CASE REPORT

An unusual recurrence of dysembryoplastic neuroepithelial tumours after a seizure-free period of 8 years

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SUMMARY

Dysembryoplastic neuroepithelial tumours (DNETs) are grade 1 central nervous tumours, which can be diagnosed judiciously based on clinical features and radiological investigations, having a good prognosis after surgical resection. However, the recurrence of tumours due to variable epileptic zones leading to persistence of seizures can make the management of such cases challenging. We present a case of DNET where the patient had recurrence of the tumour with worsening of seizure activity 8 years after initial surgical resection.

BACKGROUND

This is a case of dysembryoplastic neuroepithelial tumour (DNET) in a patient who presented with complex partial seizures with secondary generalisation due to the recurrence of DNET, after a seizure-free period of 8 years. We present this case due to its unusual recurrence after a long-term seizure-free period suggesting the importance of regular follow-up of these patients.

CASE PRESENTATION

A 27-year-old man presented with a history of first seizure occurrence at the age of 7; the frequency of seizures being 2–3 episodes/week. He was diagnosed to have a right temporo-parietal intracranial space-occupying lesion for which he was operated in 2004. The details of the surgery and histopathology reports were not available. Postoperatively, the patient was continued on carbamazepine 200 mg twice a day, and he reported no further episodes of seizure for 8 years.

However, by the end of 2011, there was a recurrence of seizure episodes with a frequency of 2–3 episodes/week. The frequency of seizures increased to 2–3/day. The ictal semiology was suggestive of complex partial seizures, with each episode lasting for a brief period of 15–60 s with a secondary generalisation lasting for a short time. There was no history of motor deficit, tongue bite or urinary incontinence. There was no history of any other significant medical illness or surgical history.

He was admitted for further evaluation and management. Central nervous system examination revealed normal higher mental functions: bilateral 3 mm, isocoric pupils with normal reaction to light. Extraocular movements were full. Fundi were normal. No craniopathy, cerebellar signs, sensory or motor weakness were identified. Plantars were

flexor bilaterally and deep tendon reflexes were brisk.

INVESTIGATIONS

Routine laboratory investigations were within normal limits. Serum carbamazepine level was sub-therapeutic (3.6 µg/mL; normal range 8–12 µg/mL). An MRI brain plain and contrast (gadolinium enhanced) revealed postoperative gliosis in the right temporal grey and white matter with secondary encephalomalacia exvacuo enlargement of the right temporal horn and lateral ventricle (figure 1A). A video EEG was performed which was suggestive of right temporo-frontal focal onset seizures with a rapid but brief secondary generalisation. An ictal brain single photo emission CT scan revealed absent perfusion in right parieto-temporal lobe region which corresponded to the craniectomy site and reduced perfusion was noted in the right parietal lobe region with an area of focally increased perfusion adjacent to that region.

TREATMENT

Antiepileptic medication was stepped-up to obtain therapeutic levels and a craniotomy was planned. A right temporal lobectomy was performed which incorporated the gliotic tissue around the temporal horn of the lateral ventricle. Histopathology specimen of the brain tissue showed nodular collections of small round oligodendroglia-like cells, in sheets and columns, at places lying within a pale myxoid matrix with microcystic changes (figure 2). No nuclear atypia or abnormal mitotic activity was observed; tumour necrosis or vascular endothelial proliferation was not present. The findings were consistent with DNET.

OUTCOME AND FOLLOW-UP

Postoperatively, the patient was continued on levitiracetam 1000 mg twice daily. The patient was discharged 5 days after surgery. The short-term follow-up was uneventful. The patient has been on regular follow-up for the last 6 months and has been seizure free.

Since there is no clearly defined long-term follow-up period, in our case we plan to continue observing the clinical presentation of the patient every 6 months and repeat an MRI after 1 year and then extend the period of follow-up, if asymptomatic.

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Figure 1 (A) MRI brain plain showing postoperative gliosis in right temporal grey and white matter with secondary encephalomalacia exvacuo enlargement of the right temporal horn. (B) EEG recording showing right temporo-frontal focal onset seizures with rapid brief secondary generalisation.

DISCUSSION

DNETs are rare neuro-epithelial tumours, which was first described by Daumas-Duport in 1988.¹ They are classified as grade 1 tumours of the central nervous system according to the WHO. They are benign, commonly supra-tentorial tumours with the most common site being the temporal lobe. Cerebellar DNETs have been reclassified as Rosette-forming glioneuronal tumour of the fourth ventricle.² Most of the patients belong to the age group of less than 20 years and invariably have a history of pharmacologically intractable seizures.

DNETs can be histologically classified into three types namely simple, complex and non-specific.^{3, 4} They are usually stable tumours; however, they can rarely recur after surgical resection and some cases of malignant transformation have been reported as well.⁵ According to one study by Nolan *et al*⁶ in 2004, the rate of recurrence was 62.5% on a long-term follow-up after tumour resection. However, on the contrary, a study had reported an incomplete tumour removal in 28% of cases, but in none of these cases the tumour recurred on a follow-up of 7 years.⁴ In our case, the postoperative MRI report after the first surgery was not available. The recurrence of the tumour in our case may be due its incomplete resection in 2004, as it has been a common finding according to certain studies, but

recurrence of the tumour as well as seizures not amenable to antiepileptics after a symptom free period of long duration, as in our case, is very rare. We suspected a malignant transformation of the tumour; however, the biopsy after tumour resection was negative for any malignant transformation.

An MRI-based scheme for epilepsy surgery for DNET has recently been suggested, since the main prognostic factors for a seizure-free outcome were complete tumour and epileptic zone removal.⁷

Even though some studies suggest that patients with DNET who are seizure free for 3 years after tumour resection could be considered cured⁸, our patient presented with the recurrence of seizure after a seizure-free period of 8 years. Being a relatively new disease described in the literature in 1988, the long-term outcomes are not that well defined. However, follow-up is very significant in these patients to diagnose a recurrence. Recurrent DNET has a much better prognosis compared to other intracranial tumours, if it is operated upon immediately.

Furthermore, they can rarely recur or have a malignant transformation, of which a neurologist should be vigilant about on follow-up. Finally, short duration of seizures has been found to favourably influence the outcome⁶, which make a prompt diagnosis and apt surgical resection of paramount importance.

Figure 2 H&E stain showing loosely dispersed oligodendroglial-like cells lying within an acellular stroma.

Learning points

- Dysembryoplastic neuroepithelial tumours (DNETs) are benign grade 1 tumours, which have good surgical outcome and prognosis.
- The aim during the surgery should, however, be to completely excise the tumour mass along with any epileptic zone which may extend beyond the tumour mass.
- DNET may, however, occur after a seizure-free period of many years, which reiterates the importance of a long-term follow-up.

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