Progressive dysembryoplastic neuroepithelial tumour

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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 radiologic 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 neuronal-glial tumours classified as World Health Organization grade I [11]. 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, seizure type event whilst at kindergarten aged 5 years. From the age of 10 years she described regular episodes of visual blurring, 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...
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.
ance, containing floating neurons which appeared dysplastic (Fig. 2D–F). Ki67 staining remained less than 1% throughout the sample. The samples were reviewed independently by two neuropathologists and were consistent with DNET.

Postoperatively she recovered well but was noted to have a persistent left inferior quadrantanopia.

3. Discussion

This patient represents a rare case of true progression of a DNET after a period of 10 years during which its behavior was typically benign.

The biology of DNET is poorly understood. There is controversy regarding whether DNET represent true neoplasms or hamartomas. The original description of DNET by Daumas-Duport reported 39 patients and postulated a germinal origin for the glioneuronal element, however the histogenesis remains uncertain [5]. There has been further classification into three subtypes, namely simple, complex and non-specific [6,7]. Simple DNET contain only the “specific glioneuronal element”, while the complex DNET also contains glial nodules and cysts. The non-specific DNET is controversial in that it is said to contain none of the classical hallmarks of DNET and rather appears as a diffuse glioma but is confined to the cortex. By this classification our patient represents a complex DNET however the clinical relevance of this classification is unclear.

Progression of DNET has been reported but remains rare [8]. One potential explanation is incorrect diagnosis. Histological diagnosis can be difficult especially with small samples from biopsy. Features of glial lesions that can lead to mis-classification of DNET include the nodular and microcystic pattern in some oligodendroglioma; additionally, secondary changes in cortex caused by glioma may be difficult to distinguish from DNET. In this case a large cortical sample was taken at the initial biopsy, as opposed to needle aspiration, and full excision was achieved during the second surgery. This makes sampling error and misdiagnosis unlikely.

In this patient, change in MRI enhancement was seen over time. This has been previously reported in DNET without any changes in the histology or clinical characteristics. A more concerning phenomenon is malignant transformation within the tumour [9]. As DNET represents a mixed glial-neuronal tumour at least within the “specific glioneuronal element” there is potential for development of astrocytoma or oligodendroglioma components, and this has been previously reported [4,10,11]. Some authors have suggested markers, such as the Ki67 proliferation index, that may indicate tumours which demonstrate more aggressive characteristics, but this remains to be validated [4]. In the patient described above the subsequent histological examination showed unequivocal DNET features with low Ki67 index rather than transformation into a more malignant variant, indicating true progression of a benign DNET.

This patient demonstrates that while DNET are believed to be benign tumours, progression can occur and does not always represent misdiagnosis or malignant transformation.

Conflicts of Interest/Disclosures

The authors declare that they have no financial or other conflicts of interest in relation to this research and its publication.

References

Ventriculo-peritoneal shunt malfunction due to complete migration and subgaleal coiling of the proximal and distal catheters

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Ventriculo-peritoneal (VP) shunt malfunction due to proximal and distal catheter migration has been rarely reported in the literature. Shunt migration has been proposed to occur as a result of a combination of various mechanisms, including the windlass effect, retained memory of the shunt tubing, inadequate shunt fixation, and increased intra-abdominal pressures. We describe a rare case of a 6-week-old child who presented in our department with VP shunt malfunction due to complete proximal migration and coiling of the peritoneal and ventricular VP shunt catheters within a subgaleal pocket at the left occipital area.

1. Introduction

Intraventricular hemorrhage (IVH) may be complicated with post-hemorrhagic hydrocephalus. Once diagnosed, hydrocephalus is frequently treated with ventriculo-peritoneal (VP) shunt insertion. VP shunt malfunction due to proximal shunt migration has been proposed to occur as a result of increased intra-abdominal pressure, inadequate shunt fixation, the windlass effect, and retained shunt tubing memory [1–4]. A variety of mechanisms have been hypothesized to be responsible for VP shunt catheter migration. Shahsavaran et al. suggested retained memory of the shunt tubing as the main cause of migration [5]. Various factors have been proposed to be responsible for this phenomenon, including the windlass effect, retained memory of the shunt tubing, inadequate shunt fixation, and increased intra-abdominal pressures. We describe a rare case of VP shunt migration due to complete proximal and distal migration within the subgaleal space leading to signs and symptoms of VP shunt malfunction.

2. Case report

2.1. History and physical examination

A 6-week-old boy with a history of IVH and posthemorrhagic hydrocephalus managed with VP shunt insertion at an outside hospital when he was 3 days old was brought to the emergency room (ER) with a 24 hour history of progressive apathy and recurrent vomiting. Admission physical examination was significant for a large subgaleal mass at the left occipital area (Fig. 1). Non-contrast head CT scan obtained in the ER was significant for ventricular dilatation, and for failure to identify the ventricular catheter within the ventricular system, suggestive of proximal VP shunt catheter migration within the subgaleal fluid filled pocket (Fig. 2). Lateral skull, thoracic and abdominal radiographs (Fig. 3) identified the distal VP shunt catheter also within the subgaleal pocket.

2.2. Treatment and follow-up

The child underwent uncomplicated insertion of a new VP shunt accompanied with dissection of the subgaleal fluid collection and removal of the old shunt. His post-operative course was unremarkable and he was discharged home on post-operative day 3 with instructions for regular follow-up visits.

3. Discussion

Complete migration of the proximal and distal VP shunt components into the subgaleal space leading to signs and symptoms of VP shunt malfunction has been infrequently reported in the literature [2–4]. A variety of mechanisms have been hypothesized to be responsible for VP shunt catheter migration. Shahsavaran et al. proposed that the presence of a subgaleal fluid collection with dissection of the subcutaneous space around the catheter may promote catheter migration to the subgaleal space [4]. Dominguez et al. suggested retained memory of the shunt tubing as the