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Multifocal Dysembryoplastic Neuroepithelial Tumour: A Case Report and Review of Literature

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Abstract: Dysembryoplastic neuroepithelial tumour (DNET) is an unusual brain tumour occurring commonly in younger age groups. We report a case of medically resistant epilepsy in a young male that was radiologically diagnosed of having the typical features of DNET at multiple locations. The clinical features and imaging findings of the multifocal DNET has been discussed along with review of literature.
Keywords: DNET, MR, Epilepsy.

INTRODUCTION

DNET are benign slow growing tumours corresponding to grade I in WHO classification of CNS tumours. It is characterized as mixed neuronal-glial tumour in the current WHO classification and arises either from cortical or deep gray matter. Other CNS tumours having mixed neuronal-glial origin are ganglioglioma, paraganglioma and central neurocytoma [1]. DNET clinically presents as focal or generalized seizures in a child. DNET was first proposed by Daumas-Duport in 1988 [2]. Leung et al. first reported multifocal DNET in 1994 [3]. To the best of our knowledge, this is the ninth case of DNET having multifocal origin.

CASE REPORT

An 18yr old male Indian child came to the neurology outdoor with a complaint of recurrent episodic involuntary movements for 2 years, which was not controlled, on anti-epileptic medication. The seizures were usually generalized without any post-ictal deficit. The patient showed no obvious abnormality in physical examination and his developmental milestones were not delayed. His prenatal, intranatal and post-natal history was unremarkable. No other family member had a similar history.Routine blood examinations were within normal limits. On MRI (Magnetic Resonance Imaging) multiple lesions of varying sizes were found in right frontal region (Fig-1), tectal plate (Fig-2) and right thalamic region. The larger lesions had a typical bubbly cystic appearance with expansion of the overlying cerebral gyri without any significant mass effect. The lesions appeared hypo intense on T1 weighted images, heterogeneously hyper intense on T2 weighted images, show neither diffusion restriction on DWI (Fig-3) nor enhancement on post-contrast study (Fig-4). The clinical and radiological features were consistent with a diagnosis of multifocal dysembryoplastic neuroepithelial tumour. The patient continued on anti-epileptic medication and neurosurgical consultation was sought for further management.

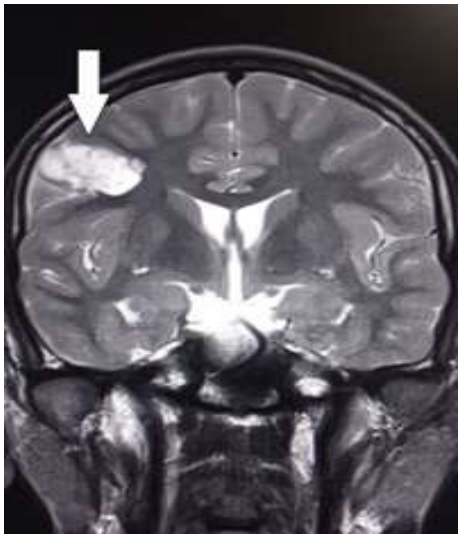

Fig-1: Coronal T2WI section of the brain shows a bubbly cystic appearing mass lesion expanding the gyrus at the right frontal region (white arrow)

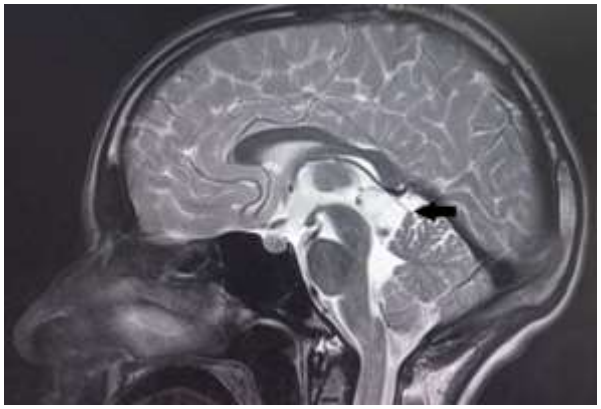

Fig-2: Sagittal T2WI section of the same patient shows another bubbly cystic mass lesion (black arrow) expanding the tectal plate

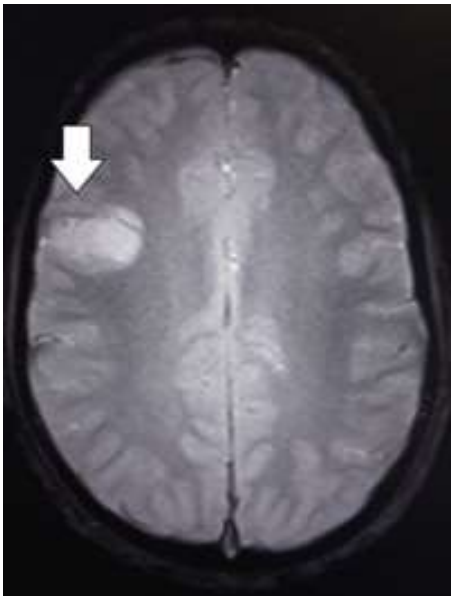

Fig-3: Axial DWI sectional the level of the right frontal lesion shows no significant diffusion restriction (white arrow)

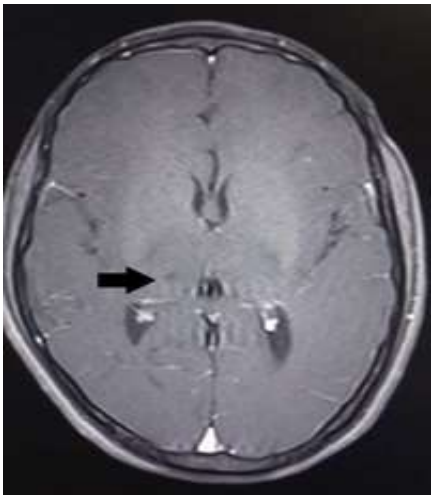

Fig-4: Axial T1C+ image at the level of the thalami shows no significant enhancement of a lesion present in right thalamus (black arrow)

DISCUSSION

DNET have a benign course and an excellent prognosis. The precise pathology is unknown. It is thought to be a hamartomatous malformation consisting of normal neuronal and glial components with a multinodular architecture. However there have been reports of malignant transformation as well as regrowth following subtotal resection, which favours a neoplastic nature of DNET. There have been three histological forms of DNET namely 1. Complex, consisting of specific glioneuronal elements with glial nodules and a multinodular architecture, 2. Simple consisting of specific glioneuronal elements only and 3. Non-specific that has the same clinical and neuroimaging features of complex DNET but no specific glioneuronal elements [4].

The temporal lobe is the most common location, however any part of the CNS can get effected, namely frontal lobe, caudate lobe, cerebellum and pons [5].

Although localized DNET is reported, multifocal DNET is extremely rare with previously only 8 cases reported in literature. Both sporadic as well as syndromic cases of multifocal DNET have been reported [6]. In our case we could not identify any specific syndrome, thereby suggesting a sporadic variety of multifocal DNET. Surrounding areas of cortical dysplasia are usually associated with multifocal DNET [7].

DNET are typically seen as a cortically based lesion with hardly any perilesional edema. On MRI they appear hypo intense on T1weighted, hyper intense with a bubbly appearance on T2 weighted and mixed signal intensity with bright rim sign on FLAIR (fluid-attenuated inversion recovery) sequences. Only 20-30% of the DNET shows heterogeneous or mural enhancement [4]. No restriction on DWI and non-specific MRS are usually seen. However MRS may contribute in diagnosis by showing a no elevation of choline to creatine ratio, normal creatine peak and a low N-acetylaspertate peak [8].

In our case the imaging findings were very much suggestive of a diagnosis of multifocal DNET, as reported earlier in literature. On detailed inter-departmental discussions and patient party counseling, a decision of conservative medical approach was taken without the need of any biopsy or surgical resection at present. The patient to the best of our knowledge is doing well at present and is kept on follow up.

CONCLUSION

Multifocal DNET is an extremely rare condition causing intractable seizures in young adults. Due to its multifocal presence, medical therapy remains the mainstay of treatment. Due to the indolent nature of the lesion, biopsy is not always recommended. As a

radiologist, the main aim should be to follow the progression of the existing lesions and appearance of any new lesion.

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