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Review article

Angiocentric glioma transformed into anaplastic ependymoma: Review of the evidence for malignant potential

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ABSTRACT

Angiocentric glioma (AG) is a low grade glioma, that was first described in 2002. Since this description, 83 patients with AG have been described, including ours. AG typically presents in childhood with medically refractory seizures that are cured with gross surgical resection. Whilst the natural history is that of a benign tumour, there have been reports of recurrence, transformation, and malignant features that suggest that AG is potentially malignant. We add to the literature a case of a 16-year-old girl who presented in May 2011 with a 3-month history of complex partial seizures, with MRI showing a T2-weighted hyperintense lesion in the left insula and inferior frontal lobe. This was confirmed on biopsy as AG and was followed with surveillance imaging. In April 2012, she presented with disease progression and underwent a left temporal lobectomy, with histology showing both AG and grade II astrocytoma. Adjuvant radiotherapy of 50 Gray in 28 fractions was administered. A small area of contrast enhancement appeared in the left parietal lobe in December 2012, which progressed over subsequent months. In June 2013, she underwent a near total excision, with histology showing anaplastic ependymoma. She received six cycles of adjuvant temozolamide. Despite this, the tumour continued to progress, with her seizure control deteriorating, and the development of a right hemiparesis. The patient died in January 2014, aged 19 years.

1. Case report

A previously well 16-year-old girl presented in May 2011 with complex partial seizures. A CT scan of the brain showed a left frontal and insular hypodense region and subsequent MRI showed an area of T2-weighted hyperintensity in the left insular region and inferior frontal lobe involving both white matter and cortex. There was no contrast enhancement (Fig. 1). She was commenced on carbamazepine and a stereotactic biopsy showed a moderately hypercellular glial tumour with accumulation of cells around blood vessels (mimicking perivascular pseudorosettes) and immediately deep to the pia. There was also prominent perineuronal satellitosis. Tumour cells had uniform round/oval vesicular nuclei with finely stippled chromatin and coarse processes. There was no necrosis, endothelial cell hyperplasia or mitoses. Immunohistochemistry showed strong staining of tumour cell processes for glial fibrillary acidic protein (GFAP), no staining for isocitrate dehydrogenase 1 (IDH-1), patchy staining for p53 and a topoisomerase labelling index of 3% (Fig. 2). After expert neuropathological review and external consultation from the University of California San Francisco (UCSF), the diagnosis was angiocentric glioma (AG) (World Health Organization Grade I). The initial treatment plan after multidisciplinary review was to observe the lesion with serial imaging. Her seizures were difficult to control, with frequent complex partial and occasional secondarily generalised seizures, requiring the addition of leveteracetam, and later topiramate.

Surveillance MRI in October 2011 was stable, however MRI in April 2012 showed marked progression of the tumour throughout the insula, thalamus, frontal and temporal lobes with midline shift, and was accompanied by symptoms of raised intracranial pressure (Fig. 3). A left temporal lobectomy for decompression was undertaken. The histopathological examination of the resected tissue showed features similar to the original AG as well as larger areas more consistent with grade II astrocytoma. The tumour remained immunonegative for IDH-1 and there was some non-specific staining for epithelial membrane antigen (EMA) in perivascular tumour cells (Fig. 4). The proliferative index was 1%. She was again reviewed by the multidisciplinary team and post-operative radiotherapy was recommended. She received 50 Gy in 28 fractions.

A small area of contrast enhancement appeared in the deep left parietal lobe in December 2012 (Fig. 5). This enlarged over

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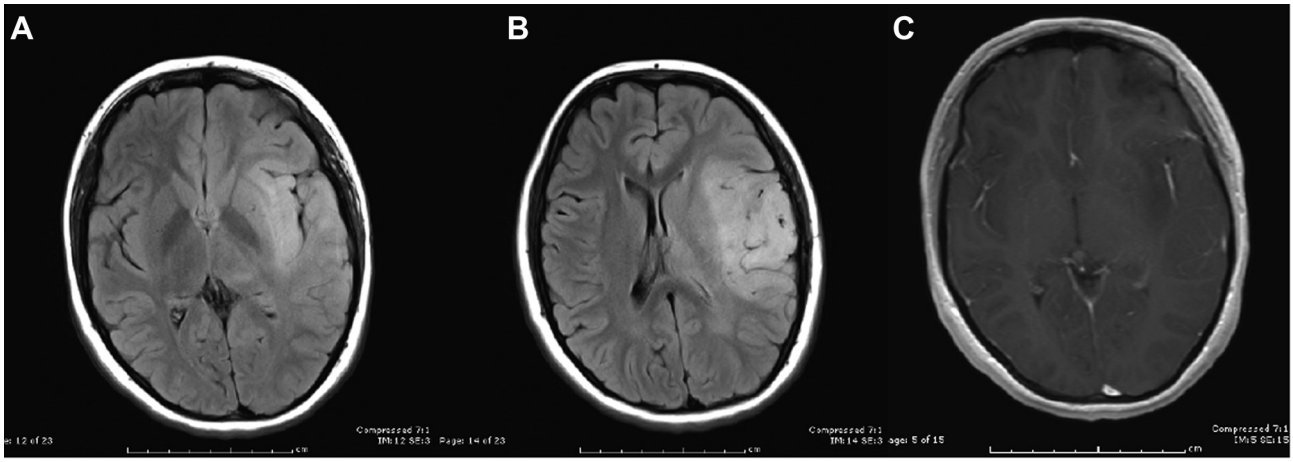

Fig. 1. (A and B) Left inferior frontal, temporal and insular region of FLAIR hyper intensity involving both white matter and cortex. (C) There was no contrast enhancement on axial T1-weighted gadolinium enhanced MRI.

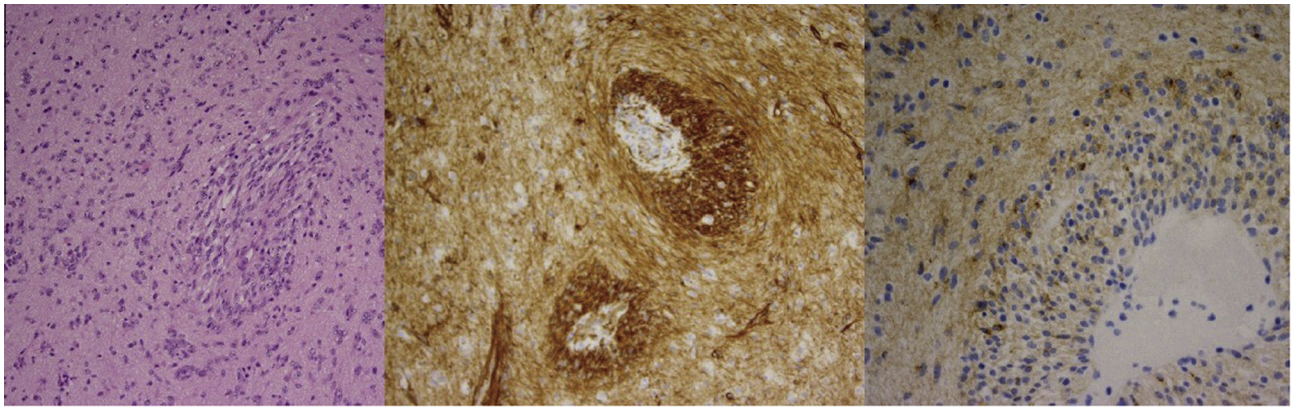

Fig. 2. (A) Haematoxylin and eosin stained section of the first biopsy surgical specimen (×400) showing sleeve-like aggregations of tumour cells around small blood vessels in white matter. (B) Immunostaining for glial fibrillary acidic protein (GFAP) highlighting astrocytic differentiation and the angiocentric accumulation of tumour cells and (C) immunostained for epithelial membrane antigen (EMA) showing perinuclear dot staining suggesting ependymal differentiation.

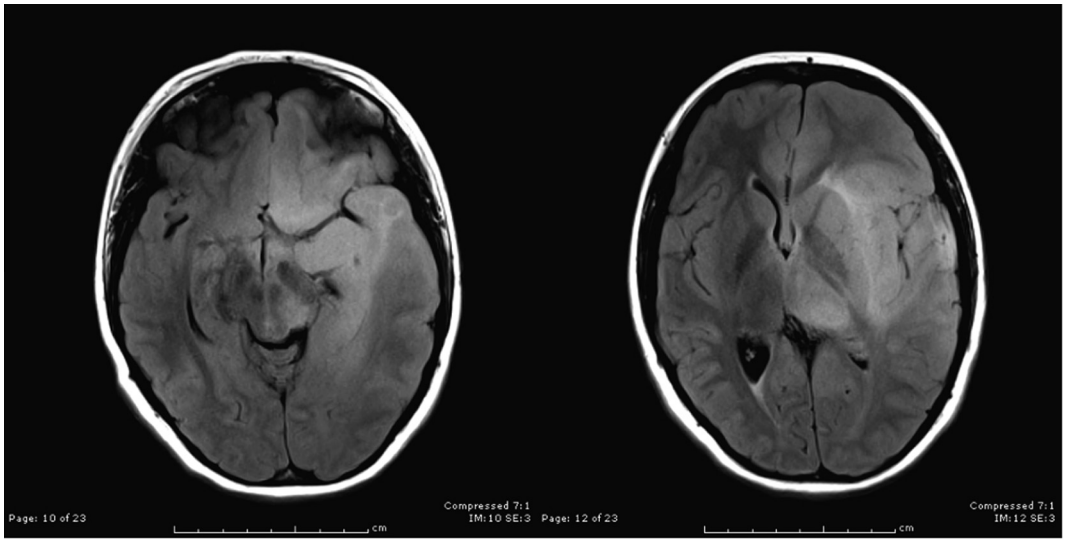

Fig. 3. Axial fluid attenuated inversion recovery MRI of the brain (April 2012, at progression after biopsy) showing progression of left insular lesion with extension into the thalamus, frontal and temporal lobes, causing mass effect upon left ventricle, and associated midline shift.