CASE REPORT

Dysembryoplastic Neuroepithelial Tumor: A Rare Brain Tumor Presenting with Atypical Radiological Findings

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Abstract

Dysembryoplastic neuroepithelial tumor (DNET) is a benign glioneuronal tumor frequently associated with intractable localization-related seizures in children and young adults. Complete surgical resection without any adjuvant treatment remains the treatment of choice. The authors present a case in which DNET occurred in a 35 year old female. CT scan of the brain revealed left parietal para-falcine discoid lesion with peripheral enhancement. Micro-decompression of the tumor was performed. Histologically, the tumor exhibited features of WHO grade I dysembryoplastic neuroepithelial tumor.

Key Words

DNET, Tumor

Introduction

Dysembryoplastic neuroepithelial tumor (DNET) is a rare low-grade, mixed neuronal and glial tumor, usually seen in young adults and associated with pharmacologically intractable, complex partial or generalized seizures (1-4). The first description of this entity dates back to1988 (1). The favored locations for these lesions are the temporal or frontal lobes; though parietal lobe involvement is also documented. Other sites are very rare. Usually the lesions are clinically and radiologically stable for years. Grossly, DNETs are mucinous or gelatinous multinodular lesions of very friable consistency and microscopically all DNETs exhibit multiple intracortical nodules of varying size. It is generally regarded as an essentially benign lesion with complete resection being the treatment of choice without any need for chemotherapy and/or radiation therapy (1, 5). The identification of DNET has therapeutic and prognostic implications because aggressive therapy can be avoided, thus sparing these young patients of the deleterious long term effects of radio- or chemotherapy.

Case Report

35 year old female presented with three months history of 6-7 episodes of sudden onset generalized tonic clonic seizures associated with loss of consciousness, frothing and incontinence of urine. This was followed by weakness of right side of body. She landed in emergency wing of our hospital where on examination she was found to have right sided hemiplegia (grade-0 power). Vitals were stable & other systemic examination was normal. Patient was thoroughly investigated. CT scan of brain revealed left parietal para-falcine discoid lesion (Figure 1 and 2). She was subjected to left fronto parietal craniotomy with micro-decompression under general anesthesia on 17/08/06. Small half mm. several specks of dirty grey color were found in sub-arachnoid space in left para falcine area around cerebral vein. Microscopically grayish pink & fleshy growth seen deep in white & grey matter extending at about an area of 2.5 X 2cm. moderately vascular. Veins were thrombosed. Histopathological Examination revealed grossly a few grayish white bits of brain tissue. Microscopically the sections showed fragmented tumor bits comprising of large neurons and oligodendroglia like cells (OLC), focal microcystic

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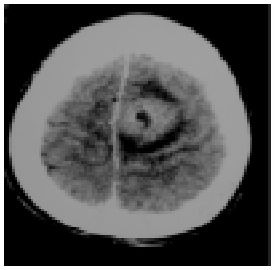
change and a mucinous matrix (Figure 3). Findings were diagnostic of Dysembryoplastic neuroepithelial tumor (WHO Grade 1).



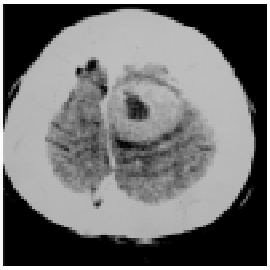


Figure 1 and 2: CT scan of brain showing a thick walled enhancing lesion with central necrosis and peri-lesional edema seen in left high para-sagittal region

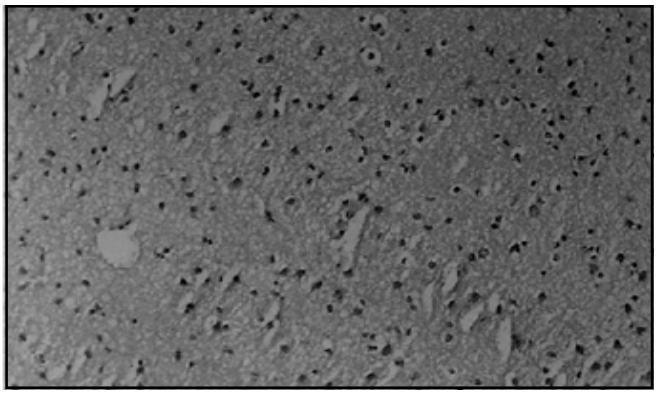
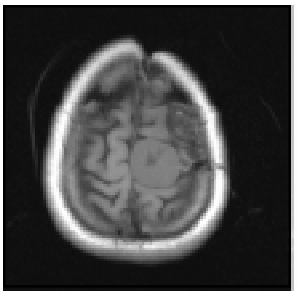


Figure 3: Photomicrograph of histopathological specimen of brain tissue (HE X 100) showing features of DNET.

Patient was referred to Department of Radiation Oncology for further management. She was admitted in ward for evaluation & management. Post-operative CT scan done on 15/12/06 revealed well circumscribed spherical enhancing lesion seen in left high parietal cortical region with adjacent non enhancing hypo density (gliotic changes).Impression of residual mass was made. Patient was discussed amongst doctors of concerned specialties and patient was put on close follow-up. MRI of brain done in June 2007 revealed a well defined lesion in high left parietal region (para-falcine lesion) which is iso-intense on T1 and T2 weighted images with few signal void areas within it. Also there is central hyper intense area on T2 weighted images and hypo intense area on T1 weighted images within the lesion. Peri-lesional edema is also noted (Figure 4, 5). Patient presented for last follow-up in June 2007 and is doing well.



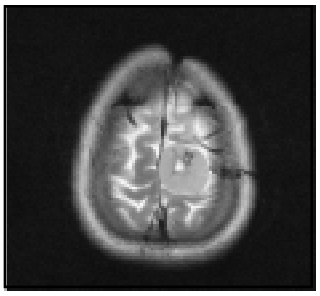


Figure 4 and 5: MRI brain showing left high parietal iso-intense lesion with few signal void areas and peri-lesional edema on T1 and T2 weighted images

Discussion

The term dysembryoplastic neuroepithelial tumor was proposed by Daumas-Duport et al (1). These lesions were originally thought to have a dysembyogenetic origin, but debate still continues about their true nature (6). According to WHO classification of tumors (2000), DNETs are included in the category of neuronal and mixed neuronal glial tumors, corresponding to Grade I (7). It is a benign supratentorial tumor characterized by its intracortical location, multinodular architecture, and heterogeneous cellular composition occurring in young patients with medically intractable epileptic seizures with the temporal lobe being the most common site (1-5). However, these tumors can occur in other areas of the CNS because of their putative origin in secondary germinal layers. Recent case studies have documented existence of DNET in caudate nucleus and other sub-cortical regions including cerebellum and brain stem, corresponding to the topography of secondary germinal layers (8-11). Our patient was a 35 years old female who presented with a lesion in left parietal para-falcine area with a history of generalized tonic clonic seizures. Macroscopically, DNETs are mucinous or gelatinous multinodular lesions of very friable consistency (2) .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. Though, in our case the neuroimaging features were not typical of DNET, enhancement and edema (12) as inconsistent findings have been reported in literature. As DNETs are

seen in young patients and their behavior is mostly benign, surgery forms mainstay of treatment thus avoiding side effects of adjuvant treatment. However, recent reports have shown malignant transformation in histologically proven DNETs (13). This points to incompletely understood natural history and clinical behavior of this entity; as such these patients should be put on life long follow-up.

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