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Case Study

Astroblastoma: a rare case report

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Abstract

Background: An astroblastoma is a rare primary glial tumour occurring preferentially in young adults. It is characterised by a perivascular arrangement of tumour cells forming perivascular pseudorosettes mimicking ependymomas. The histogenesis of astroblastoma is unclear.

Case description: We present the history of a 13-year-old girl with chief complaints of headache associated with vomiting, blurring of vision on the left eye and a history of diplopia on the right eye. She underwent left parietal parasagittal craniotomy and near-total excision of tumour. She was planned for postoperative radiotherapy 5,940 cGy in 28 fractions along with concurrent temozolamide100 mg. She had no neurological deficit or complaints during her last visit.

Conclusion: Astroblastomas are a distinct clinic pathologic entity, with well-described radiologic, pathologic and cytogenetic features. Its recurrence is high, and efforts must be made to elucidate the role and usefulness of radiotherapy and chemotherapy in these tumours.

Keywords: astroblastoma; glioma; histological study; temozolamide

INTRODUCTION

Astroblastoma is a rare glial tumour affecting children and adolescents whose histogenesis remains uncertain. It accounts for 0·5% of all glial neoplasms. These tumours were initially described by Bailey and Cushing and further supported by Bailey and Bucy.[1] Generally, it is a solid well-circumscribed tumour defined histologically by the presence of astroblastic pseudorosettes and prominent perivascular hyalinisation of glial fibrillary acidic protein (GFAP)-positive astrocytic cells with broad non-tapering processes radiating towards a central blood vessel.[2] Two different subtypes of astroblastoma have been defined on the basis of histological characteristics. These tumours are usually located in the cerebral hemispheres, but have also been described in the cerebellum, brainstem, corpus callosum, hypothalamus and the ventricular system. The neuroradiological appearance is characteristic; typically, they present as supratentorial multi-lobulated lesions with both solid and cystic components.[3] Here, we present the clinical features of a child with left lateral ventricle astro-blastoma treated at our institution.

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CASE HISTORY

History and presentation

We present the history of a 13-year-old girl with chief complaints of headache, progressive in nature, associated with vomiting since 6 weeks, and blurring of vision on the left eye and a history of diplopia on the right eye since 1 week. She had an uneventful past medical and surgical history. On examination, her Glasgow Coma Scale was 15. Clinically, no focal neurological deficit was observed. Fundoscopy was suggestive of papilloedema in the left eye; other system examinations were clinically normal. Complete laboratory investigations and cardiac evaluation were normal.

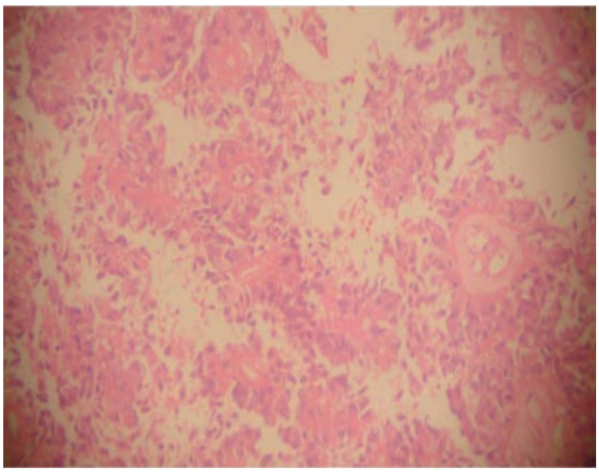

Figure 1. HPE findings—pseudorosette.

Radiological findings

Magnetic resonance imaging (MRI) findings revealed a well-defined intra-axial heterogeneous altered signal intensity lesion in the left occipital measuring ~6·7 × 4·0 cm. Anteriorly, it caused compression on the left occipital horn of the lateral ventricle. No midline shift was seen. It was predominantly cystic with solid areas within. The solid area appeared isotense on T1W1 and T2 fluid-attenuated inversion recovery (FLAIR) imaging with the cystic area showing hyperintensity in FLAIR imaging. It showed heterogeneous enhancement after contrast.

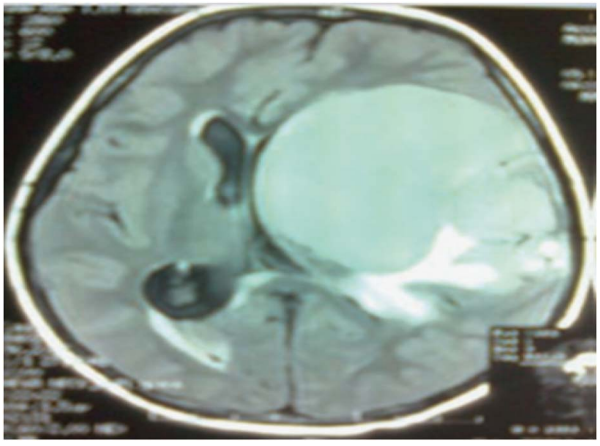

Figure 2. Magnetic resonance imaging pre-op: T2-weighted image.

Surgical treatment

She underwent left parietal parasagittal craniotomy and near-total excision of tumour. The patient withstood the procedure well. Postoperatively, the patient developed periorbital oedema, which resolved. Hospital stay was uneventful. Post-op computed tomography scan revealed post-op decompression with ring-enhancing area noted in the post-op region. Oedema was observed on the post-op site.

Histopathology status

Post-op pathological examination of the tumour suggested astroblastoma in the left lateral ventricle. Microscopic sections showed a cellular tumour with plump cells having eosinophilic cytoplasm and large vesicular nuclei arranged around sclerosed blood vessels. Stromal hyalinisation was also seen. The enclosed groups of tumour cells showed lobular/acinar configuration. Immunohistochemical analysis with S100 and vimentin showed uniform diffuse labelling of tumour cells. Glial Fibrillary Acidic Protein (GFAP) showed patchy labelling in a small group of cells. Electron Microscopic Array (EMA) was group membrane labelling cells with focal paranuclear dot. MIB Index (MIB-1) 15%. Cytokeratin and P53 were negative, and INI-1 expression was retained (Figures 1 and 2).

Radiotherapy treatment

She was planned for postoperative radiotherapy 5,940 cGy in 28 fractions along with concurrent

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temozolamide 100 mg. She had no neurological deficit or complaints during her last visit.

DISCUSSION

Astroblastoma is a rare type of glial tumour that was first described by Bailey and Buey in 1930.[1] Although its existence has been questioned by some investigators, it is now recognised as a distinct clinicopathological entity, probably representing <0·5% of gliomas. It usually appears in young adult patients, although congenital cases have also been described. No sex preference has been observed. Characteristic features for a diagnostic orientation are young age and localisation of the lesion, as astroblastoma almost always presents as an intra-axial peripheral supratentorial lesion, more frequently in the vicinity of the convexity, and more often involves the frontal and parietal lobes of a single hemisphere, or presents in a medial-sagittal site. Exceptionally, other localisations have been reported.

Histologically, astroblastoma has characteristic perivascular pseudorosettes with short, thick cytoplasmic processes that show blunt-ended footplates and are attached to the basal lamina of the blood vessels, frequently by hyalinisation. Borders of the lesion more often compress rather than infiltrate the brain parenchyma.[2] This observation is fundamentally important for understanding the behaviour of this tumour and in particular allows its radical surgical excision. Two histological types have been recognised: (1) differentiated astroblastoma and (2) anaplastic astroblastomas. The latter show cytological atypias, compact cellularity, perivascular cells with high mitotic rates and hypertrophy of the vascular endothelium.

In practical terms, the prognosis may be good and the evaluative course may be prolonged if total resection of astroblastoma is performed; however, a longer period of follow-up observation is necessary in such cases. This patient presented to us with postoperative residual disease and she was not reliable for regular follow-up. On performing a literature search, we could not find any case report addressing this situation. In this regard, our treatment decision is unique. In some selected cases, the efficacy of radiotherapy has been widely demonstrated, but mainly with reference to high-grade cases. In fact, the only patient who had been subjected to radiotherapy after biopsy alone was still alive 12 years after diagnosis. Moreover, the only patient with a high-grade astroblastoma who had not been subjected to adjuvant radiotherapy presented the worst prognosis, with a survival time of 1·5 years. No evidence of adjuvant chemotherapy was found in the literature.

Sughrue et al. performed a systematic comprehensive search of the published literature in English on patients undergoing surgery for astroblastoma to summarise what is known about these tumours, and to provide some framework for future efforts in this area. A total of 62 references met their inclusion criteria, and contained individual patient data on 116 patients with astroblastoma.[3]

Lau et al. described the case of a patient with a low-grade lesion removed totally, but not treated with adjuvant radiotherapy, who presented with recurrence 12 months after surgery. Subtotal removal of the recurrence was followed by radiotherapy. Although follow-up MRI showed tumour residue, the disease appeared to be under control. It therefore appears evident that adjuvant radiotherapy is effective and undoubtedly advisable for high-grade lesions that warrant a more aggressive approach. There have been some reports suggesting local relapses in low-grade astroblastoma patients not undergoing radiotherapy.[4]

CONCLUSION

Astroblastoma is a distinct type of glial tumour, usually well circumscribed and supratentorial in young adults, with the characteristic histopathological pattern of perivascular arrangements of tumour cells. In this report, we have described and discussed our clinical, radiological and pathological findings and management in a patient with astroblastoma.

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