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Progressive dysembryoplastic neuroepithelial tumour



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ABSTRACT

Dysembryoplastic neuroepithelial tumour (DNET) is a benign tumour characterised by cortical location and presentation with drug resistant partial seizures in children. Recently the potential for malignant transformation has been reported, however progression without malignant transformation remains rare. We report a case of clinical and radiological progression of a DNET in a girl 10 years after initial biopsy.

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1. Introduction

Dysembryoplastic neuroepithelial tumour (DNET) is a benign tumour characterised by its cortical location and presentation with drug resistant partial seizures in children. They are mixed neuro-glial tumours classified as World Health Organization grade I [1]. These lesions are generally considered benign or hamartomatous in nature and surgical removal for tissue diagnosis and seizure control is considered curative.

There is a common belief that progression or post-surgical recurrence of these tumours is not seen and indicates an incorrect diagnosis. Recently a number of case reports have described recurrence or progression in the setting of a malignant transformation of these tumours [2–4]. The patient reported here illustrates

clinical and radiological progression of a DNET without malignant transformation.

2. Case report

The patient presented at 16 years of age complaining of intermittent visual disturbances with two episodes of loss of consciousness associated with the visual blurring. She was otherwise well with no other medical conditions and physical examination, including visual field examination, was unremarkable.

Her past history was unremarkable prior to the age of 15 months when she suffered febrile convulsions followed by a seizure type event whilst at kindergarten aged 5 years. From the age of 10 years she described regular episodes of visual blurring, particularly aggravated by physical activity.

MRI of her brain revealed a 5.2 cm mass in the right occipital lobe with cystic changes and remodelling of the overlying skull (Fig. 1A). There was no enhancement with gadolinium or mass

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effect. These findings were regarded as consistent with DNET. Inter-ictal electroencephalogram was unremarkable.

The patient was started on carbamazepine for control of visual seizures. While resection of this lesion was not thought to be appropriate due to the risk of visual field defects she did undergo biopsy to obtain a tissue diagnosis. At surgery the cerebral cortex appeared pale and expanded. A generous sample was taken for histological examination and was reported as a DNET with Ki67 proliferation index less than 1% (Fig. 2A–C).

Postoperatively she had no deficit and remained clinically well for an extended period with only occasional visual seizures. She remained on carbamazepine and 12 monthly MRI scans were performed. During this time the pattern of enhancement of the lesion varied with the overall size remaining constant

but with periodic enhancement of nodules within the tumour (Fig. 1B).

Ten years following the original surgery she re-presented with headache and lethargy following a mild head injury. There had been no change in seizure activity and no visual field deficit was present on examination. MRI revealed progression in the lesion size with enlargement of both cystic areas and an enhancing nodule (Fig. 1C). There was mass effect and oedema.

She underwent craniotomy and gross total resection of the lesion without complication. At surgery the cortex appeared tense and bulging. The tumour was soft with variable appearance of pale grey/pink and golden brown areas. The cyst contained golden fluid. Histological examination revealed cortical nodules with variable stromal myxoid change and oligodendroglioma-like appear-

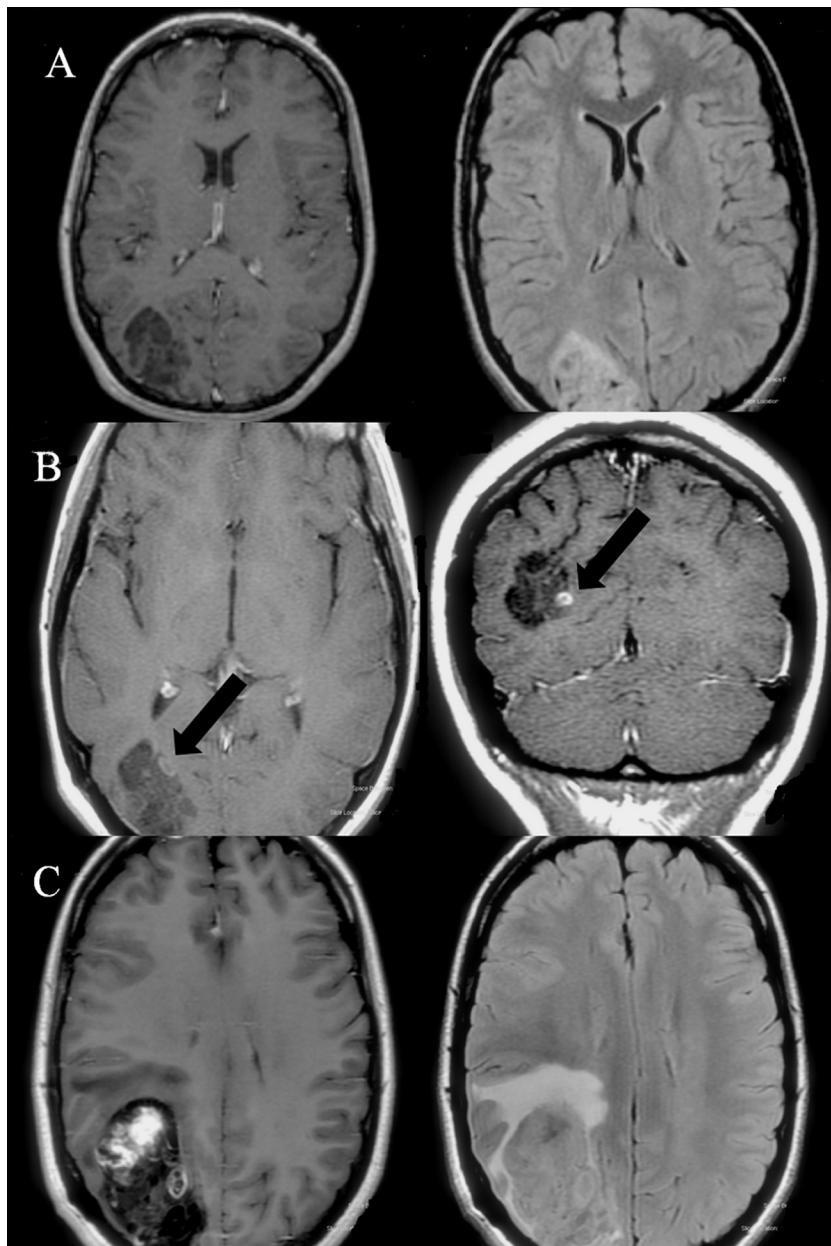


Fig. 1. MRI of the lesion. (A) Axial T1-weighted with gadolinium (left) and fluid attenuated inversion recovery (FLAIR; right) images at presentation demonstrating the 5.2 cm mass in the right occipital lobe with cystic changes and remodelling of overlying skull. (B) Axial (left) and coronal (right) T1-weighted with gadolinium contrast images taken at different time points demonstrating varying intensity of the enhancing nodule without change in size. (C) Axial T1-weighted with gadolinium (left) and FLAIR (right) images show progression in the lesion size after 10 years with enlargement of both cystic areas and the enhancing nodule with mass effect and oedema.

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