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Cerebellar Pilomyxoid Astrocytoma

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Pilomyxoid astrocytomas (PMA) are new class of Pilocytic Astrocytoma (PA.), which typically have their origin in hypothalamus and Chiasmatic region. There are very few case reports of PMAs arising from cerebellum. Their imaging features are similar to PA but they behave more aggressively than PA. To increase awareness of PMA within the neurosurgical community, the authors reviewed a case of 11-year-old male child who presented with truncal and cerebellar ataxia and vomiting and right cerebellar tumor diagnosed as PA radiologically but PMA on histopathology examination. These findings indicate that PMA may be a unique entity that is distinct from PA, or it may be an unusual variant.

Key words: -Pilocytic astrocytoma • pilomyxoid astrocytoma • diagnosis • cerebellar
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Introduction

Pilocytic astrocytomas are generally benign, typically showing 20-year survival rates of 70 to 80%, even when only a subtotal resection is achieved.[1] These highly treatable and potentially curable pediatric tumors are considered Grade I neoplasms according to the World Health Organization tumor classification system.[2, 3]Prior to its recognition, PMA was grouped with PA, because the two display similar histological features.

In 1999, Tihan and colleagues provided the sentinel description of PMAs.[1] Because of their similar histological and radiological features, [2] PMAs were classified as PAs prior to acceptance of this publication. . Earlier reports pointed out unusual features of some pediatric astrocytomas, particularly those within the hypothalamic/chiasmatic region, but did not specifically use a term to distinguish them.[4, 5,9]

In contrast to PAs, PMAs demonstrate a more aggressive clinical course[8-9] and appear to be associated with a higher incidence of leptomeningeal spread.[9] Pilomyxoid astrocytomas are considered to be WHO Grade II neoplasms, and most often arise from the hypothalamic/chiasmatic region. However, PMAs sometimes originate from the posterior fossa.[4]whereas hypothalamic PMAs have been well described in the literature, there is a relative paucity of information about PMAs that arise from the cerebellum. Although limited clinical experience makes it difficult to generate conclusive prognostic data regarding this recently described pediatric tumor, PMA has been shown to behave more aggressively than PA .[11] Here we report a case of cerebellar PMA in a 11 year old male child and discuss literature review

Case report

This was an 11 year old male patient who was relatively in a good state of health prior to his presentation to neurosurgical referral clinics at which time he presented with global headache,projectile vomiting of ingested matter of one moth duration, these symptoms were associated with difficulty of maintaining balance, and blurring of vision.Physical examination revealed papilledema with positive Romberg's test.Abnormal rapid alternating movements, and tandemwalk were indicative of cerebellar ataxia and truncal ataxia. Brain CT scan with contrast was done,

Figure 1: First CT scan done on January 12, 2015 showed a cystic mass at cerebellar vermis measuring 5 by 4 cm which had compressed the 4th ventricle with obstructive hydrocephalus

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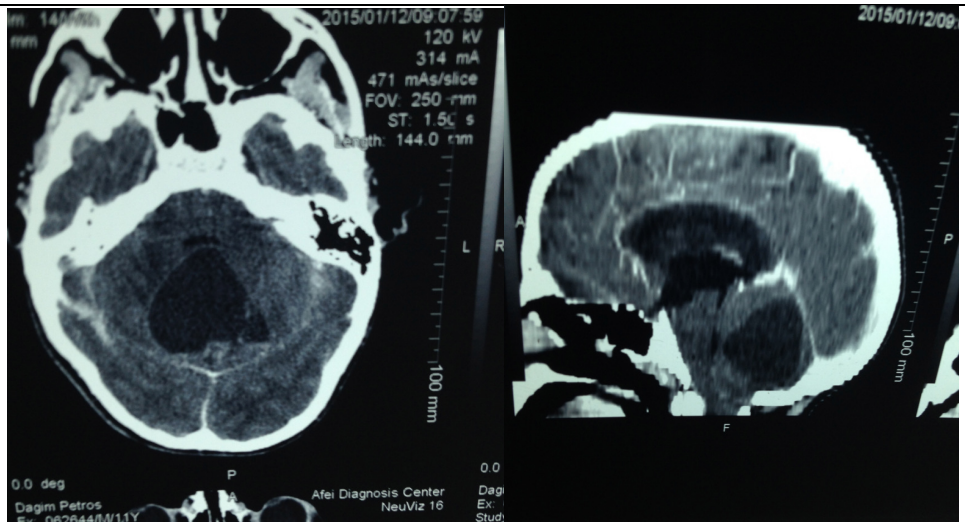
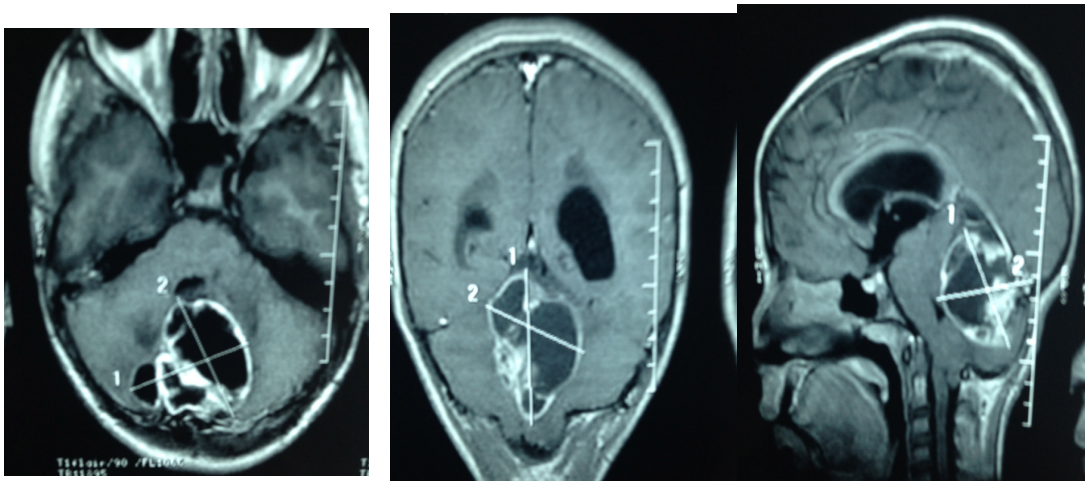


With diagnostic impression of cerebellar Pilocytic astrocytoma, he was operated on 07/05/2015 midline sub-occipital craniectomy and gross total resection of the tumor was done, and patient was discharged with improvement after 10days of hospital stay. Initial biopsy result was Pilocytic Astrocytoma. After discharge patient resumed his daily normal activity and re started his education. On follow-up for 8 months he was perfectly healthy at which time he started to have similar previous symptom such as global headache with difficulty of maintaining balance with frequent vomiting of ingested matter. Physical examination revealed papilledema with positive cerebellar sign.

Figure 2: MRI with contrast of the brain, A-Axial View, B-Coronal View, C Sagittal view: all showing predominantly cystic posterior fossa mass with markedmass effect, protrusion of the mass through the surgical defect posteriorly tonsilar herniationand severe hydrocephalus



With diagnostic impression of recurrent posterior fossa mass with mass effect and severe Hydrocephalus re-do surgery was decided and operative findings are described as follows.

Operation:

Patient under went reoperation on October 30,2015. The patient was placed on the operating table in prone position and in a pin head holder. Midline sub-occipital craniotomy and frizer bur hole done and temporary external ventricular drain was placed in the right lateral ventricle via posterior horn to lower the intracranial pressure by draining CSF.The dura mater did not show any abnormality. Despite the tumor had no capsule, it demarcated clearly with the normal tissues, so that it was not difficult to remove the tumor from the cerebellum. Gross total resection of the tumor was done. Tumor was grayish,firm non-

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suckable mass.

Postoperative period:

The patient started to shoot low-grade fever at 3rdpostoperative day CSF analysis was taken and EVD was removed at 4th day. Cell count was high but culture was negative .He was treated with IV antibiotic with presumptive diagnosis of post operative meningitis and discharged improved after 18 days of hospital stay. Figure 3: shows Postoperative control Brain CT scan image done on 2 Nov 2015. Showing Gross total resection withsmall hematoma and questionable residual.

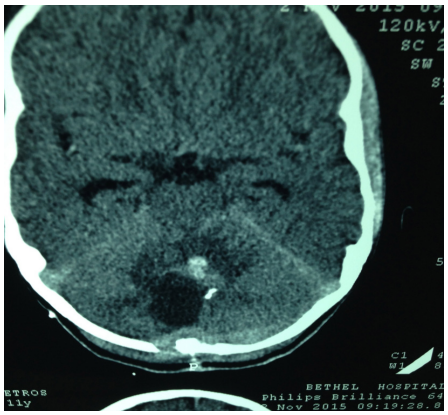

Figure 3. brain CT scan, Axial View, showing postoperative state with small hematoma and with surgical bone defect

Pathology report

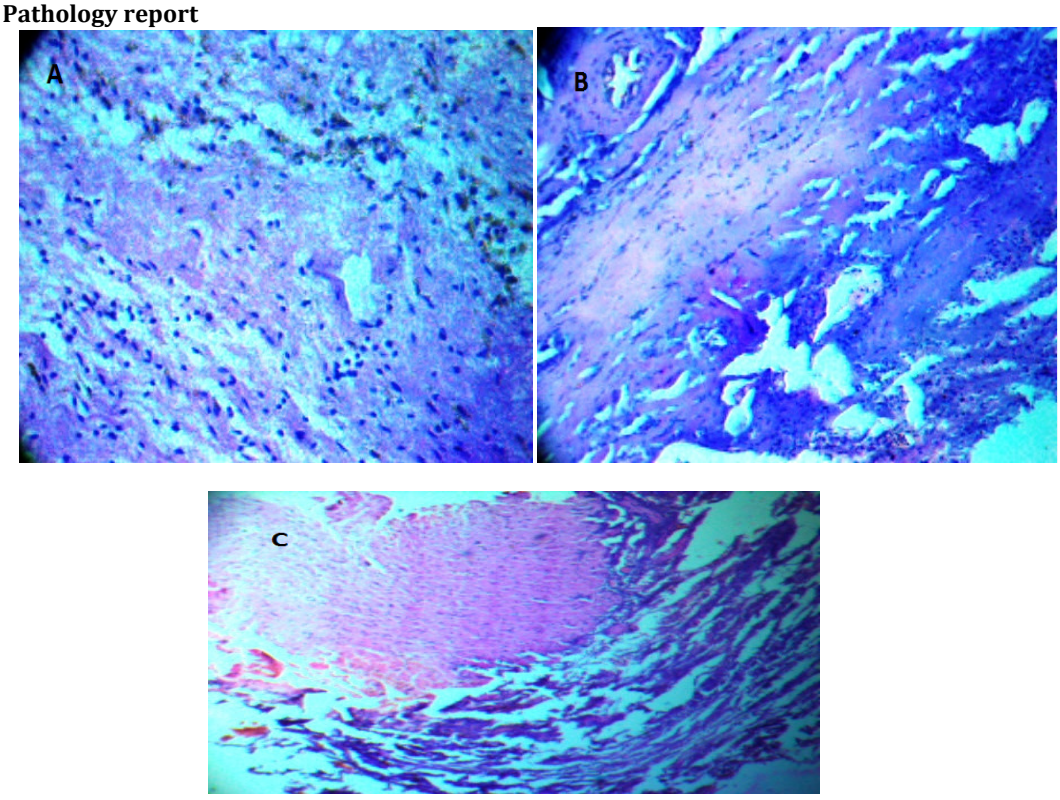

Figure 4. H and E (hematoxylin and eosin stain) 200X. Parallel bundles of bipolar spindle cells with fibrillar cytoplasmic process set in myxoid stroma and aniocentric nature of he tumor cell close to the meninges is also demonstrable.

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Histopathological examination of the specimen from reoperation wasdone on 8 November 2015: formalin fixed paraffin embedded, hematoxylin and eosine stained section demonstrate parallel band of bipolar spindle tumor cell with fibrillary cytoplasmic process set in myxoid stroma in places the tumor cell are arranged near the blood vessels. The angiocentric nature of the tumor cell the myxoid stromal and the proximity of the tumor to the meninges point to the diagnosis of pilomyxoid astrocytoma.

Patient stayed 3 weeks in the hospital and treated for meningitis, finally discharged with marked improvement. Patient was sent to Radiotherapy center for Radiation therapy, but the oncologist decided not to give him radiation treatment. Currently, 4 months postoperatively he become asymptomatic and in a good state of health attending school.

Discussion

PMA have been reported in the English literature and the overwhelming majority of the patients were children aged from 2 months to 4 years.[10]Under the current WHO grading scheme, PAs are designated Grade I and PMAs are designated Grade II neoplasms[8]. Because of similar radiological and histological features, PMAs were considered PAs prior to their sentinel description in 1999.[6] In contrast to the fibrillary background and biphasic cellular pattern observed with PAs, PMAs display a predominantly myxoid background with a perivascular arrangement of tumor cells, reminiscent of ependymomatous pseudorosettes. The myxoid background and angiocentric tumor cell arrangement are typically discernible on a low-magnification examination of H & E–stained material. Examination at high power then reveals a monomorphous population of small bipolar cells and an absence of Rosenthal fibers and Eosinophilic granular bodies.[11] PMAs were typically seen in the chiasmatic-hypothalamic region, but they were also found in other locations, including the spinal cord, the temporal lobe, occipital lobe and sellar-suprasellar region.[12, 13]Histopathological differentiation within the pilomyxoid spectrum is based on morphological analysis of H & E–stained material; the utility of other methods of differentiation—including immunohistochemical staining and molecular analysis—remains speculative at this time. [14,15]

In this case report, the diagnosis of PMA was made predominantly on the basis of histological features after first surgery. Histopathological distinction between PA and PMA is useful because PMAs have been associated with increased rates of local recurrence and leptomeningeal dissemination, as well as decreased overall survival relative to PAs[9]Although it is not uncommon for conventional PAs to spread locally to involve the leptomeninges, the incidence of leptomeningeal dissemination in cerebellar PAs is extremely rare, [17] and many neurosurgeons do not routinely obtain MR imaging of the neuraxis to rule out leptomeningeal dissemination before or after resection.[8] Thus, pathological designation in these patients may influence the frequency and scope of surveillance imaging. Histological features influence the risk-benefit analysis of post resection adjuvant therapy as well. Complete surgical excision of a PA is believed to obviate the need for adjuvant therapy[18]; however, adjuvant chemotherapy has been recommended as initial therapy in the treatment of PMAs.[16]Whereas the literature is replete with clinical series of pediatric patients with hypothalamic/chiasmatic PMAs, descriptions of cerebellar PMAs are rare. In 1 compilation describing the radiological features of 21 patients with PMAs from 7 different institutions, 2 patients (10%) had tumors that arose from the cerebellum.[2] In another compilation of 84 patients with pathological findings of pediatric astrocytomas with pilomyxoid features, 2 patients harbored cerebellar tumors.[15]In our case, the tumor was located in the midline region of the cerebellum, that blocked circulation pathway of cerebrospinal fluid and the initial symptoms were induced by increased intracranial pressure .

On MRI, PMAs also have similar signal patterns to PA, and they both show isointensity on T1WI sequences, hyperintensity on T2WIssequences and on FLAIR images. However, some features could be used to distinguish PMA from PA on MRI. PAs are usually cystic with solid mural nodules and are surrounded by edema. When contrast medium was administered, PA often showed intense enhancement in the nodule or the cyst wall. On the contrary, PMAs are often demonstrates solid or cystic, tend to be well circumscribed, and has little to no calcification rarely with peripheral edema. In PMAs, 40% cases showed homogenous enhancement and 30%-60% cases displayed heterogeneous enhancement. [19,20] Furthermore, PMAs may exhibit peritumotral edema, mass effect and necrosis.

our case, the tumor showed cyst with a mural nodule. Nodule displayed homogenous enhancement while cyst wall was non-enhancing. However, neuroimaging features cannot yet distinguish between PMA and PA, [19] and the diagnosis of PMA is made predominantly on the basis of distinctive histological features[19, 21]

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Similar to Tihan T and collegues our patients with PMA, regardless of total or partial resection, a majority of the postoperative recurrences took place within one year and the local recurrence rate was 76%. [3] The average survival time was 6 months when MRI demonstrated a recurred tumor. [6, 13]

Conclusion

PMA generally exhibits more aggressive biological behavior. Currently, there is no standard of care in treating patients with PMA. Surgical intervention remains the first step, with complete resection being the goal. Given the uncertainties about the prognosis of PMA, it is not possible to provide strict management guidelines at this stage. More definitive guidelines are certain to emerge as clinicians gain more experience with PMA in the future.

Recommendation

Histopathologist should look for features of PMAs whenever they consider the diagnosis of PAs, as this finding is important for the patient and surgeon for

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Prognostication and further treatment especially in cases of recurrence

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