

Case report

Dysembryoplastic Neuroepithelial Tumor with Intractable Epilepsy: A case Report and Review of the Litreature

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ABSTRACT

The dysembryoplastic neuroepithelial tumor (DNET) is a newly recognized brain tumor with distinctive clinicopathological features and a favourable prognosis. All DNTs are benign supratentorial tumor most commonly located in the temporal lobe and usually cause no neurologic deficit. Clinically, DNT is usually associated with chronic intractable epilepsy in adolescents and young adults. DNET common age of presentation range from 119- year.

Key words: Dysembryoplastic neuroepithelial tumor, intactable epilepsy, child, neurosurgery Sudan.

Eight years old girl presented with intractable generalized tonic clonic convulsion with loss of consciousness since the age of one and half year. The condition was diagnosed as epilepsy, started on cocktail of antiepileptic drugs without response.

On examination the patient was fully conscious; there was no evidence of neurological deficit, no signs of increase intracranial pressure and no stigmata of neurocutaneous malformation.

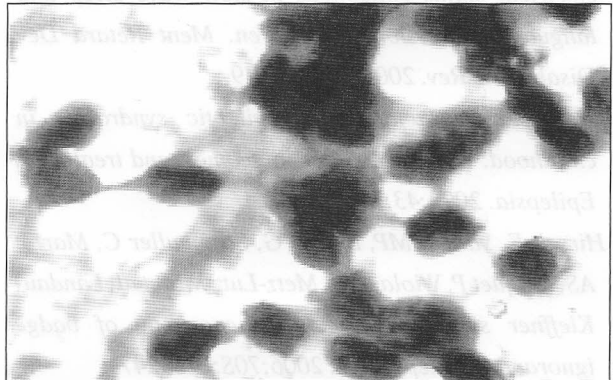
Investigations: MRI showed a mass occupying the right frontal lobe fig (1)

On surgery it is proved to be a cystic mass. A histopathology and cytology showed fibrillary astrocytoma grade II fig (2,3)

One month postoperative the

patient referred to paediatric oncology, where another MRI was done and the report showed featurer of dysembryerioplastic neuroepithelial tumor (DNT).

Figure (2)
cytology showing fibrillary astrocytoma grade II



A paediatric neurooncologist at St . Jude children research hospital (USA) was asked for a second opinion and he confirmed the diagnosis of DNT. The patient was referred to the neurosurgeon and planned for frontal lobectomy yet.

Discussion:

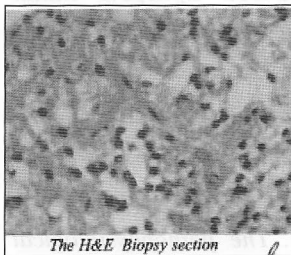


Figure (3) histopathology showing
fibrillary astrocytoma grade II

The dysembryoplastic neuroepithelial tumor (DNT) is a newly recognized brain tumor with distinctive clinicopathological features and a favorable prognosis. Regarding control of the tumor itself, as well as control of the associated epilepsy. The term

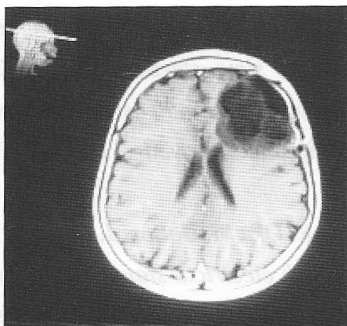


FIGURE (1) MRI brain
showing a mass occupying
the right frontal lobe

dysembryoplastic neuroepithelial tumor (DNT) was introduced by Daumas-Duport et al in 1988 (1). Clinically, DNT is usually associated with chronic intractable epilepsy in adolescents and young adults. Common age of presentation range from 119- year. (Mean age 9 year). All DNTs are benign supratentorial tumor most commonly located in the temporal lobe (62%), frontal lobe (31%), (1) and usually cause no neurologic deficit.

Macroscopically the tumor is obvious at cortical surface. On cut section the gross appearance of the tumor is variable with some of them are well demarcated while the other are poorly demarcated. Some were soft; some were firm and some with cystic changes (1).

Histopathologically, DNT is a benign multinodular lesion composed of glial and neuronal elements (1, 2–3). DNTs share several important features with gangliogliomas and glioneuronal malformations, such as glioneuronal hamartomas and hamartomas (small glioneuronal malformations) (4). These consist of glial cells and highly differentiated ganglion cells and are frequently associated with chronic focal epilepsy.

The pathological diagnosis was based on the revised WHO classification of brain tumors [5]. This classification describes DNT as a histological variant of neuronal and mixed neuronogial tumors characterized by the following criteria: cortical location, multilobular architecture – the nodule being made of multiple variants looking like astrocytomas, oligodendrogliomas or oligoastrocytomas, foci of dysplastic cortical disorganization and the presence of a glioneuronal element showing a columnar structure perpendicular to the cortical surface. According to Mischel's criteria, identification of the presence of cortical dysplasia was based on nine microscopic abnormalities: cortical laminar disorganization, single heterotopic white matter neurons, neurons in the cortical molecular layer, persistent remnants of the sub-pial granular cell layer, marginal glioneuronal heterotopia, polymicrogyria, white matter neuronal heterotopia, neuronal cytomegaly with associated cytoskeletal abnormalities, and balloon cell

change [6].

High-resolution MR imaging has significantly contributed to the recognition of low-grade tumors and tumor like lesions as a major cause of chronic focal epilepsy. Modern MR techniques with high resolution and high contrast permit detection of lesions measuring no more than a few millimeters in size, and because of this, MRI should be routinely used to examine patients

with chronic focal epilepsy. The neurora-diologic characterization of such lesions is essential for the treatment of these patients because it aids in identifying those who are likely to benefit from epilepsy surgery. Following hippocampal sclerosis, benign tumors are the second most frequent brain lesions that are associated with epilepsy (7). The high prevalence of gangliogliomas in patients with chronic focal epilepsy contrasts with the fact that they constitute only 0.4% to 1.3% of all brain tumors in general neurosurgical series (8, 9).

The overall seizure free rate was 90% post operatively. Recurrence was reported in those who received chemotherapy or radiotherapy.(10)

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