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Cerebellar pilomyxoid astrocytoma

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Abstract: Pilomyxoid astrocytomas (P.M.A) are new class of Pilocytic Astrocytoma (P.A.) 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. The authors report a case of 10 year old male child who presented with right cerebellar tumour diagnosed as PMA on histopathology.

Introduction

Tihan et al, in year 1999, first reported PMA as a new variant of pilocytic astrocytoma. (1) It has been recently defined as a distinct entity among brain tumours and is classified as a WHO grade II glial tumour. (2, 3) PMA may occur anywhere along the neuraxis but they are predominantly present in hypothalamic – chiasmatic region. (3-5) Very few cases of cerebellar PMA are mentioned in literature. (6-8) Herein, we report a case of cerebellar PMA in a 10 year old male child and discuss literature review.

Case report

A 10 year old male child presented with history of unsteadiness of gait and frequent falls for last 3 months. Also, he had developed severe headache and vomiting for last 20 days. Physical examination revealed grade II papilloedema in both eyes with right cerebellar signs.

Imaging: NCCT head revealed a large solid-cystic mass in right cerebellum pushing the IV ventricle to the opposite side with hydrocephalus (Figure 1). The tumour was heterogeneously iso to hypo intense on T1WI (Figure 2A). DWI revealed anterior cystic and posterior solid tumor (Figure 2B). Solid portion was hyper intense on T2WI (Figure 2C) and heterogeneous contrast enhancement was seen on gadolinium administration. The cyst wall was nonenhancing (Figure 2D).

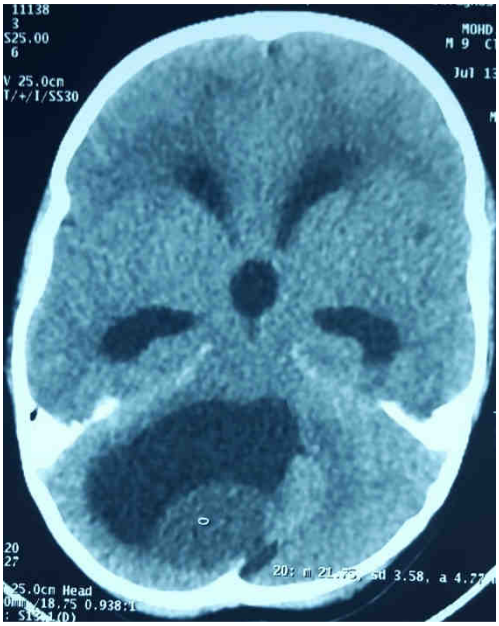

Figure 1 - CT image shows solid cystic lesion in right cerebellum with hydrocephalus

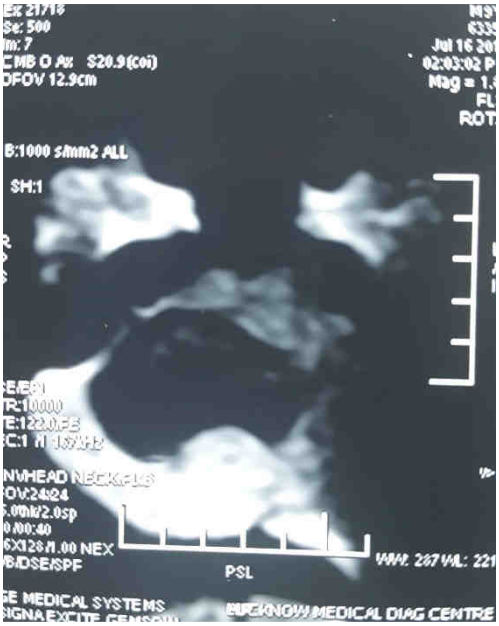

Figure 2 B - Diffusion weighted image (DWI) revealing anterior cystic and posterior solid portion

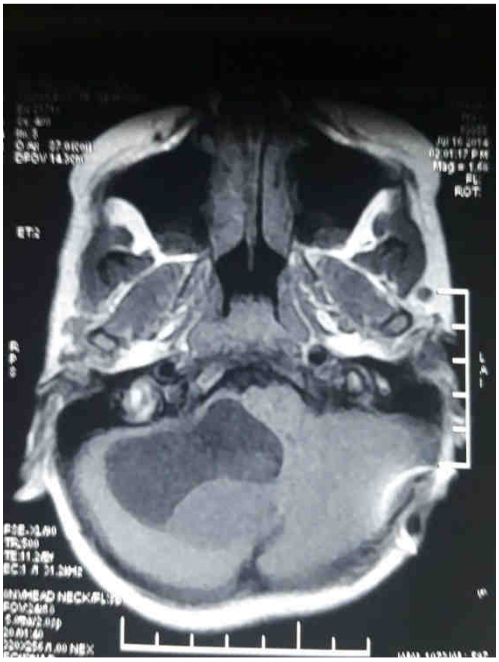

Figure 2 A - T1WI showing iso to hypointense lesion

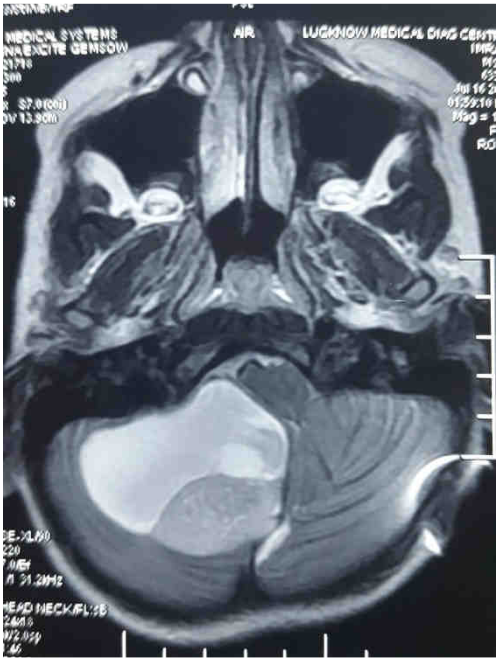

Figure 2 C - Gadolinium enhancement present in solid portion

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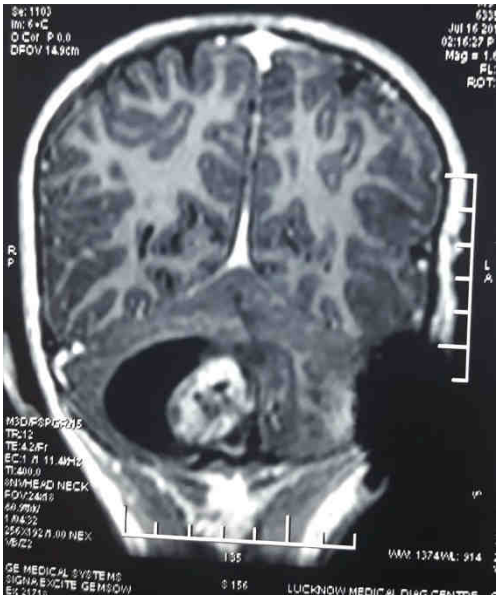

Figure 2 D - Non-enhancing cystic portion on Gadolinium administration

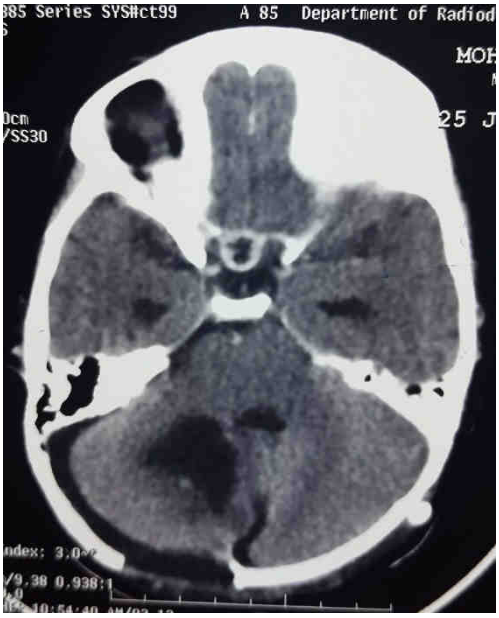

Figure 3 - CT image showing gross total excision of tumour

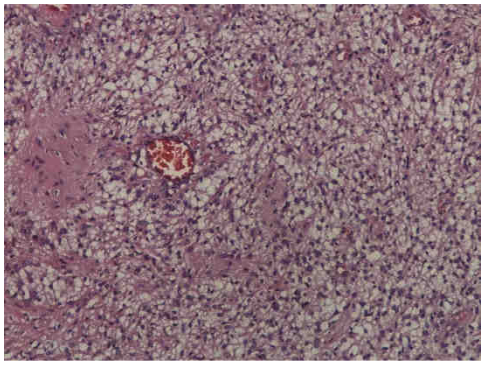

Figure 4 - Section shows tumor cells displaying monomorphic nuclei with piloid morphology with perivascular arrangement of cells with myxiod areas. (H&E X200)

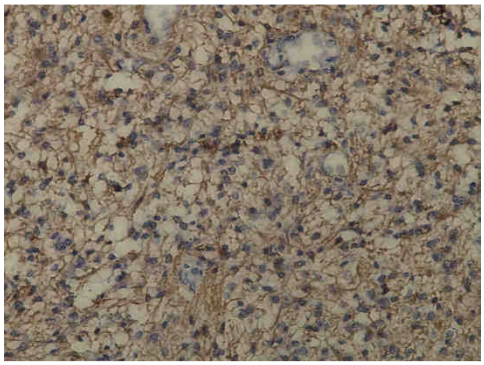

Figure 5 - IHC GFAP: tumor cells display diffuse cytoplasmic expression of GFAP

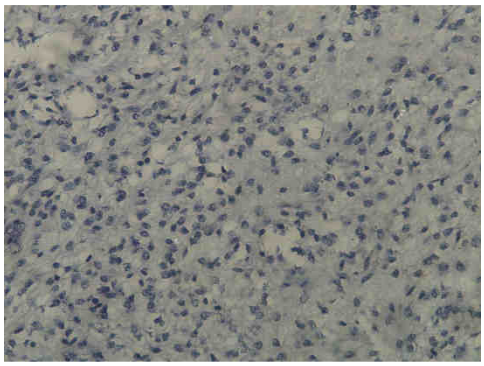

Figure 6 - IHC Ki67: Ki67 expression is low < 1%

Operation: Patient underwent left sided ventriculo peritoneal shunt done to relieve raised intracranial pressure and further deterioration. After one week, Right retromastoid suboccipital craniectomy was done with gross total excision of tumor (Figure 3). Tumour was greyish pink, moderately vascular, non suckable mass with clear cystic fluid.

Post operative: The post operative period was uneventful. Headache and vomiting were relieved immediately after shunt surgery. Child was discharged on 8th postoperative day.

Histopathological examination revealed it a case of pilomyxoid astrocytoma. In view of this report patient received 40 Gray of radiation to whole brain with 14 Gray boost to tumour bed. During follow up, his gait improved within 2 months. After one year of follow up, patient remained asymptomatic without any recurrence on contrast CT.

Histopathology: Haematoxylin and eosin stained section displayed a tumor composed of piloid cells with monomorphic nuclei. Cells were converged near blood vessels in myxoid background. Rosenthal fibres and eosinophilic bodies were not observed (Figure 4). These tumor cells displayed diffuse cytoplasmic immunoexpression for Glial Fibrillary Acidic Protein (GFAP) (Figure 5) with low Ki-67 index; less than 1% (Figure 6). Neuronal markers like Synaptophysin and neuron-specific enolase were negative.

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. (9) 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. (10-12, 19) Recent reports have indicated that PMAs can occur in adults; a tumor in the amygdala/uncus region has been reported in a 28-year-old man, (13) and a spinal cord PMA has been reported in a 45-year-old woman. (14) The case of a 25-year-old man with a fourth ventricular PMA has also been reported. (15) PMA in cerebellum were reported from 2-12 years as in our case whose age was 10 years.

PMA has some specific features in histology, which include monomorphous growth of piloid cells with an angiocentric pattern, being rich in myxoid background, and lacking of Rosenthal fibers or eosinophilic granular bodies. (16) Similarly, PAs also consist of piloid cells, but these cells grow in a biphasic pattern, mixed with Rosenthal fibers and eosinophilic granular bodies. (16) Occurrence of necrosis is more commonly associated with PMA, while cyst formation, calcification and perilesional edema are more common in classic PA. (4) In immunohistochemical studies, PMA stains strongly positive for glial fibrillary acidic protein and vimentin, and is negative for neuronal markers. (2) In our case tumor tissue was diffusely positive for GFAP and negative for neuron-specific enolase.

On MRI, PMAs also have similar signal patterns to PA, and they both show isointensity on T1W sequences, hyperintensity on T2W sequences and on FLAIR images. However, some features could

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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 solid, rarely with peripheral edema. In PMAs, 40% cases showed homogenous enhancement and 30%-60% cases displayed heterogeneous enhancement. (4, 17) In 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, (4) and the diagnosis of PMA is made predominantly on the basis of distinctive histological features. (2, 4)

For the 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%. (18) The average survival time was 6 months when a recurred tumor was demonstrated by MRI. (1, 12) Our patient remained asymptomatic without any recurrence on contrast CT at one year of follow up.

Conclusion

Histopathologist should look for features of PMAs whenever they consider the diagnosis of PAs as this finding is important for patient and surgeon for prognostication and further treatment especially in cases of recurrence.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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References

1.Tihan T, Fisher PG, Kepner JL, et al. Pediatric astrocytomas with monomorphous pilomyxoid features and a less favorable outcome. J Neuropathol Exp Neurol 1999;58:1061–8.

2.Brat DJ, Scheithauer BW, Fuller GN, et al. Newly codified glial neoplasms of the 2007 WHO classification of tumors of the central nervous system: angiocentric glioma, pilomyxoid astrocytoma and pituicytoma. Brain Pathol 2007;17:319–24.

3.Komator RJ, Mocco J, Carson BS, et al. Pilomyxoid astrocytoma: a review. Med Gen Med 2004;6:42.

4.Komakula ST, Fenton LZ, Kleinschmidt-DeMasters BK, Foreman NK: Pilomyxoid astrocytoma: neuroimaging with clinicopathologic correlates in 4 cases followed over time. J Pediatr Hematol Oncol 29:465–470, 2007

5.Komotar RJ, Mocco J, Jones JE, Zacharia BE, Tihan T, Feldstein NA, et al: Pilomyxoid astrocytoma: diagnosis, prognosis, and management. Neurosurg Focus 18(6A):E7, 2005

6.Ge PF, Wang HF, Qu LM, Chen B, Fu S, Luo Y. Pilomyxoid astrocytoma in cerebellum .Chin J Cancer Res. 2011 Sep;23(3):242-4.

7.Ajani OA, Al Sulaiti G, Al Bozom I. Pilomyxoid astrocytoma of the cerebellum. J Neurosurg Pediatr. 2011 May;7(5):539-42.

8.Forbes JA, Mobley BC, O'Lynnger TM, Cooper CM, Ghiassi M, Hanif R, Pearson MM. Pediatric cerebellar pilomyxoid-spectrum astrocytomas. J Neurosurg Pediatr. 2011 Jul;8(1):90-6.

9.Morales H, Kwock L, Castillo M. Magnetic resonance imaging and spectroscopy of pilomyxoid astrocytomas:

case reports and comparison with pilocytic astrocytomas. J Comput Assist Tomogr 2007; 31:682-7.
10.Linscott LL, Osborn AG, Blaser S, et al. Pilomyxoid astrocytoma: expanding the imaging spectrum. AJNR Am J Neuroradiol 2008; 29:1861-6.
11.Omura T, Nawashiro H, Osada H, et al. Pilomyxoid astrocytoma of the fourth ventricle in an adult. Acta Neurochir (Wien) 2008; 150:1203-6.
12.Buccoliero AM, Gheri CF, Maio V, et al. Occipital pilomyxoid astrocytoma in a 14-year-old girl--case report. Clin Neuropathol 2008; 27:373-7.
13.Komotar RJ, Mocco J, Zacharia BE, Wilson DA, Kim PY, Canoll PD, et al: Astrocytoma with pilomyxoid features presenting in an adult. Neuropathology 26:89–93, 2006.
14.Sajadi A, Janzer RC, Lu TL, Duff JM: Pilomyxoid astrocytoma of the spinal cord in an adult. Acta Neurochir (Wien) 150:729–731, 2008
15.Omura T, Nawashiro H, Osada H, Shima K, Tsuda H, Shinsuke. A: Pilomyxoid astrocytoma of the fourth ventricle in an adult. Acta Neurochir (Wien) 150:1203–1206, 2008
16.Koeller KK, Rushing EJ. From the archives of the AFIP: pilocytic astrocytoma: radiologic-pathologic correlation. Radiographics 2004; 24:1693-708
17.Arslanoglu A, Cirak B, Horska A, et al. MR imaging characteristics of pilomyxoid astrocytomas. AJNR Am J Neuroradiol 2003; 24:1906-8.
18.Tsugu H, Oshiro S, Yanai F, et al. Management of pilomyxoid astrocytomas: our experience. Anticancer Res 2009; 29:919-26.
19.Alimohamadi M, Bidabadi MS, Ayan Z, Ketabchi E, Amirjamshidi A. Pilomyxoid astrocytoma with involvement of the sella turcica in an adolescent. J Clin Neurosci. 2009 Dec; 16 (12):1648-9.