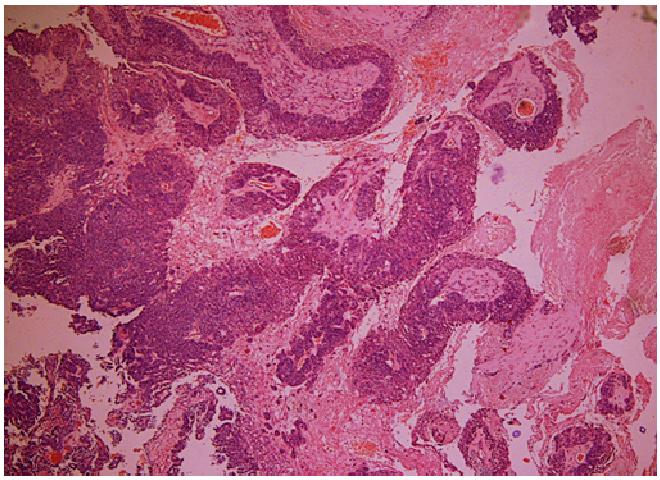

Figure 5. Low-power field slide – Papillary architecture around central hyalinized thickened blood vessels notice the large area of necrosis

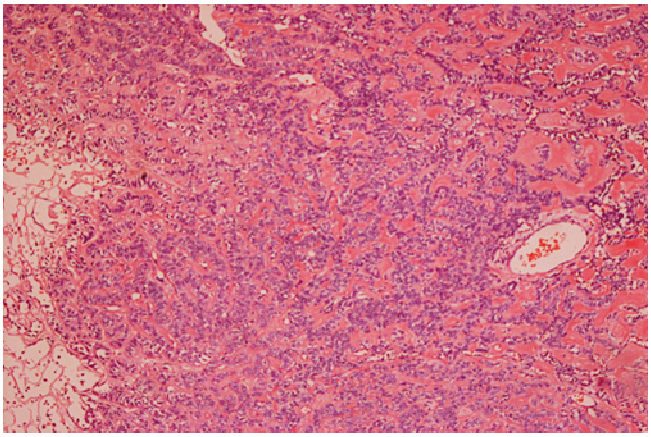

Figure 6. Low-power field slide – cellular area with extensive hyalinization and thickened blood vessels

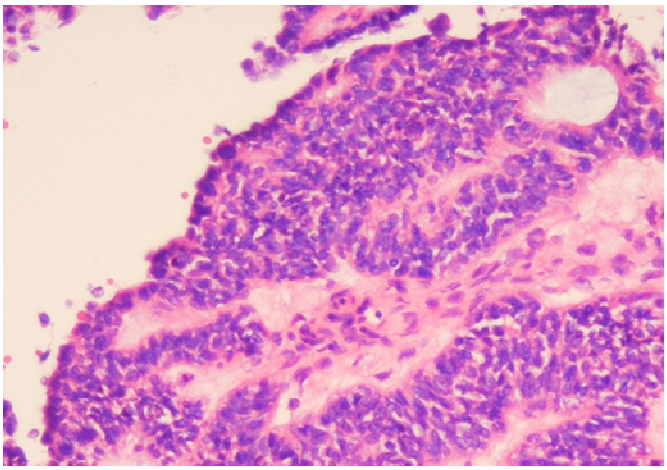

Figure 7. Histopathologic slide – High-power field pseudorosettes around central hyalinized thickened blood vessels with individual cells being polygonal to spindled, showing moderate eosinophilic cytoplasm and eccentrically placed nuclei

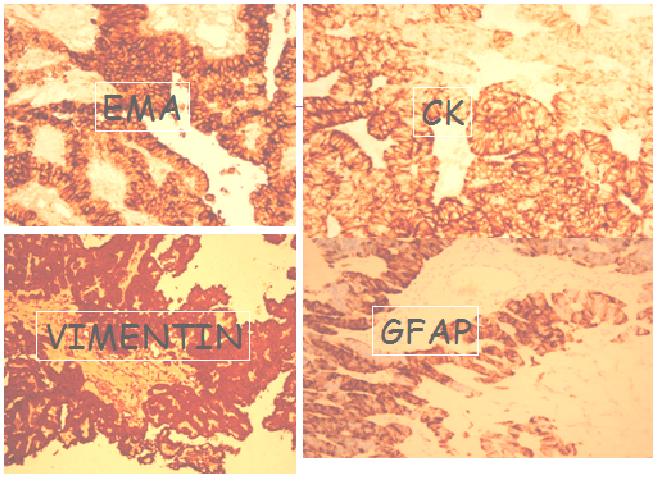

Figure 8. The broad GFAP-positive glial processes surround blood vessels (×400).The tumor cells are positive for EMAprotein ,vimentin and LMWCK

neurofilament protein (NFP), pan-cytokeratin, high molecular weight keratin (HMWK) immunostains.

The patient had adjuvant radiotherapy. At the latest follow up,14 months after the surgery, no recurrence was depicted on MRI.

DISCUSSION

Approximately 40 cases of astroblastoma have been reported in the literature since Bailey and Bucy reported the condition for the first time in 1930. Bailey and Bucy believed that astroblastoma originated from astroblasts, an intermediate stage between glioblasts and astrocytes (Bailey and Bucy, 1930).

These are rare glial tumors usually located in the cerebral hemisphere. However, tumor invasion has also been reported into corpus callosum, cerebellum, brain stem, and optic nerve (Pizer et al., 1995; Bonnin and Rubinstein, 1989). Clinical signs and symptoms depend on the location and size of the neoplasm, with headache and seizures being the most frequently encountered symptoms. Astroblastomas are mostly seen in children and young adults like in ourcase, but congenital cases have also been reported rarely (Pizer et al., 1995).

Bell *et al*. reported the largest imaging series with 12 cases of astroblastomas. As per their report, astroblastomas are almost exclusively seen supratentorially and are peripheral in location with both solid and cystic components (Bell et al., 2007). Our case showed typical solid cystic lesion with rim enhancement without calcification. Based on imaging, the differential diagnoses for astroblastomas include high-grade astrocytoma, pilocytic astrocytoma, oligodendroglioma, primitive neuroectodermal tumor, ependymoma, and atypical rhabdoid tumor. Unlike in high-grade tumors,

perilesional edema is usually less in astroblastomas including high-grade variants.

Astroblastomas are defined histologically by the presence of perivascular pseudorosettes and prominent perivascular hyalinization (McLendon et al., 1998). They may resemble astrocytic tumors, ependymomas, and non-neuroepithelial tumors due to their astroblastic components. Lack of fibrillarity is an essential feature in distinguishing astroblastomas from other glial neoplasms. Immunohistochemically, astroblastomas are immunoreactive for GFAP, S-100 protein, and vimentin. The majority displays a focal cytoplasmic immunoreactivity for EMA.

Astroblastomas along with gliomatosiscerebri and polar spongioblastoma are included in neuroepithelial tumors of uncertain origin and are grade 4 WHO classification of brain tumors .Bonnin*et al*. reported two distinct histological types: A low-grade type with better differentiated pattern and favorable postoperative prognosis and a high-grade type showing more anaplastic microscopic features with short postoperative survival. High-grade lesions show focal or multifocal regions of high cellularity, anaplastic nuclear features, elevated mitotic indices, vascular proliferation, and necrosis with pseudopalisading (Bonnin and Rubinstein, 1989).

Our case was considered in High-grade group as it had well orderly growth pattern with no evidence of necrosis and a high mitotic activity. Although malignant astroblastomas may show infiltration of brain parenchyma, most of them are noninfiltrating (Burger and Scheithauer, 1994).

Natural history of astroblastoma seems to place it in between astrocytoma and glioblastoma (Burger and Scheithauer, 1994). Total resection is the aim of surgery (Cabello et al., 1991). Regular follow-up is required even in low-grade variants due to unpredictable behavior. Adjuvant therapy is recommended for high-grade and recurrent cases (Unal et al., 2008). Favorable prognosis is almost invariably associated with well-circumscribed tumors which permit total resection of tumor in all grades. In a series of 23 patients reported by Bonnin and Rubinstein, patients with high-grade astroblastomas who did not receive postoperative radiotherapy had a shorter survival time (Bonnin and Rubinstein, 1989). Caroli*et al*. reported a high-grade astroblastoma with a 5-year survival without recurrence after total resection, radiation therapy, and temozolamide usage (Caroli et al., 2004). Our case was a High-grade astroblastoma, Ourpatient has a rather long free survival time without recurrence. Combination of total excision and radiotherapy seems gratifying with norecurrence detected 14 months after surgery.

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