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A CASE REPORT ON ASTROBLASTOMA

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ABSTRACT

Background: Astroblastoma is a rare neuroepithelial tumor that often originates in the cerebral hemisphere of children and young adults. Diagnosis of this obscure neoplasm can be difficult because these tumors are so infrequently encountered and share common radiological and neuropathological features of other glial neoplasms. Case Description: We present a case of a low-grade astroblastoma diagnosed in a 30-year-old female with complaints of headache, vomiting and vision changes. CT was done which revealed anintraaxial neoplasm in the left frontal lobe. Further CE MRI brain was done which showed a heterogeneously enhancing solid &cystic mass in the left frontal lobe with septationsand predominant cystic component. There was mild peritumoral edema. She underwent gross total resection. There was noevidence of high-grade features hence adjuvant therapy was not planned postoperatively. Conclusion: Astroblastoma is a very rare primary brain tumor. Its diagnosis is often challenging because of the astroblastic aspects that can be found in astrocytic tumors, in ependymomas, and in non-neuroepithelial tumors. Astroblastoma therefore must be considered in the differential of supratentorial tumors in children and young adults. Treatment of such, as suggested by most recent literature, includes gross total resection and adjuvant radiotherapy for lesions exhibiting high-grade features.

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INTRODUCTION

Astroblastoma is a controversial and an extremely rare central nervous system neoplasm^[1-4]. It was first described by Bailey and Cushing in 1926, accounting for approximately 0.4–2.8% of primary brain tumors ^[5]. Although classically considered as pediatric brain tumors, astroblastomas tend to display a bimodal incidence ^[6]. This tumor is commonly found in the frontoparietal hemispheres, although other locations, such as brainstem, cerebellum, hypothalamus, and intraventricular, have been documented ^[7-9]. The clinical presentation is often related to signs of elevated intracranial pressure including headaches, nausea, and vomiting. Focal neurological deficits, seizures, and hemorrhage may also exist at the time of presentation. Much confusion has centered on the cell of origin as well as histopathologic criteria for diagnosis because they

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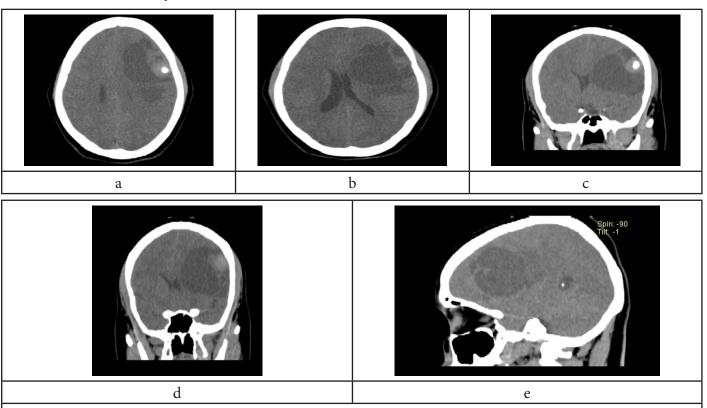
share features of both astrocytomas and ependymomas [10]. In the literature on brain tumor classification, this tumor has been categorized as follows: as a stage in the process of glioma dedifferentiation, as an astrocytoma of large cells producing fibers, or as a rare tumor, probably originating from tanycytes or ependymal astrocytes. Finally, it was listed among "other neuroepithelial tumors" in the WHO Classification of Tumors of the Central Nervous System. Astroblastomas can be graded as either a low-grade or high-grade variant. We present a case of an astroblastoma in a young woman, focusing on neuroimaging and neuropathological features.

CASE REPORT

A 30-year-old woman presented with history of seizure, left scalp paresthesias, left-sided headache, blurred vision, nauseaandvomitings. Her neurological examination was unremarkable, however, formal ophthalmological testing demonstrated left homonymous quadrantanopia.

She underwent a non-contrast computed tomography (CT), with results indicating an

intra-axial left frontal lobe neoplasm.

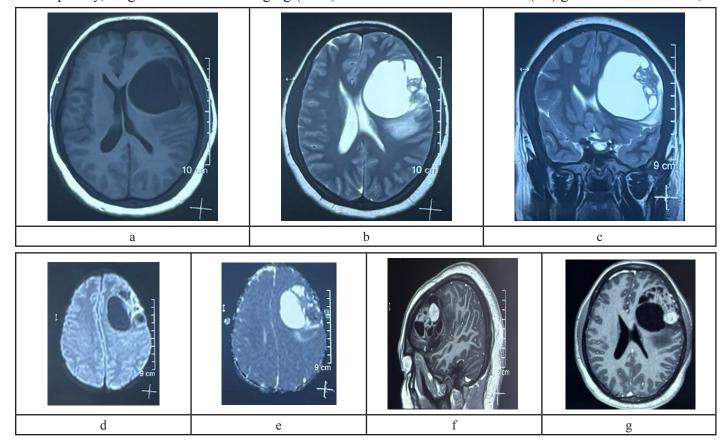


NCCTBRAIN: (a, b)- axial; (c, d)- coronal; (e)-sagittal reformatted images.

Single well defined intra axial mixed solid cystic lesion in left frontal lobe measuring 53x52x55mm with focal calcification in solid component and hypodense vasogenic edema surrounding the lesion. CT attenuation of the solid component is 55HU and cystic component is 30HU. There is mass effect on adjacent brain parenchyma and left lateral ventricle with midline shift of 6mm.

INTRA AXIAL LESION OF NEOPLASTIC ETOLOGY.

Subsequently, magnetic resonance imaging (MRI) of the brain with intravenous (IV) gadolinium was done,



CE MRI brain:

Large well defined mixed solid & Department of the solid amp; cystic space occupying lesion with variable thickness internal septations seen in left frontal lobe Measuring 5.4x5.4x5.2cms with predominant cystic component not suppressed on FLAIR. There was mild peritumoral T2 hyper-intensity, signifying peritumoral edema.

Lesion shows (a)T1, (b, c)T2, FLAIR hypo intensity with blooming on GRE measuring 5.5x5.5mm in posterolateral aspect representing calcification.

(d, e)- DWI, ADC showing patchy restricted diffusion in the solid component.

Lesion is causing mass effect in the form of compression of frontal horn and body of left lateral ventricle and subfalcine midline shift of 6mm to right side.

On contrast, there is (f, g) enhancement of both solid component and septations

DIFFERENTIAL DIAGNOSIS

Ependymoma

Ganglioglioma

Oligodendroglioma

Astroblastoma

The patient subsequently underwent a left craniotomy for resection and biopsy, with pathology described as a low-grade astroblastic tumor.

HISTOPATHOLOGY REPORT

Gross/ Macroscopic Description

Received bottle contained single grey white soft tissue mass measuring 2 x 1.6×1.3 cm.

Cystic wall measuring 4x3.5x1cm.

Tiny grey white to hemorrhagic bits altogether measuring 1.5x1x0.3cm.

Microscopic Description

Multiple sections from the specimen submitted studied showed a well circumscribed lesion comprised of cells arranged radially around vessels (pseudorosettes) and in sheets. The lesional cells are polygonal with round to oval vesicular nuclei and moderate to abundant eosinophilic cytoplasm. Perivascular sclerosis and hyalinizationis seen. Areas of hemorrhage are also noted.

IHC

GFAP - Positive

Vimentin - Positive

EMA - Negative

D240 - Positive

OLIG2 - Positive

Ki67 - 14%

FLUORESCENCE IN SITU HYBRIDISATION (FISH) REPORT

Specimen Type: Paraffin embedded tissue section

Test Results

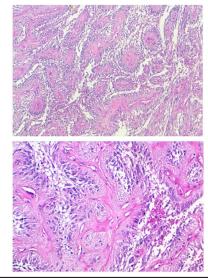
MN1 Break Apart FISH Positive

Fluorescence in situ hybridization (FISH) was performed on a paraffin embedded tissue section using MN1 dual color break apart probe (Wuhan Health care Biotechnology). 100 non-overlapping nuclei were assessed for the fused and or split green and red signals. More than 20% split signals are

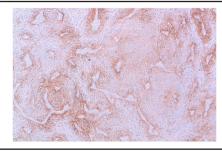
taken as positive. This test is intended to detect chromosome rearrangements involving the MN gene region long arm (q) of chromosome 22 at position 12.1. This is useful in diagnosis of ASTROBLASTOMA AND CNS NEUROBLASTOMAS

This test is intended to be used as an adjunct to existing clinical and pathologic information currently used for the differential diagnosis of round cell tumors.

Diagnosis: Astroblastoma, MN1 Altered



Microscopic appearance: tumor composed of perivascular rosettes of tumor cells with thickened and focally hyalinized blood vessel walls (hematoxylin and eosin stain; original magnification × 100)



Positive immunostaining for glial fibrillary acid protein (original magnification \times 100)

DISCUSSION

Astroblastoma is one of the rarest central nervous system gliomas. It can occur in persons of any age, with a bimodal age distribution, with one peak in infancy (between 5 and 10 years) and the other one in young adults (between 21 and 30

years)^[6]. The studies performed to date show a striking female preponderance with a male to female ratio of 1:11^[11].

The tumor usually presents as a well-circumscribed and superficial mass, usually supratentorial with occipital and frontal lobes the most frequently affected sites. However, tumor invasion has also been reported into corpus callosum, cerebellum, brain stem, and optic nerve.

Clinical signs and symptoms differ depending on the tumor's location and size, and are primarily related with increased intracranial pressure. The most typically reported symptoms include headaches, seizures, vomiting, and focal neurologic abnormalities.

Considerable confusion has surrounded the diagnosis, the histogenesis, and the classification of astroblastoma. Controversy still exists in the literature of the cell of origin of this neoplasm. Bailey and Bucy believed that astroblastoma originated from astroblasts, an intermediate stage between glioblasts and astrocytes [12]. However, Russell and Rubinstein suggested that astroblastomas are dedifferentiated from mature astroglial cells [13]. Later, in a study by Rubinstein and Herman using electron microscopy, it was proven that astroblastomas might originate from persisting groups of embryonic precursor cells, transitional between astrocytes and ependymal cells [14]. Given the lack of consensus, astroblastomas are currently classified as other neuroepithelial tumors by the WHO Classification of Tumors of the Central Nervous System2021.

On MRI, it is usually seen supratentorially and is peripheral in location. It typically appears as a large, well-demarcated, lobulated mass. It often has solid and cystic components with a characteristic bubbly appearance in the solid component, which was believed to result from the tumor vascular architecture, with inhomogeneous contrast enhancement and little vasogenic edema. It is hyperintense to white matter on fluidattenuated inversion recovery (FLAIR) imageand T2- weighted imageand hypointense to isointense on T1- weighted image.

On macroscopic examination, astroblastomas were described as superficial, well-demarcated, lobulated, solid, or cystic masses

On histologic examination, an astroblastoma is defined by the presence of perivascular pseudorosettes and prominent perivascular hyalinization. The perivascular pseudorosettes give the characteristic "cartwheel" appearance. They exhibit characteristic epithelioid cells with cytoplasmic processes having blunt-ended foot plates attached to the basal lamina of blood vessels. The amount of perivascular hyaline formation varies from case to case; but in the most severe forms, expansive, acellular hyalinized zones will be seen without any residual tumor architecture. Another feature of diagnostic importance is lack of fibrillary background. Higher grade lesions will occasionally have clusters of tumor cells extending marginally into surrounding brain parenchyma.

Immunohistochemical features of astroblastoma have some variability throughout the literature. Immunostaining for GFAP is positive, lending support to the theory that the tumor cell is derived from an astrocyte cell line. Astroblasts also consistently stain positive with vimentin, suggesting derivation from a more primitive astroblast, and for S-100 protein.

The diagnosis of astroblastoma is often difficult. In fact, astroblastic features are not unique to astroblastoma and can also be found in other tumors. Therefore, the combination of the radiologic and the histopathologic characteristics is necessary for making a correct diagnosis.

Data on the molecular genetics of astroblastoma are rare and only recently available from the literature. A study by Brat et al. demonstrated that astroblastomas have characteristic chromosomal aberrations because they exhibit gain of chromosomes 19 and 20. These anomalies are different from those of the ependymomas or astrocytic tumors, suggesting that astroblastoma is a distinct entity rather than a variant of ependymoma. Other alterations noted were losses on 9q, 10, and X chromosome. Shuangshoti et al. found loss of heterozygosity at the D19S412 locus on 19q in a cerebral astroblastoma. More recently, an absence of IDH 1/2 and TP53 mutations, which are known to be involved in the development of low-grade gliomas, was shown in astroblastomas.

MANAGEMENT

Total resection is the best treatment^[1,2]. It provides excellent tumor control rates. Subtotal resection should be avoided, if possible. The addition of adjuvant focal radiotherapy after subtotal resection does not appear to provide equivalent outcomes to gross total resection. Adjuvant therapy for high-grade and recurrent cases is recommended^[15]. Regular follow-up is required even in low-grade variants due to unpredictable behavior.

CONCLUSION

Astroblastoma is a very rare primary brain tumor. Its diagnosis is often challenging because of the astroblastic aspects that can be found in astrocytic tumors, in ependymomas, and in non-neuroepithelial tumors. Considerable confusion surrounds its histogenesis and classification. Thereby Astroblastoma must be considered in the differential of supratentorial tumors in children and young adults. Treatment of such, as suggested by most recent literature, includes gross total resection and adjuvant radiotherapy for lesions exhibiting high-grade features.

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