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**Case Report**

# Dysembryoplastic neuropithelial tumor: a rare case report

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## ABSTRACT

Dysembryoplastic neuropithelial tumor (DNET) is a rare recently described, benign glioneural tumor frequently associated with intractable seizures in children and young adults which is important to recognise clinically and radiologically as it is surgically curable without need for adjuvant chemoradiotherapy. We hereby present a case report of a 10year old male child who presented with intractable seizures and right parietal space occupying lesion which was diagnosed DNET radiologically, treated by microsurgical excision and confirmed histopathologically as DNET, thus emphasising multidisciplinary role in management of this rare entity.

**Keywords:** DNET, Epilepsy, Histopathology, MRI

## INTRODUCTION

DNET is a rare low grade neoplasm included in recent WHO 2007 classification as Grade 1 neuronal and mixed neuronal -glial tumor usually seen in children and young adults presenting with seizures which are often intractable, complex partial or generalized.[1-5]

The most common location is temporal or frontal. Parietal and especially occipital lobe involvement is rare. They have also been reported in cerebellum, pons and basal ganglia. The importance of identifying this rare entity lies in the favourable outcome with complete surgical resection without any need for chemoradiotherapy.[1]

## CASE REPORT

A 10-year-old male presented with history of generalized Tonic-clonic convulsions for one month in August 2014. On clinical examination, no deficit was found. CT Scan brain revealed right parietal hypodense lesion. MRI brain showed circumscribed macrolobular cortical subcortical tumor involving Right parietal lobe 4.5cm×4.2cm×5.4cm, hypointense on T1W and heterogeneously hyperintense on T2W Images with few intralesional cyst like components (pseudocyst) without contrast enhancement or mass effect and surrounding edema. Radiologically it was Diagnosed as DNET.

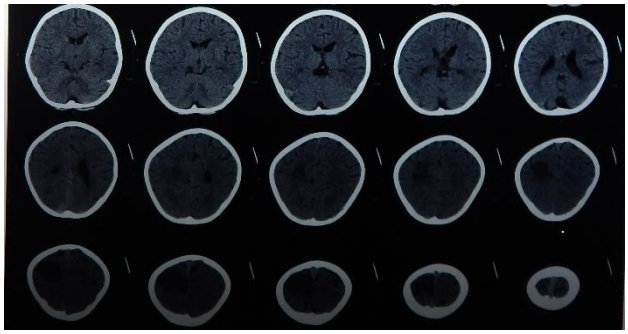

**Figure 1: Plain CT brain.**

Patient was given Phenytoin and advised surgery but patient failed to follow up for 2 years. He presented again in July 2016 with medically intractable seizures and was given phenytoin, Sodium Valproate, Levetiracetam and midazolam infusion.

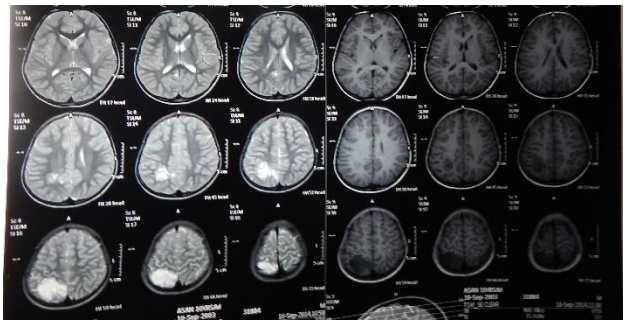

**Figure 2: T1W and T2W MRI 2014.**

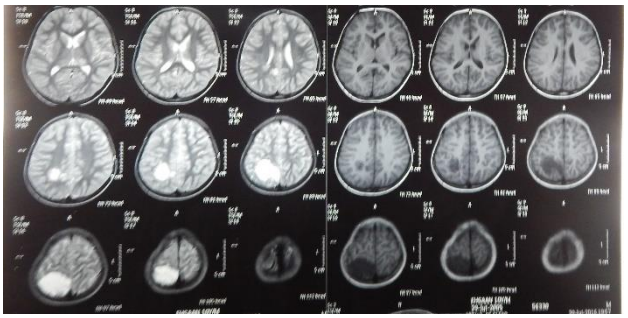

**Figure 3: T1W and T2W MRI 2016.**

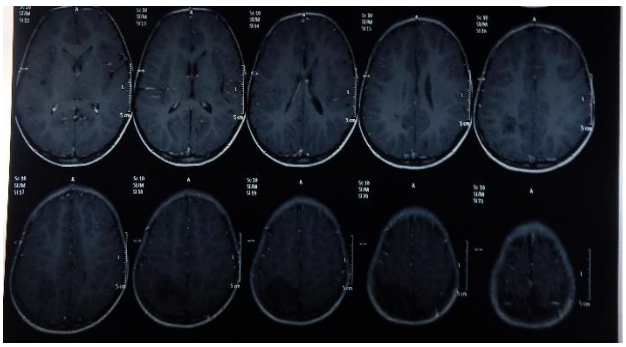

**Figure 4: Contrast MRI Pre-operative.**

Repeat MRI revealed no significant interval changes. MR Spectroscopy revealed minor elevation in Lipid-Lactate range (0.9-1.4ppm), no abnormal signal intensity of Choline peak and Creatinine –NAA peaks marginally reduced. He underwent right parietal craniotomy and microsurgical total excision of tumor on 30-08-2016 Grossly tumour was Greyish white, gelatinous, friable with clear demarcation from surrounding normal cortex. Histopathologically, section showed intracortical lesion composed of Oligodendroglia like cells, large neurons, focal microcystic areas with mucinous matrix. Findings were diagnostic of DNET (WHO Grade 1). Patient has been under regular follow up since then and seizures are under control.

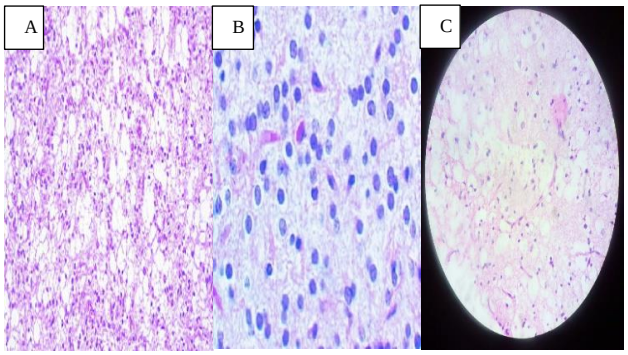


**Figure 5: Histopathology pictures.**

## DISCUSSION

The term dysembryoplastic neuroepithelial tumor was proposed by Daumas-Duport et al, 1st described in 1988.[1] Originally thought to have dysembryogenetic origin from secondary germinal layer but debate continues about their true nature.[6] Malignant transformation and regrowth following subtotal resection supports the hypothesis of neoplastic origin.[7-9]

It is a benign Supratentorial tumor characterized by its intracortical location, multinodular architecture and heterogenous cellular composition occurring in young patients with medically intractable seizures. DNET ´s are more commonly seen in temporal lobe.[1-4] Parietal and especially occipital lobe involvement is rare. Other rare sites include Caudate Nucleus, Septum Pellucidum, Brainstem and Cerebellum.[10] Occasional multifocal DNETs are associated with Neurofibromatosis, XXY syndrome and Intradural spinal lipoma.[11] Males are more commonly affected than females, usually in 6-20 years' age range.[10] Patients typically present with longstanding medically intractable epilepsy, usually complex partial or generalized Tonic-clinic. Raised ICP and neurological deficit are uncommon which may suggest malignant transformation.[10,12]

Our patient was 10 year old boy who initially presented with GTCS and was put on phenytoin. MRI revealed DNET for which surgery was advised but patient failed to follow up. After 2 years, he came with medically intractable seizures and repeat MRI showed that lesion remained same, emphasising two important facts of Natural history of these lesions

- Growth of these tumors remains stable over extended period of time. However malignant transformation has been reported rarely.
- Seizures become medically refractory over period of time.

Radiologically, conventional diagnostic criteria include supratentorial cortical especially temporal location without any sign of mass effect, peritumoral edema or tumoral enhancement. On CT, DNETs are typically well demarcated, hypodense, cortical lesion that can be

associated with deformation of overlying skull.MR images often show solid cystic mass with cystic portion appearing slightly more hyperintense than CSF. The solid components often appear multinodular, hypointense on T1W Images, hyperintense on T2W Images and occasionally weakly enhancing.[13-14] Present case showed all typical features except for atypical location in parietal lobe. Atypical features including mass effect and peritumoral edema with contrast uptake as well as multifocal lesion are reported in literature.[15] Three histological forms of DNET have been described: Simple, Complex and non specific.[6] The hispathological hallmark is bundles of axons lined by oligodendroglia-like cells, forming columns in a pale mucoid matrix in which isolated neurons float. Glioneural elements are seen both in simple and complex DNET. Heterogeneity of latter is due to additional glial or neuronal cell population which mimic low-grade gliomas. Present patient presented with typical features including glioneural elements.

The principal differential diagnosis of DNET are Oligodendrogliomas and Gangliogliomas. Typically, the diagnosis is established by pathology. Absence of floating Neurons with GFAP and synaptophysin negativity can rule out oligodendroglioma. Gangliogliomas are positive for GFAP and synaptophysin but radiologically present as cystic mass with mural nodule. They show neoplastic ganglion cells and perivascular lymphocytic infiltrate.[16]

Best course of treatment is complete surgical resection which provides long term seizure control.

## CONCLUSION

DNETs are clinically benign, non-recurring tumor of children and young adults. It is important to accurately diagnose DNET by multidisciplinary approach with contribution from clinician, radiologist and pathologist to reach definite and conclusive diagnosis so that surgical cure by complete excision may be achieved. Recurrence and malignant transformation are rarely reported, hence emphasing need for long term follow up.

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