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CASE REPORT

Parietal pilomyxoid astrocytoma with recurrence in 10 months: A case report and review of literature

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ABSTRACT

Pilomyxoid astrocytoma (PMA) is a new entity described in WHO 2007 classification of brain tumors. Pilocytic astrocytoma (PA) and PMA share many histopathological features with a few differences in histopathology and behavior of the tumor. This tumor is commonly located in the hypothalamic chiasmatic region. PMA behaves more aggressively than PA, with shorter progression-free survival as well as a higher rate of recurrence and CNS dissemination. We describe a case of PMA in a 10-year-old male involving left parietal lobe presenting with raised ICP features along with the follow-up. Patient was symptom free after 7 months of postoperative and 5½ months of post-radiation. The unusual site and atypical Magnetic resonance imaging features are distinctive in this case report.

Key words: Pilocytic astrocytoma, pilomyxoid astrocytoma, recurrence

Introduction

Pilomyxoid astrocytoma (PMA) is a new entity described in World Health Organization (WHO) 2007 classification of brain tumors. Pilocytic astrocytoma (PA) and PMA share many histopathological features with a few differences in histopathology and behavior of the tumor. This tumor is commonly located in the hypothalamic chiasmatic region. We describe a case of PMA in a 10-year-old male involving thalamic region presenting with raised intra cranial pressure (ICP) features with recurrence in 10 months following first surgery. PMA behaves more aggressively than PA, with shorter progression-free survival as well as a higher rate of recurrence and central nervuous system (CNS) dissemination.

Case Report

A 10-year-old boy presented with early morning headache of 2-month duration, difficulty in walking due to weakness of right upper limb and lower limb. On examination, he had right hemiparesis of grade 4/5, upper limb weaker than lower limb, right plantar up going. Contrast-enhanced computerized tomography (CT) [Figure 1] of brain revealed an iso- to hyper-dense irregular, solid and cystic lesion with heterogeneous enhancement on contrast administration with moderate perilesional edema with mass effect and midline shift. Magnetic resonance imaging (MRI) showed large solid cystic T2/FLAIR heterogeneously hyperintense lesion noted in the left parietal region extending from atrium of left lateral ventricle to parietal convexity. Lesion was iso- to hypo-intense on T1 with a few hyperintense areas within, suggestive of hemorrhage with moderate perilesional edema. Cystic component is seen anteroinferior to the solid component of the lesion and hyperintense on T2/FLAIR and hypointense on T1 WI. On contrast administration, there was intense heterogeneously enhancing solid region [Figure 1]. The patient underwent a left parietal craniotomy and gross total excision. Per-operatively, the tumor was soft suckable, grayish white, moderately vascular lesion noted in the left parietal region going deep up to thalamus with compression over the left ventricle. Histopathological examination (HPE) showed prominent myxoid background, cells are arranged in cords, trabeculae, and as perivascular rosettes; focal areas show marked cellular pleomorphism; few cells showed intra-cytoplasmic inclusions. However, biphasic pattern, eosinophilic granular bodies, Rosenthal fibers, calcification, and necrosis were absent. On immunohistochemistry, tumor showed focally positive Glial fibrillary acidic protein (GFAP), strong positivity of epithelial membrane antigen (EMA) with synaptophysin negative, and anti-Ki-67 monoclonal antibody

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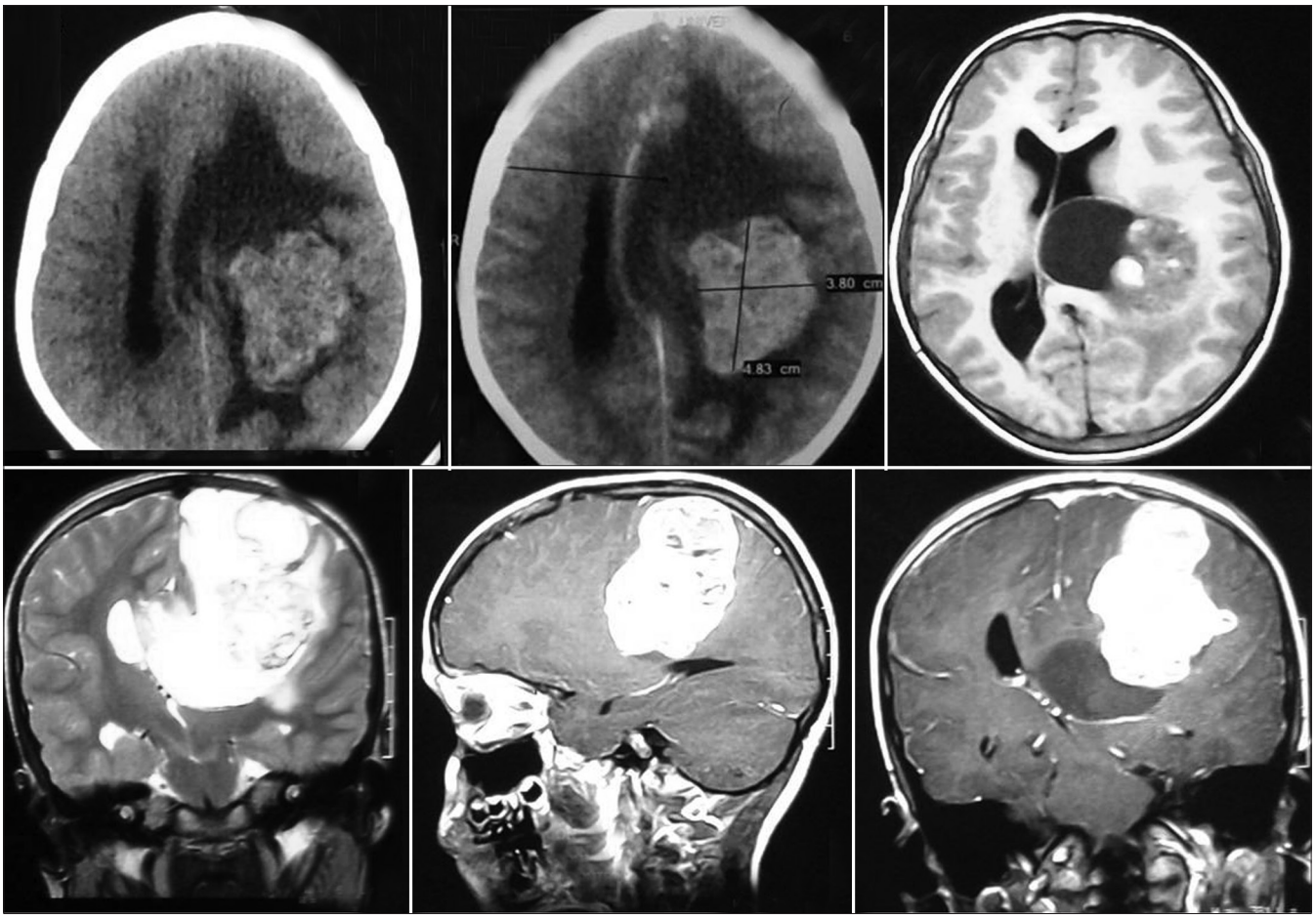

Figure 1: CT scan plain and contrast study, MRI Brain T1WI axial plain section, T2WI coronal plain section, contrast sagittal and coronal sections showing solid cystic lesion in left parietal region extending deep up to the periventricular region

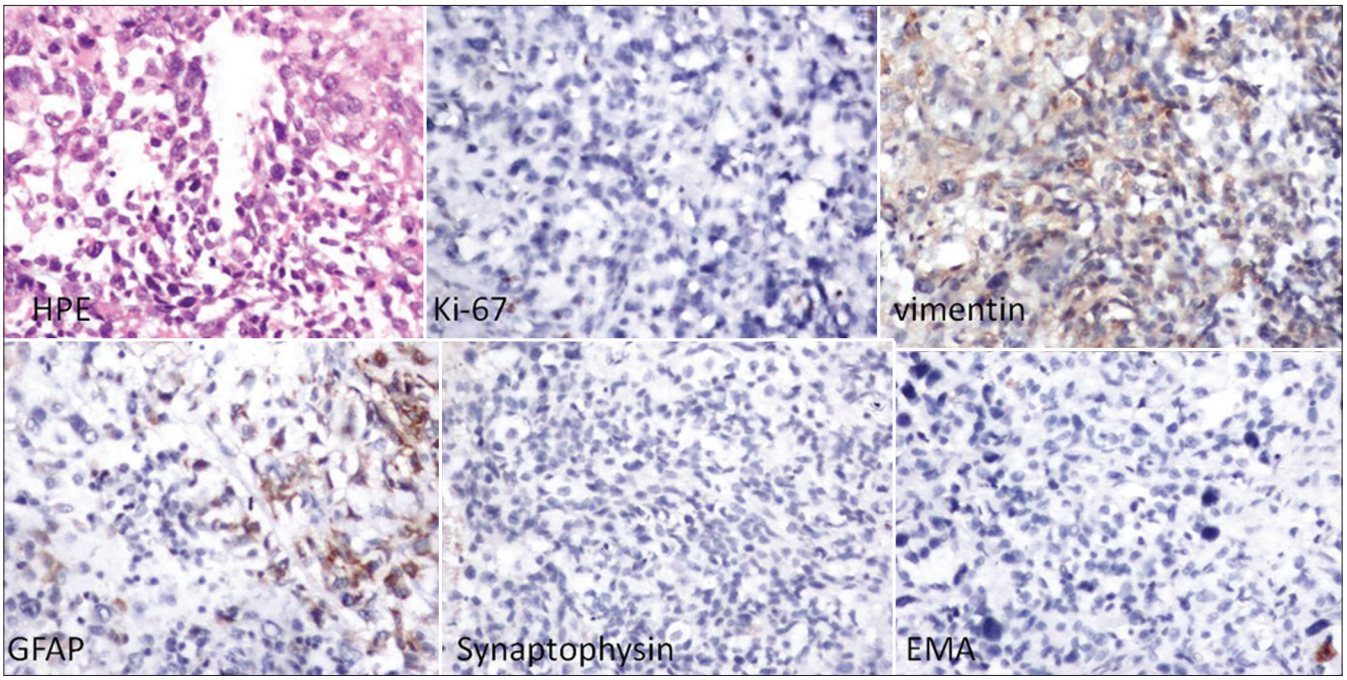

Figure 2: Histopathological study – HPE ×400 section showed prominent myxoid background, cells are arranged in cords, trabeculae, and as perivascular rosettes. The staining index for the anti-Ki-67 monoclonal antibody MIB-1 was 2%. Immunohistochemistry showed focally positive GFAP, strong positivity of EMA, vimentin with synaptophysin negative

MIB-1 staining was 2% suggestive of PMA [Figure 2]. Patient was discharged after suture removal and followed up at regular intervals. 10 months postoperatively, patient developed right hemiparesis and headache of 1-month duration. We investigated with the MRI brain plain and contrast which showed multiple ring-enhancing cystic lesions and solid component suggestive of the recurrence of the lesion [Figure 3]. He was taken up for re-exploration and subtotal excision of the lesion was performed. Histopathology was confirmed as PMA [Figure 4]. Post operative CT immediately following first and second surgery shows gross total excision and subtotal excision. [Figure 5] Repeat staining index for the anti-Ki-67 monoclonal antibody MIB-1 was 12%. The child was relieved of his symptoms and was referred for radiotherapy after suture removal. Patient was symptom free after 7 months of postoperative and 5½ months of post-radiation.

Discussion

PMA was first acknowledged in 1999 as a different one from the PA.[1] PMA is a new entity described in WHO 2007 classification of brain tumors and classified under Grade II.[2] PMA is a pediatric tumor found mostly in hypothalamic chiasmatic region, but also described in diencephalon, posterior fossa, spinal cord, and cerebral hemisphere (right parietal lobe).[3-5] In our case, the tumor is located from the parietal lobe to the atrium of the lateral ventricle. PMA is commonly found in pediatric population but can also be found in any age group.[4,6] Patients usually present with failure to thrive, developmental delay, altered sensorium, vomiting, and generalized weakness along with focal neurological symptoms, visual and endocrine disturbances.[3] In our case, the child presented with headache and right hemiparesis.

On imaging, PMA presents as a homogenously enhancing solid mass in suprasellar region without significant cystic areas or necrosis.[7] Previously reported right parietal lobe PMA showed well-defined tumor with T2 hyper-intensity having low intratumoral ratios of Cho/Cr, with no enhancement on contrast.[5] In our case, the tumor showed a solid and cystic area which was atypical for the PMA. Classically, there will be no perilesional edema in PMA. However, in this report, there is significant amount of perilesional edema with mass effect.

HPE is the main diagnostic tool along with the immunochemistry. HPE of the PMA characteristically shows monomorphous proliferation of bipolar spindle cells in a loose myxoid stroma that will be stained with Alcian blue.[7] Microvascular changes such as telangiectatic clusters in the cyst wall and necrosis without pseudopalisading can be seen.[8] Mitosis and necrosis are rare or absent. Formation of pseudorosettes is also one of the characteristic histological features.[9] PA will have compact biphasic nature demonstrating compact cellular areas with loose cystic regions. However, PMA lacks the biphasic appearance. Eosinophilic granular bodies, Rosenthal fibers, and calcification are uncommon or absent.[1,9] Tumor cells and fibrillary background show immunopositivity for GFAP, EMA, and vimentin.[1,2,10] In our case, the histopathology and immunohistochemistry have the classic features.

Main modality of treatment is gross total resection (GTR) of the tumor. The excision rate depends mainly on the location of the lesion. When the lesion is located in cerebellar hemisphere and cerebral lobes away from the eloquent region, gross total excision is possible. However, if the lesion

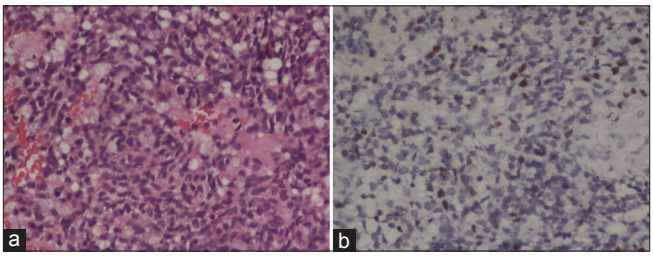

Figure 4: HPE × 400 section and anti-Ki-67 monoclonal antibody MIB-1 staining of the recurrent lesion showed similar findings

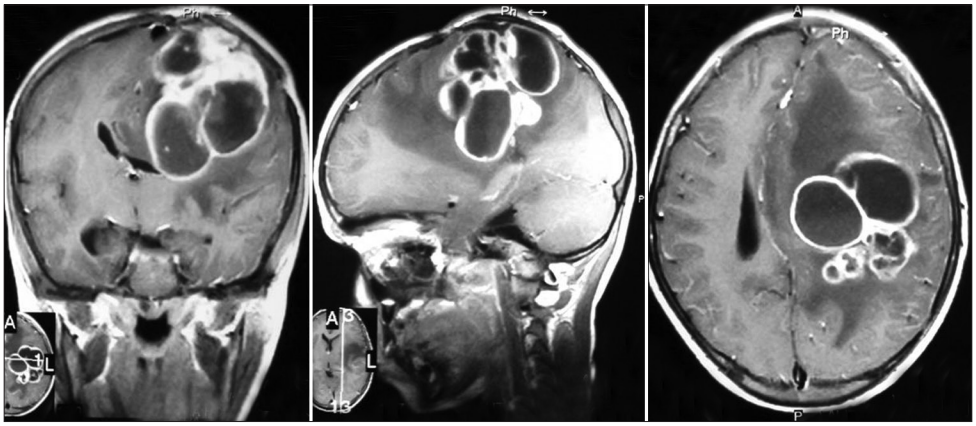

Figure 3: Ten months postoperative MRI Brain contrast study—sagittal, coronal, and axial sections showing recurrent ring enhancing multicystic lesion in the left parietal operative site

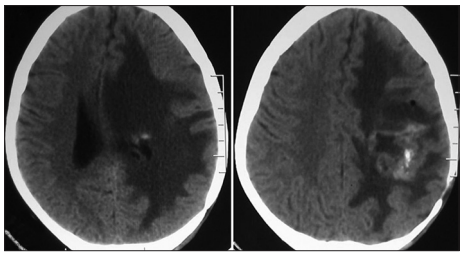

Figure 5: Postoperative CT scans of the patient immediately after 1st surgery and 2nd surgery

is in hypothalamic chiasmatic III ventricle region, fourth ventricle, and other eloquent region, then the excision rate is compromised. The indications for adjuvant therapy may vary as per the patient requirement and can be instituted in at least three situations: (1) tumor recurrence following initial GTR; (2) partially resected tumors that are causing neurological impairment; and (3) partially resected tumors that demonstrate high Ki-67 index or growth on follow-up imaging, even in the absence of symptoms. Chemotherapy is often used now to treat infants and very young children.[3] Adjuvant radiotherapy is generally limited to patients older than 3 to 5 years of age whose disease shows progression.

Patients with PA will have a longer progression-free survival with GTR of the lesion. When compared the hypothalamic or chiasmatic region PA and PMA, the later was associated with shorter overall survival (233 vs. 60 months), shorter progression-free survival (147 vs. 26 months), and higher frequency of recurrence (50% vs. 76%) along with substantial rate of cerebrospinal fluid dissemination.[7,9] Hence, the histopathological and immunohistochemical differentiation of PMA from PA is very important in planning the treatment and to advise the prognosis.

Conclusions

The above case highlights the clinical, radiological, and pathological features of PMA in parietal region extending deep up to the left lateral venricular atrial region. The aggressive nature, less progression-free interval, and high recurrence rate as well as CSF spread needs the differentiation of the pilomyxoid from PA. HPE of the tumor tissue is the mainstay in differentiating the two entities. The unusual site and atypical MRI features are distinct in this case report.

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