ISSN No:-2456-2165

Astroblastoma: Case Report and Review of Literature

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Abstract:-
Background: Astroblastomas are regarded as uncommon neuroepithelial tumors with an uncertain origin. These tumors primarily affect young adults and children, mainly in the cerebral hemisphere. When it comes to neuroglial tumors, they represent a small percentage, ranging from 0.45% to 2.8%. Because of their rarity in clinical practice and their resemblance to other glial neoplasms in terms of radiological and histopathologic appearance, astroblastomas can be prone to misdiagnosis.
Case presentation: We report a 38-year-old female patient who presented with vomiting and left-sided weakness. Following a brain MRI, we observed a moderate-sized area of T2 heterogeneous hyperintensity and T1 isointensity acute intraparenchymal bleed with mild surrounding edema in the right high fronto-parietal region. The dimensions of the bleed measured approximately 4.1x3.9x3.4 cm. The MRI also revealed some areas of mild diffusion restriction and faint inhomogeneous areas of enhancement upon post-contrast imaging. Furthermore, there was evidence of mild mass effect on the right Posterior cingulate gyrus and a thin streak of flair hyperintense subarachnoid bleed. To address this condition, the patient underwent near-total resection of the lesion and evacuation of the clot through right fronto-parietal craniotomy. The diagnosis of low-grade astroblastoma was confirmed through histopathological examination combined with immunohistochemical study.
Conclusion: Astroblastoma is an extremely rare primary brain tumor. Its diagnosis often poses challenges due to the astroblastic aspects found in astrocytic tumors, ependymomas, and non-neuroepithelial tumors. The histogenesis and classification of this tumor are surrounded by considerable confusion. Conducting studies to investigate tumor characteristics is difficult due to its low incidence rate.

I. INTRODUCTION

Astroblastomas are uncommon neuroepithelial tumors [1].The incidence of this condition is relatively low, although there is a lack of reliable estimates. It is appropriate to estimate that the incidence falls below 1% of primary brain tumours. Based on numerous case series, it is consistently observed that females are more frequently affected by this condition compared to males [2]. Additionally, it is important to note that this tumour primarily affects individuals during childhood and young adulthood [2].

II. CASE PRESENTATION

A 38 old female presented to us with vomitings followed by left sided weakness(Left upper limb weakness 3/5 and Lower limb 4/5),rest no defecits. Brain magnetic resonance imaging (MRI) revealed moderate sized area of T2 heterogeneous hyperintensity and T1 isointensity acute intraparenchymal bleed with mild surrounding edema seen in right high fronto-parietal region showing significant blooming on susceptibility weighted images, approximately measuring 4.1 x 3.9 x 3.4 cm (AP× TR×CC). Interspersed areas of mild diffusion restriction. Post-contrast images show faint inhomogeneous areas of enhancement in mild right cerebral hemispheric. Mild mass effect on right Posterior cingulate gyrus. Thin streak of FLAIR hyperintense subarachnoid bleed seen along right posterior frontal sulci.MRI reported as Neoplastic lesion with tumour bleed.

The patient underwent a gross total resection of the lesion through a right fronto parietal free bone flap craniotomy and evacuation of the clot. The tumor was found to be rubbery, vascular, and well demarcated from the surrounding brain. The postoperative period was without any complications, and the neurological examination showed improvement in motor power . The histopathologic diagnosis revealed a low-grade astroblastoma. Under the microscope, the section displayed a predominantly circumscribed glial neoplasm composed of tumor cells arranged in a pseudopapillary, perivascular pattern. These cells had eosinophilic cytoplasm and short stub-like processes extending to a central blood vessel. The presence of thrombosed and sclerosed vessels, as well as areas of hemorrhage, were also noted. The immunoprofile confirmed the diagnosis with positive results for GFAP, Synaptophysin, and focal EMA, while p-53 showed 0-2% positivity and Ki-67 showed 10-12% positivity. The neoplasm was classified as suggestive of Astroblastoma, although the WHO Grade has not been assigned. The patient was advised regular followup without adjuvant radiotherapy. To date, there has been no recurrence of the tumor as confirmed during the last follow-up done 6 months after the surgery.

ISSN No:-2456-2165

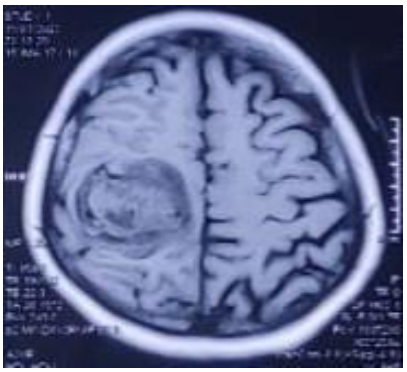
Fig 1: T1W axial image.

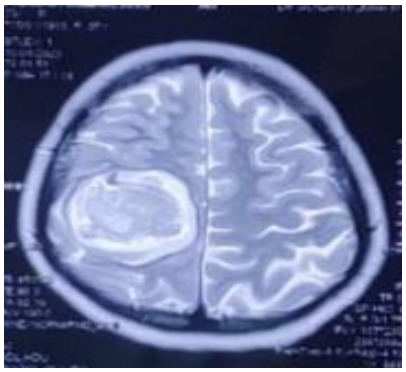
Fig 2: T2W axial image.

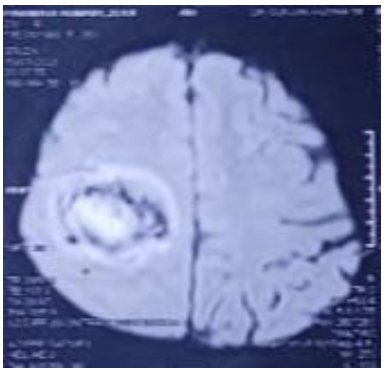
Fig 3: T2 flair axial image

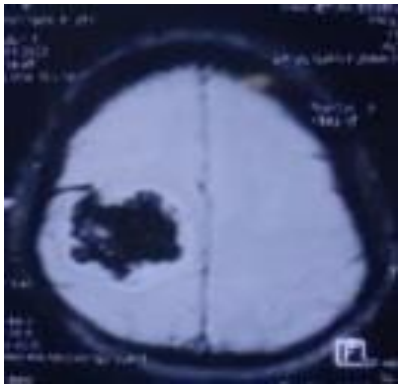
Fig 4: SWI axial image

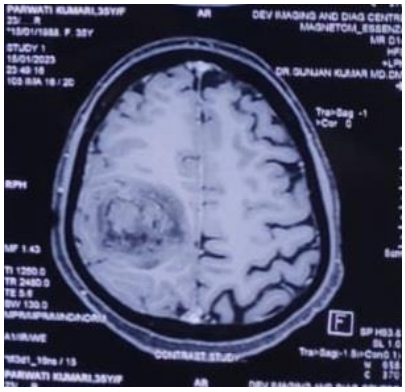
Fig 5: T1W axial Post-contrast image

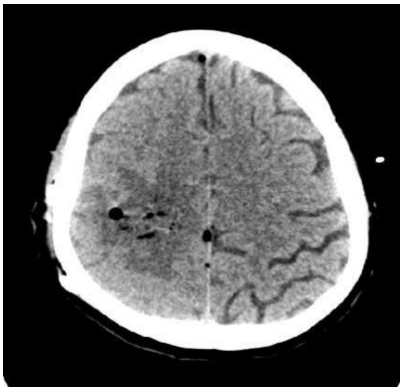
Fig 6: Post op CT scan axial.

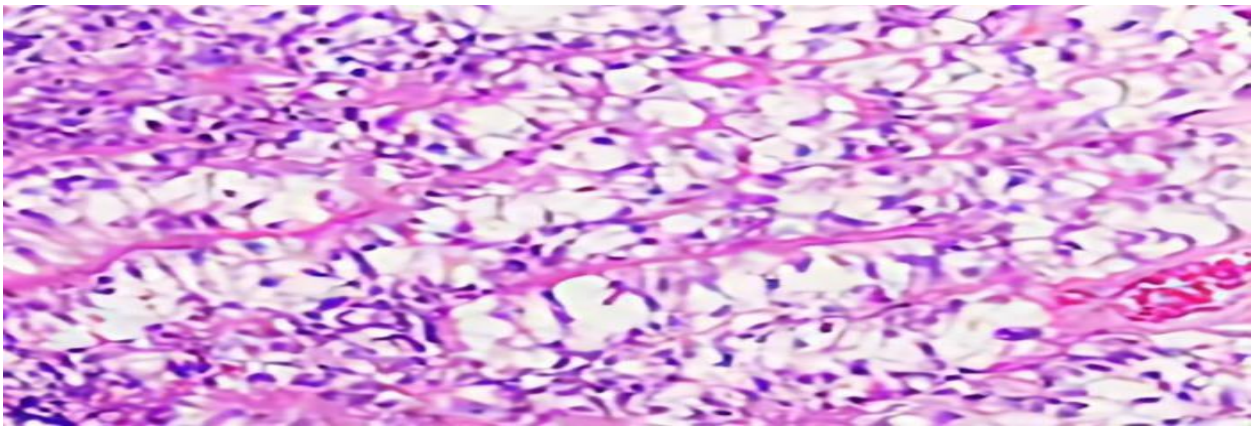
Fig 7: Histopathologic slide – High-power field pseudorosettes around central hyalinized thickened blood vessels with individual cells being polygonal to spindled, showing moderate eosinophilic cytoplasm and eccentrically placed nuclei

III. DISCUSSION

Astroblastoma is a rare tumor originally described by Bailey and Cushing [15] that has been the subject of debate, but it maintains its status as part of the 2021 World Health Organization (WHO) classification. Its histological features are described as a glial neoplasm characterized by the presence of GFAP positive astrocytic cells with distinct radiating processes towards a central blood vessel. Typical features include perivascular pseudorosettes and prominent perivascular hyalinization. The tumors show strong positive staining for S-100, glial fibrillary acid protein (GFAP), and vimentin. Based on nuclear atypia, cellularity, and mitotic figures, astroblastomas are categorized as either low-grade or high-grade. Necrosis can be present in any histological type. While the clinical significance of the grading of these tumors is not completely understood, anaplastic histology has been found to be a poor prognostic feature with higher recurrence rates in larger studies. Surprisingly, rapid recurrences have been reported in low-grade astroblastoma. It is important to distinguish these tumors from ependymomas, astrocytomas, and certain non-neuroepithelial tumors [3,4,5]. Recurrent lesions often show increased anaplasia and can be misdiagnosed as glioblastoma or gliosarcoma.

In light of limited comprehensive studies on the pathogenesis of these lesions, there is anecdotal evidence suggesting potential factors. One study examined chromosomal abnormalities in seven cases using classical comparative genomic hybridization[5]. Results showed that the most common alterations were gains in chromosome arm 20q (four out of seven cases) and 19 (three out of seven cases). There were also two cases[6] with a gain in chromosome 9p and one case with a loss of heterozygosity in chromosome 9.

The incidence of astroblastomas is relatively low, although reliable estimates are lacking. It is reasonable to assume that the incidence is below 1% of primary brain tumors. Multiple case series[2] consistently show that females are more commonly affected than males, as observed in this case. These tumors are primarily found in the cerebral hemispheres, particularly the frontal or parietal lobes, but can also occur in the occipital or temporal lobes. They have also been reported in the brainstem, intraventricular compartment, and spinal cord[3,7].

The clinical presentation[7] of astroblastoma varies depending on location and growth rate, making it nonspecific. Some patients may have symptoms that do not clearly indicate the affected area, possibly suggesting increased intracranial pressure. Others may experience focal deficits or seizures.

Bell et al. [8] have recently conducted an imaging series consisting of 12 cases of astroblastomas, making it the largest study on this topic. According to their findings, astroblastomas are predominantly observed in supratentorial regions and tend to be located at the periphery. These tumors typically present with both solid and cystic components. In our case, we observed moderate sized area of T2 heterogeneous hyperintensity and T1 isointensity acute intraparenchymal bleed with mild surrounding edema seen in right high fronto-parietal region showing significant blooming on susceptibility weighted images, with interspersed areas of mild diffusion restriction. Post-contrast images show faint inhomogeneous areas of enhancement in mild right cerebral hemispheric.Thin streak of FLAIR hyperintense subarachnoid bleed seen along right posterior frontal sulci. However, in contrast to high-grade tumors, astroblastomas generally exhibit less perilesional edema, including in high-grade variants.

Astroblastomas, gliomatosis cerebri, and polar spongioblastoma are classified as neuroepithelial tumors of uncertain origin. In the 2007 WHO classification of brain tumors, these tumors are considered grade 4, in contrast to grade 1. Bonnin et al [3]. conducted a study and identified two distinct histological types: a low-grade type with a well-differentiated pattern and a favorable prognosis after surgery, and a high-grade type with more anaplastic microscopic features and shorter postoperative survival. High-grade lesions exhibit areas of high cellularity, anaplastic nuclear characteristics, increased mitotic indices, vascular proliferation, and necrosis with pseudopalisading.

Our case was placed in the low-grade group due to its well-organized growth pattern and absence of necrosis, accompanied by a high mitotic activity. It is important to note that while certain malignant astroblastomas may exhibit infiltration into the brain parenchyma, the majority of cases are noninfiltrating.

Radical resection [9] is considered the preferred treatment for these typically non-infiltrative, well-defined, and externally located lesions. Due to these characteristics, most of these tumors are eligible for complete removal, even if they are very large. According to a comprehensive review, the five-year progression-free survival rate after complete resection was 83%, compared to 55% after subtotal resection.

The role of adjuvant treatments, specifically radiotherapy, after complete resection in low-grade lesions remains challenging due to limited retrospective case reports. Although many patients with low-grade astroblastoma who underwent complete resection have received postoperative radiotherapy and achieved long-term survival, there have also been reports of successful long-term survival without radiotherapy in certain cases[3,10,11]. Positive responses and long-term survival have been observed with radiotherapy alone or in cases with incomplete resections[3,12]. Gamma knife-based radiosurgery has shown excellent local control in recurrent astroblastoma[13]. Adjuvant postsurgical radiotherapy may be considered for anaplastic astroblastoma, except for very young children to avoid severe side effects. Radiotherapy should also be considered for cases with incomplete resections in low-grade astroblastoma. The role of radiotherapy in completely resected low-grade cases remains uncertain.

As for chemotherapy and astroblastoma, there is limited data available. Some anaplastic astroblastomas have been treated with chemotherapy as part of initial treatment. However, it is unclear whether chemotherapy has contributed to overall survival rate, as it was predominantly given to patients with more aggressive lesions. Limited data on chemotherapy outcomes for recurrent lesions show some cases of complete remissions. In a study by Bonnin and Rubinstein[3], high-grade astroblastoma patients who did not receive postoperative radiotherapy had shorter survival time. Caroli et al. also reported a case of highgrade astroblastoma with a five-year recurrence-free survival after total resection, radiation therapy, and use of temozolomide. In our low-grade astroblastoma case, we recommended regular follow-up without adjuvant radiation, and there were no signs of recurrence during the last 6-month follow-up.

IV. CONCLUSION

Supratentorial astroblastomas are extremely rare. Radiological features of these lesions can vary. Surgical removal alone is enough for low-grade types, while high-grade and recurring tumors necessitate additional radiotherapy.

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