Dysembryoplastic Neuroepithelial Tumour (DNET): A Rare Brain Pathology Presenting with Intractable Seizures

YASER-UD-DIN HOTI, KHURRAM ISHAQUE, AMIR AZIZ
Bashir Ahmed, Tariq Salah-ud-Din
Department of Neurosurgery Unit II, Lahore General Hospital, Lahore

ABSTRACT

Dysembryoplastic neuroepithelial tumour (DNET) is a benign glioneuronal tumour often related with intractable localization – related seizures in both in paediatric age group as well as young adults. However, a small number appear to have the potential for malignant transformation. Total surgical removal without any adjuvant therapy is the gold standard treatment. We report a case in which DNET appeared in a 13 year old male. Cranial CT scan showed a discoid lesion with peripheral enhancement in right parietal lobe. Micro-decompression of the tumour was done. On histopathology, the tumour revealed features of WHO grade I dysembryoplastic neuroepithelial tumour.

Key Words: *Dysembryoplastic neuroepithelial tumour (DNET), glioneural, malignant, brain tumors.*

Abbreviations: *DNET = Dysembryoplastic neuroepithelial tumour.*

INTRODUCTION

Dysembryoplastic neuroepithelial tumor (DNET) is an infrequent low grade, mixed neuronal and glial tumour, usually occurring in young adults and is accompanied with pharmacologically intractable, complex partial or generalized seizures (Daumas – Duport C. et al, 1988, Burger PC, Scheithauer BW, 1994). It was first described in 1988 (Daumas – Duport C, Lantos PL 1997). The favoured sites for these tumours are the **temporal** or **frontal lobes;** though **parietal lobe** association is also known other sites are less likely. More often, DNETs patients are asymptomatic clinically and radiologically for several years. Gross appearance of DNETs shows **mucinous** or **gelatinous multinodular lesions** of very **friable** consistency while microscopically all DNETs display **multiple intracortical nodules of variable sizes**. It is commonly considered as a fundamentally **benign lesion with total resection** being the treatment of choice **without any adjuvant therapy** (Prayson RA, Estes ML, Morris HH 1993). The identification of DNET has therapeutic and prognostic implications because aggressive therapy can be avoided, thus sparing these young patients of the harmful long term effects of radio- or chemotherapy.

CASE REPORT

Thirteen years old boy presented to us with 4 months history of 5 to 6 episodes of sudden onset generalized tonic clonic seizures accompanied with unconsciousness, frothy discharge from the mouth and urine incontinence for a period of 2 weeks. This was followed by weakness of left side of body. The epileptic fits were not responding to the medical therapy (valproic acid 500 mg thrice daily and levetiracetam 500 mg twice daily). After about 2 weeks, the patient presented in neuro emergency department in an altered state of consciousness and vomiting. On examination he was found to have left sided hemiparesis (grade – 3/5 power). Vitals were stable and other systemic examination was normal. His pupils were equal and reactive bilaterally. Both fundii appeared normal. Patient was thoroughly investigated. His cranial CT scan showed right parietal discoid hypo dense non-contrast enhancing lesion. Based on the symptomatology his MRI brain was done which revealed a hypo intense lesion in the right mesial temporal lobe with extension to the parietal area on T_1 weighted non-contrast images. This lesion on post contrast MRI study showed faint focal, punctate and ring – enhancement foci. On T_2 weighted

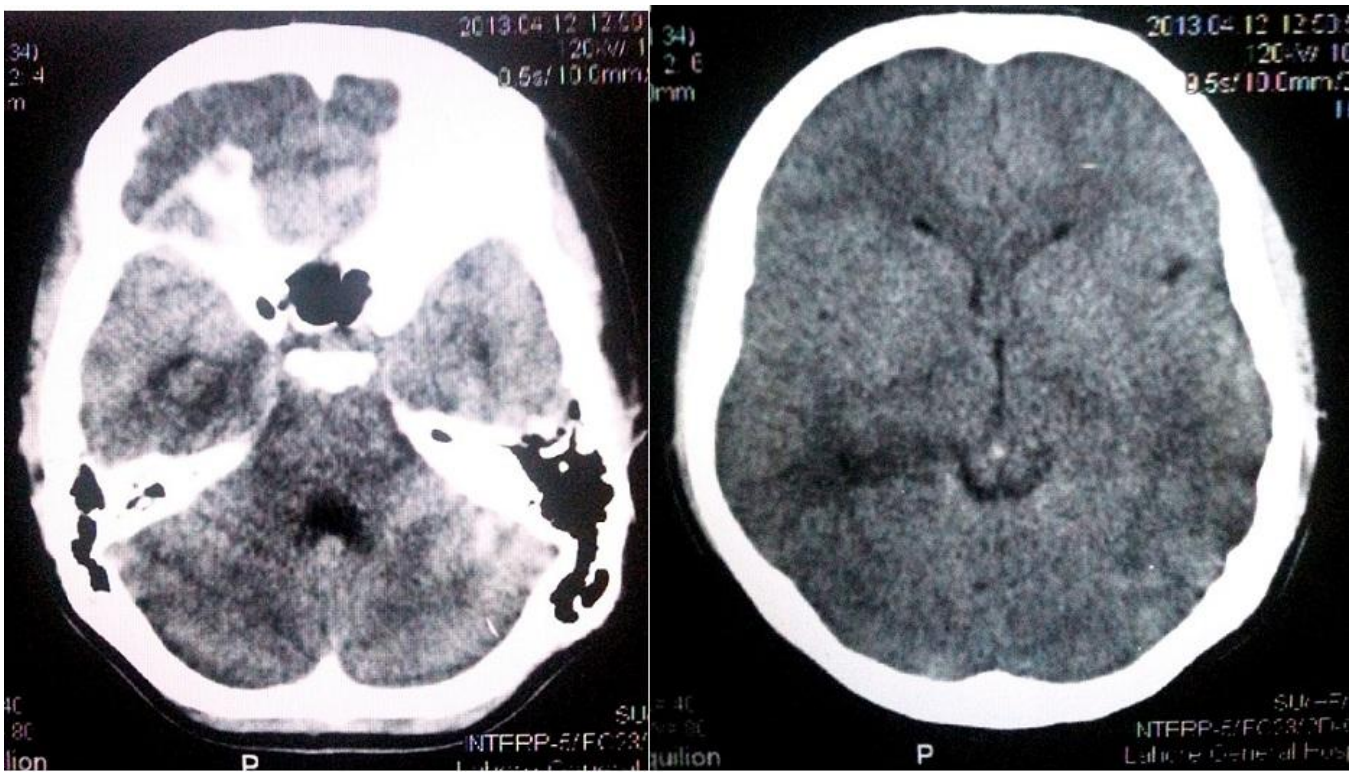

Figure 1: *CT scan Brain showing a hypo dense lesion in the right temporo parietal area.*

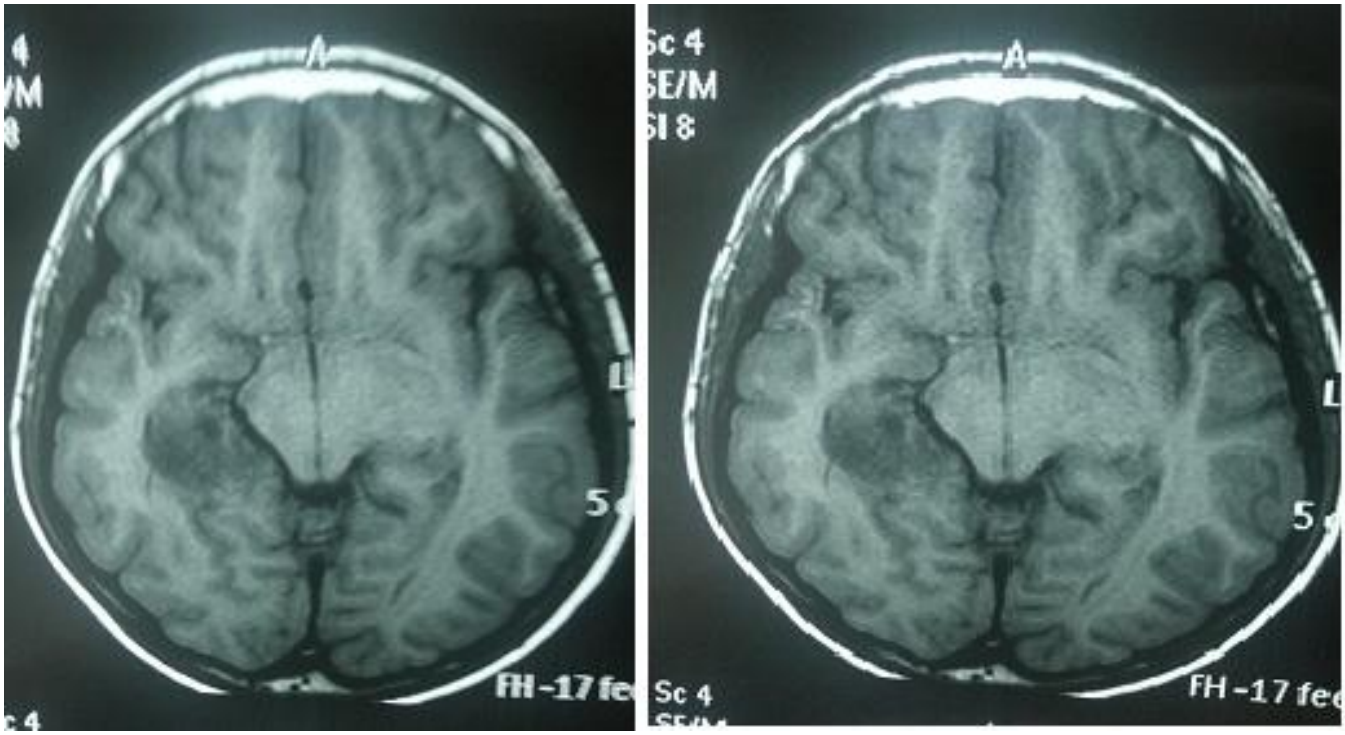

Figure 2: *Pre-operative T_1 Wieghted images showing hypointense area in to the right mesial temporal area.*

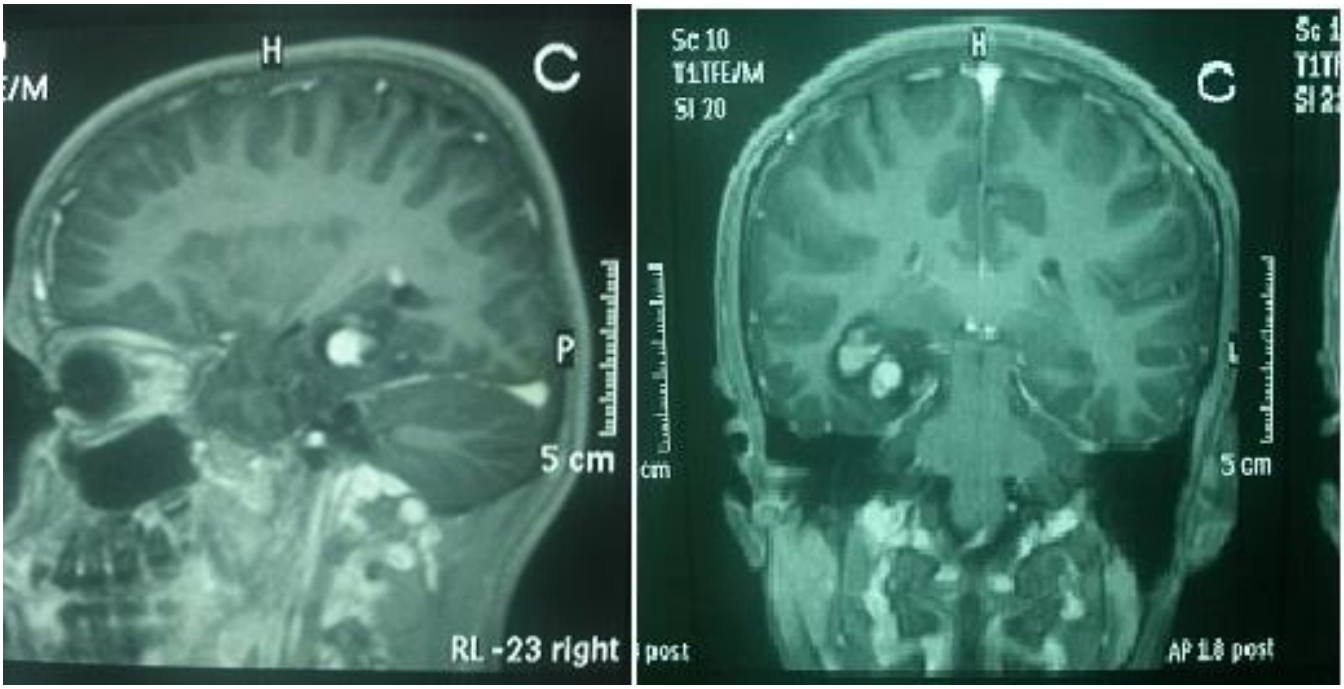

Figure 3: *T_1 weighted post contrast images showing areas of hyperintensity.*

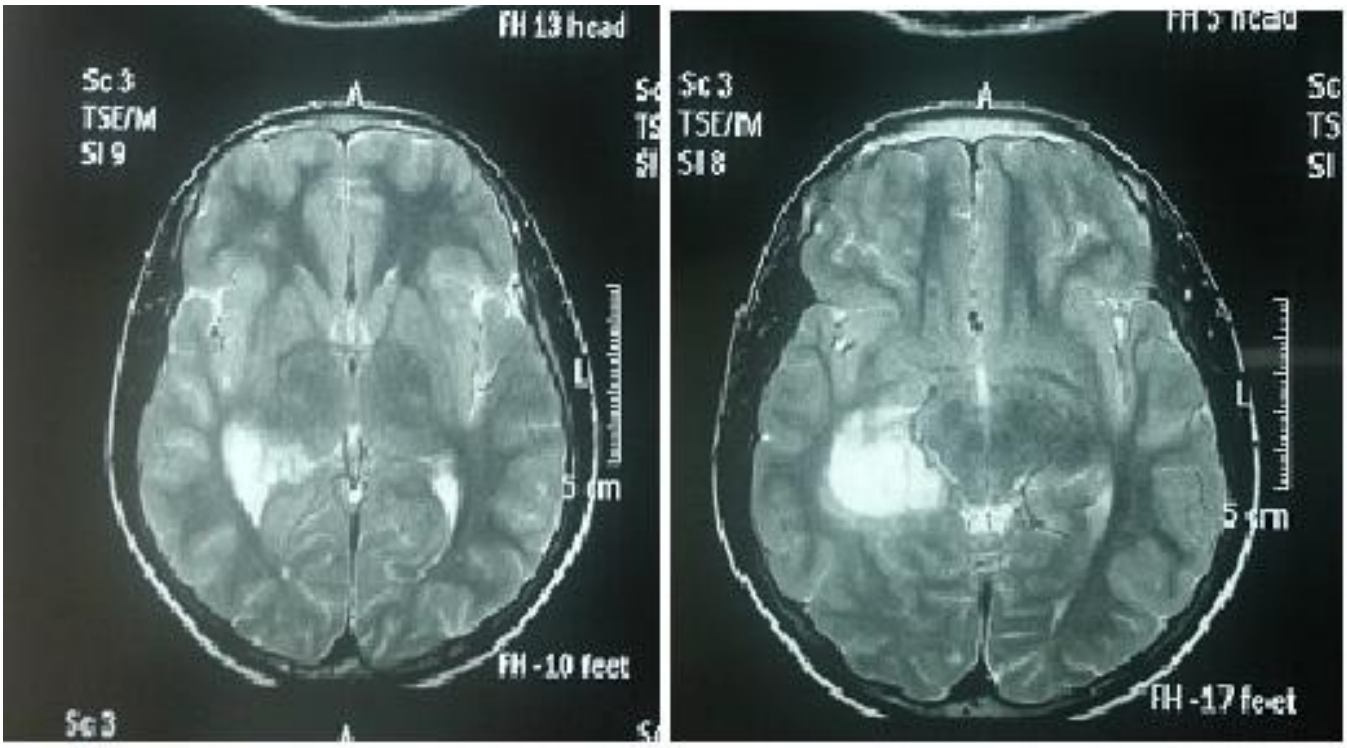

Figure 4: *T_2 weighted images showing hyperintense lesion, Right mesial temporal region.*

images the lesion appeared as hyper-intense with surrounding edema. On the basis of history, examination and radiology the first assumed diagnosis was that of oligodendroglioma.

After anaesthetic evaluation and informed consent, the patient was offered a right fronto parietal cranio-

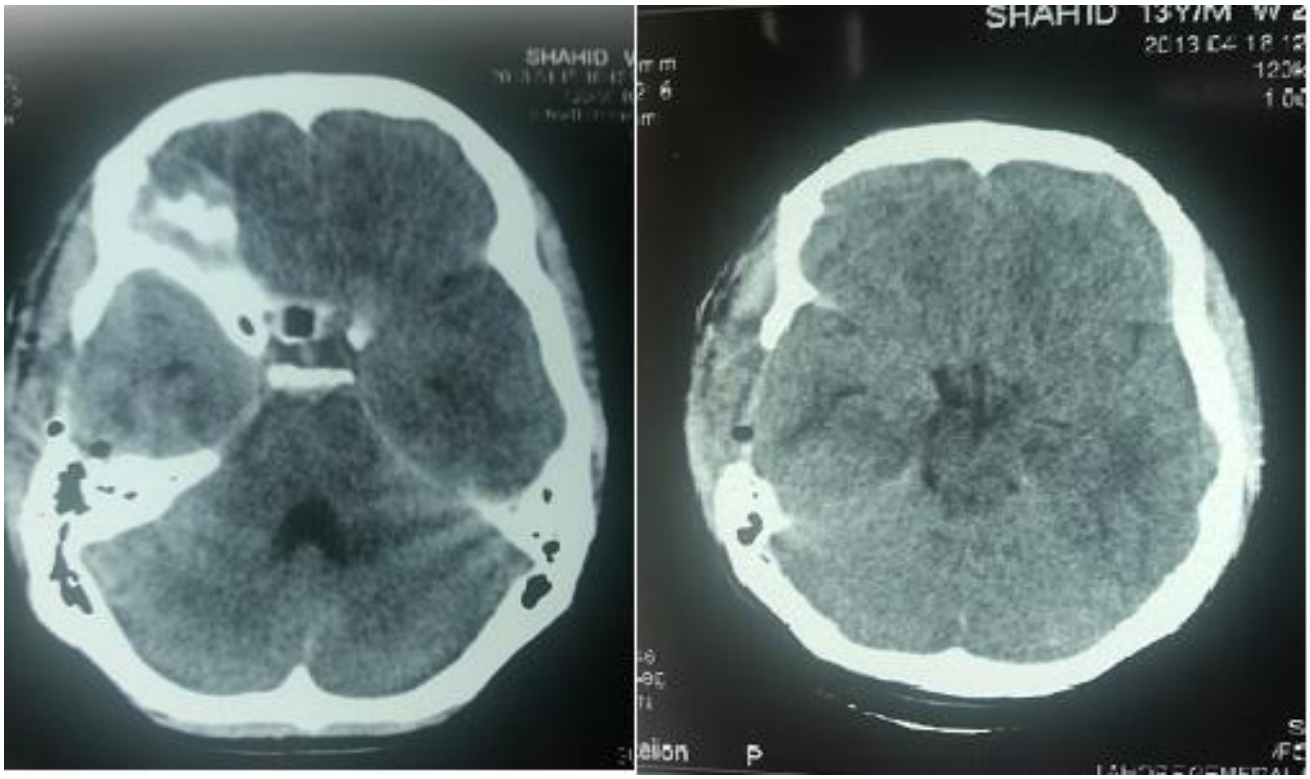

Figure 5: *Post-operative CT scan showing total removal of tumor.*

Figure 6: *Post-operative status.*

tomy with micro-decompression under G.A. Small lesion, **almost 1 cm in size, with numerous flecks of dirty grey colour were found in sub-arachnoid space in right temporoparietal area particularly in the mesial temporal lobe. The lesion was excised completely and sent for histopathology. The biopsy report showed specific glioneuronal element (SGNE), columns of heterogeneous cells oriented perpendicular to cortex. Oligodendrocyte – like cells arranged around capillaries, typical of DNET.**

Post-operative the patient recovered very well, his left sided weakness improved markedly and the power of the right upper limb became 5/5. Initially anti-convulsant were given for a period of 2 weeks and then gradually tapered off in 1 month time duration. The patient was followed up regularly every 2 weeks for 2 months and was seizure free without medication.

DISCUSSION

The term dysembryoplastic neuroepithelial tumor was first suggested by Daumas – Duport, et al, in 1988. This tumour was initially thought to have a dysembryogenetic origin, but argument still remains about their true nature (Chan CH, Bittar RG, Davis GA, et al, 2006).

According to WHO classification of tumours (2000), DNETs are classified in the category of neuronal and mixed neuronal glial tumors, consistent with Grade I (Daumas – Duport C, Pietsch T, Lantos PL, 2006). It is a benign supra-tentorial tumour having intracortical location, multinodular architecture and **heterogeneous** cellular configuration presenting in young patients with epileptic seizures not controlled by medical therapy. The temporal lobe is the most common site (Daumas – Duport C, et al, 1988, Prayson RA, Estes ML, Morris HH 1993). Nevertheless, these tumours can occur in other areas of the CNS because of their alleged origin in secondary germinal layers. Recent case studies have recognized existence of **DNET in caudate nucleus and other subcortical regions including cerebellum and brain stem, corresponding to the topography of secondary germinal layers** (Argyropoulos MI, et al, 2001, Kurtkaya – Yapicier O, et al, 2002, Cervera – Pierot P, et al, 1997).

Our patient was a 13 years old male who presented with a lesion in right temporoparietal area with a history of generalized tonic clonic seizures. Grossly, DNETs are **mucinous or gelatinous multinodular lesions of very friable consistency** (Daumas – Duport C, Lantos PL, 1997). Microscopically, all DNETs exhibit **multiple intracortical nodules of varying size.** The principal differential diagnoses of DNETs are **oligodendrogliomas** and **gangliogliomas**.

In our case, the diagnosis was established based on the histopathological findings (confirmed on review) and clinical data. In our case the neuroimaging features were typical of DNET showing enhancement and edema (Ostertun B, Wolf HK, Campos MG, et al, 1996) as consistent findings have been reported in literature. As DNETs occur in young patients and their course of progression is mostly benign, surgery forms the back bone of treatment thus avoiding side effects of adjuvant treatment. **However, recent reports have shown malignant transformation** in histologically proven DNETs (Josan V, Smith P, Kornberg A. et al, 2007). This points to partly understood natural history and clinical behaviour of this entity; as such these patients should be put on **lifelong follow-up.**

CONCLUSION

Surgery is the main treatment for **mesial temporal lobe tumours**, though invasive procedure require expertise. Total tumour resection is highly recommended for control of long-term epilepsy and tumour recurrence. The mesial temporal lobe tumours are heterogeneous in their prognosis but DNET has usually a relatively **favourable prognosis** without any radiotherapy or chemotherapy.

Address for Correspondence:
Dr. Yaser-ud-Din Hoti
Department of Neurosurgery Unit II
Lahore General Hospital, Lahore
Cell: 0345-9517899

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AUTHORS DATA

Name	Post	Institution	E-mail
Dr. Yaser-ud-Din Hoti	Senior Registrar	All at Department of Neurosurgery, PGMI / AMC and Lahore General Hospital, Lahore	yhanwer@yahoo.com
Dr. Khurram Ishaque	Senior Registrar		khurrumishaque@yahoo.com
Dr. Amir Aziz	Assistant Professor		dramiraziz247@yahoo.com
Dr. Bashir			basirakhan@yahoo.com.pk
Prof. Tariq Salah-ud-Din	Professor of Neurosurgery		tariqsalahuddin@hotmail.com