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Case Report

A pediatric case of anaplastic pleomorphic xanthoastrocytoma (Grade 3)

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ABSTRACT

Introduction: Pleomorphic xanthoastrocytoma (PXA) is a glial tumor which accounts for less than 1% of astrocytomas. PXA most commonly occurs in the temporal lobe in the outer cortex of brain and usually seen in young adults and children. As per 2020 WHO classification of CNS tumors, PXA with a mitotic activity ≥5 mitoses/10 HPF is defined as anaplastic pleomorphic xanthoastrocytoma (APXA), WHO grade 3. Here we present a rare case of Anaplastic Pleomorphic Xanthoastrocytoma developing in a female child of 11 years of age.

Case Details: A 11 years old female child presented with symptoms of continuous headache, giddiness, fever and vomiting since last 15 days. Computed Tomography of her brain revealed a soft tissue density mass lesion in right temporal lobe involving the insular cortex and capsuloganglionic region. The radiological differential diagnosis was a neoplastic lesion, most likely glioma. The patient underwent surgery for removal of temporal lobe tumor and the resected tumor bits were sent for histopathological assessment. Microscopically, many multinucleated lipidized giant cells were seen having eccentrically pushed nuclei, conspicuous nucleoli and abundant eosinophilic granular cytoplasm. A second population of few spindled cells with less amount of cytoplasm was also noted in the intervening parenchyma. Immunohistochemical workup showed the tumor to be positive for GFAP, S-100, Synaptophysin, p53 and CD-68. The tumor was negative for CD34. Ki-67 index was 15-20%. A final diagnosis of Anaplastic Pleomorphic Xanthoastrocytoma (grade 3) was made.

Conclusion: APXA is a rare astrocytic tumor and has relatively more aggressive radiological and histomorphological features than pleomorphic xanthoastrocytomas and therefore it needs to be identified and treated separately.

1. Introduction

Pleomorphic xanthoastrocytoma (PXA) is a glial tumor which accounts for less than 1% of astrocytomas. PXA most commonly occurs in the temporal lobe in the outer cortex of brain and usually seen in young adults and children.[1,2] PXA manifests itself first as seizures followed by focal neurological deficits. According to WHO classification for astrocytic neoplasms, it has been classified histologically as a grade 2 (benign) neoplasm. As per 2020 WHO classification of CNS tumors, PXA with a mitotic activity ≥5 mitoses/10 HPF is defined as anaplastic pleomorphic xanthoastrocytoma (APXA), WHO grade 3. APXA has poor prognostic outcome in comparison to pleomorphic xanthoastrocytoma (PXA). APXA is usually seen in the second decade and more rarely in the first and third decades of life.[3,4] In this case report, we present a case of Anaplastic Pleomorphic Xanthoastrocytoma (grade 3) developing in a female child of 11 years of age.

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2. Case Report

A 11 years old female child presented in Neurology OPD with symptoms of continuous headache, giddiness, fever and vomiting since last 15 days. No significant medical or surgical history was found. Physical examination of the patient was unremarkable with intact cranial nerves.

Computed Tomography of her brain revealed a soft tissue density mass lesion in right temporal lobe involving the insular cortex and capsuloganglionic region along with edema in adjacent parenchyma. There was associated effacement of right lateral ventricle as well. The radiological differential diagnosis was a neoplastic lesion, most likely glioma.

The patient was referred to Neurosurgery department and underwent surgery for removal of temporal lobe tumor. The resected tumor bits were sent for histopathological assessment.

Microscopically, the tumor was consistent with a glial neoplasm with variable morphology. Many multinucleated lipidized giant cells were seen having eccentrically pushed nuclei, conspicuous nucleoli and abundant eosinophilic granular cytoplasm. A second population of few spindled cells with less amount of cytoplasm was also noted in the intervening parenchyma. Few dense eosinophilic bodies were also seen along with prominent fibrillary processes and mitotic figures. Large areas of haemorrhage with thrombosed blood vessels were observed as well. There were no foci of microvascular proliferation or tumor necrosis.

Tumor was found to be positive for GFAP, S-100, Synaptophysin and CD-68 and negative for CD34 on immunohistochemistry. Ki-67 index was 15-20%. Based on the clinical history, typical microscopic findings and immunohistochemistry findings, a final diagnosis of Anaplastic Pleomorphic Xanthoastrocytoma Grade 3 was made. The tumor was excised completely and the patient was referred for radiotherapy.

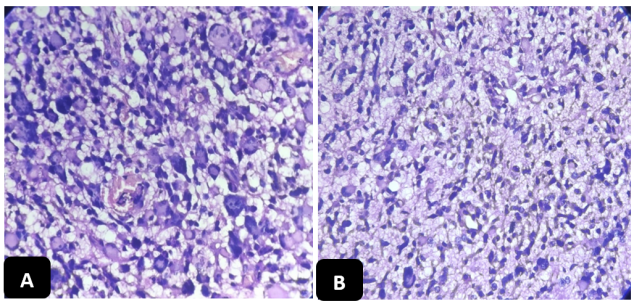

Fig. 1: A,B) Sections showing glial neoplasm with many multinucleated lipidized giant cells having eccentrically pushed nuclei, conspicuous nucleoli and abundant eosinophilic granular cytoplasm. (Hematoxylin and Eosin x400)

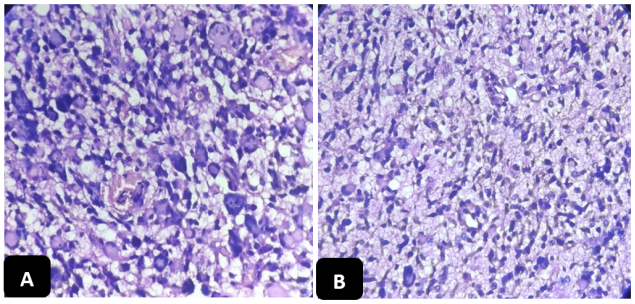

Fig. 2: A,B) Sections showing a second population of spindled cells with less amount of cytoplasm in the intervening parenchyma. Few dense eosinophilic bodies are also seen alongwith prominent fibrillary processes and mitotic figures. (Hematoxylin and Eosin x400)

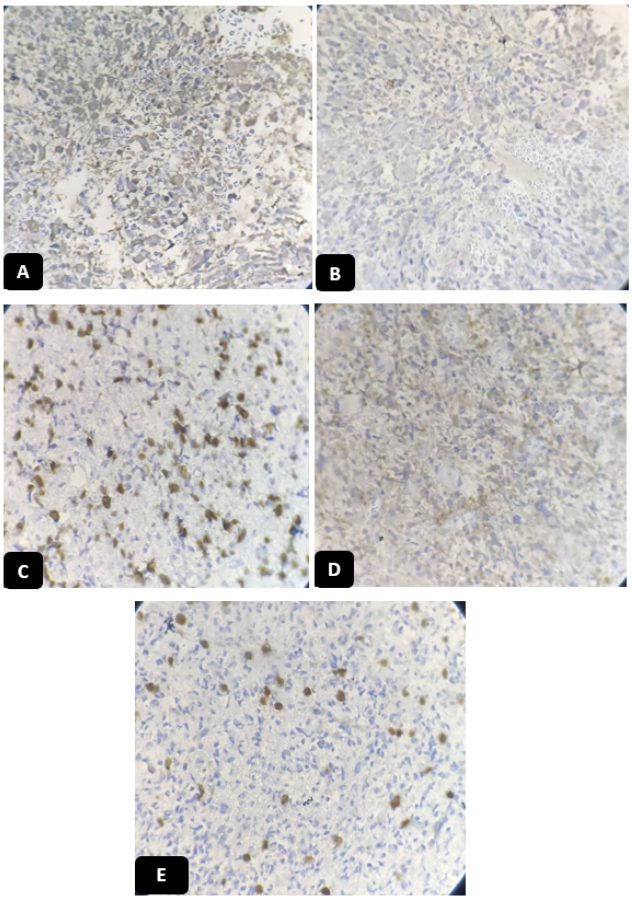

Fig. 3: Immunohistochemical findings; **A**): GFAP positive; **B**): S100 positive; **C**): Synaptophysin positive; **D**): CD68 positive; **E**) Ki67 Index= 15-20%

3. Discussion

PXA is a glial tumor which accounts for less than 1% of astrocytomas. The most common site of PXA is the temporal lobe in the outer cortex. The lesions are usually well demarcated solid or cystic masses. The most common clinical symptom seen is epilepsy which usually arise in the second decade.[5] It is hypothesised that PXA arises from neuroepithelial cells.[6] As per 2020 WHO classification of CNS tumors, PXA with a mitotic activity ≥5 mitoses/10 HPF is defined as anaplastic pleomorphic xanthoastrocytoma (APXA), WHO grade 3. The site, histomorphology, and immunohistochemical markers of APXA are same as those of PXA. Therefore, the cut-off value for mitotic activity of ≥5 mitoses/10 HPF is important to differentiate between the above two entities. APXA usually arise from malignant transformation of PXA.[7]

Histopathological features of APXA include highly pleomorphic, xanthomatous tumour cells with intracytoplasmic lipid vacuoles along with many multinucleated giant cells. There is increased mitotic activity (≥5 mitoses/10 HPF). Cellular atypia with hyperchromatism and nuclear irregularities may be seen.[8] Necrosis is usually absent but can be rarely seen. The closest differential diagnosis of APXA include Glioblastoma, most common of which are epithelioid and giant cell Glioblastoma, due to the reason that they can display the same features as APXA. Palisading necrosis and endothelial microvascular proliferation are absent in APXA which help in differentiating these tumours from most of glioblastomas.

Molecular analysis of PXA cases show BRAF gene mutations, most commonly BRAF V600E mutations. Compared with PXA, APXA has a lower BRAF mutation rate and therefore differentials of APXA include other gliomas which also harbor BRAF V600E mutations such as pilocytic astrocytoma.

APXA can show transformation to glioblastoma and typically has a poor prognosis.[9] The treatment of choice is surgical excision and post-surgical radiotherapy. Though complete surgical resection is quite effective in benign PXA without any recurrences, it is found that total surgical excision in APXA is still associated with frequent recurrences. Chemotherapy is usually not effective in APXA.[10] It has been observed that long-term control can be achieved with surgical resection and stereotactic radiation therapy.[11]

4. Conclusion

APXA is a rare astrocytic tumor and has relatively more aggressive radiological and histomorphological features than pleomorphic xanthoastrocytomas and therefore it needs to be identified and treated separately. Radiological, histopathological and immunohistochemical features along with clinical correlation can help in correct diagnosis and further treatment planning.

5. Source of Funding

None.

6. Conflict of Interest

None.

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