Case Reports

Dysembryoplastic Neuroepithelial Tumor

L. Kasturi
A.V. Kulkarni
V.A. Mashankar
U.A. Desai

Dysembryoplastic neuroepithelial tumor (DNT) is a recently described pathologically benign unique tumor arising within the supratentorial cortex and having a 100% association with partial complex seizures(1). Due to the paucity of literature on this condition and its heterogeneous cellular composition, DNT can present difficulties in diagnosis and is often confused with glial neoplasms(2). We are reporting a case of DNT in a 15 year old girl who presented with intractable partial complex seizures and who recovered completely after surgical excision of the tumor.

Case Report

A 4 year old girl presented to us in 1983 with history of recurrent attacks of afebrile generalized tonic clonic seizures of recent onset. She was born normally at term. Her neonatal period was uneventful and developmental milestones were normal. The initial investigations including EEG and CT scan of the brain were normal. She was put on antiepileptics but was poorly controlled. The seizure pattern changed from grandmal to partial complex to atonic seizures (drop attacks) over the years. She would have a cluster of attacks (50-60/day) and then would be seizure free for 2-3 weeks. Her interictal neurological examination was normal. In view of poorly controlled seizures, she was reinvestigated at 9 years of age. Her CT and EEG were normal at that time.

In 1993, at the age of 14 years she was investigated again because of her refractory partial complex seizures. This time her EEG showed focal slowing intermittently (2 cycles per second) over left parasagittal area which was suggestive of a structural lesion in the left parasagittal area. CT scan of the brain at this time also was normal. At this stage, MRI of the brain was done which showed a 1 x 1.2 cm sized rounded non-enhancing lesion in the left parasagittal frontal cortex which could either be a neuropithelial tumor or a low grade glioma *(Fig. 1).* Surgical excision of the tumor was done and the histopathology was suggestive of DNT *(Fig. 2).* Postoperatively she received anticonvulsants for a period of

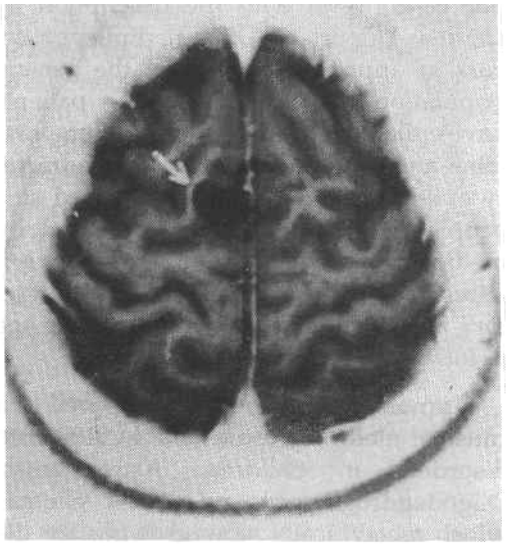

Fig. 1. MRI scans showing hypodense lesion in T1 weighted images.

From the Department of Pediatrics, BARC Hospital, Anushaktinagar, Bombay 400 094.

Reprint requests: Dr. L. Kasturi, 7-D, Everest, Anushaktinagar, Bombay 400 094.

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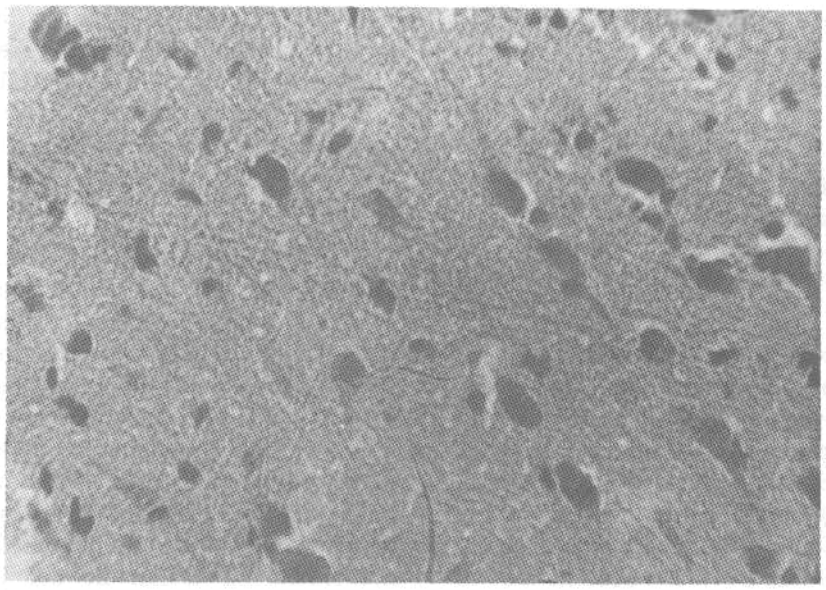

Fig. 2. Photomicrograph of DNT showing dysplastic ganglion cells in a fibrillary matrix (x 400)

one year and is at present asymptomatic and has no neurological deficit.

Discussion

The clinical and pathological profile of our case fits in with the description by Daumas-Duport as DNT(3). Epilepsy occurs in approximately 1% of the general population. One third of these patients have epileptogenic foci within the temporal lobes and of these about half have refractory epilepsy. DNT has been described as a neoplasm of childhood characterized by medically intractable partial complex seizures(3). Our case also presented as refractory partial complex seizures since the age of four years.

Tumors have been long recognized as a cause of medically intractable focal seizure disorders in children. Astrocytomas, Oligodendrogliomas and mixed gliomas which are typically slow growing are the most frequent epileptogenic tumors. DNT is a recently described pathologically benign unique tumor arising within the supratentorial cortex and having a 100% association with partial complex seizures. The lack of recognition of DNT as a clinicopathological tumor entity may be explained by the failure to detect them before the advent of modern imaging techniques and by the reluctance to consider surgery as an option for patients with medically refractory partial complex seizures(1).

Computed tomographic scanning reveals rounded hypodense lesions with varying degrees of contrast enhancement. Calcification may also be seen. Pronounced deformity of the overlying calvarium is seen when the tumor is situated superficially and is longstanding. The CT may be normal in 10% of patients (3,4) as exemplified by our case.

Magnetic resonance imaging demonstrates focal cortical mass in all patients. MRI discloses a well demarcated hypo-intense signal on T_1 weighted images

as well as hypersignal on T_2 weighted images. Some lesions may show enhancement with Gadolinium-DTPA. The margins of the tumor are well circumscribed and may remodel the adjacent calvarium(l). In our case, the MRI findings ware the same as described above. CT and MR differential diagnosis of DNT include low grade gliomas, arachnoid cysts and other forms of hamartomas(5). In cases of refractory partial complex seizures where CT is normal, MRI is indicated.

DNTs are pathologically benign and occur within the cortical regions of the brain typically in the temporal or frontal lobe. Parietal and occipital involvement is infrequent. This tumor has also been included in the new WHO classification of brain tumors and it is grouped under "neuronal and mixed neuronal glial tumors" (6). In most cases the patient's age at the onset of symptoms is less that 20 years.

The term DNT was proposed for these neoplasms because of multiple distinct cell lineages, early onset of clinical symptoms and associated presence of cortical dysplasia in most of the cases(5). Histologically, tumors are characterized by multinodular appearance with a predominant component of alveolar arrangement of oligo-dendroglial cells around delicate capillaries with mucoid matrix containing floating ganglion cells. It is hypothesized that DNT may be derived from second germinal layer which includes the dentate fascia, subpial germinal layer, subependymal germinal layer and external granular layer in the cerebellum. These matrix cells have the capacity to develop into both neurons and glial cells which can account for the divergent cell lineage in DNT(7).

The important differential diagnoses in view of the heterogeneous cytological composition are mixed oligoastrocytoma and ganglioglioma. The multinodular character and presence of cortical dysplasia are of particular assistance when the cytological features which distinguish DNT from these tumors are not entirely clear(8).

Identification of these neoplasms is of obvious therapeutic importance because it spares these young patients the deleterious long term effects of radio or chemotherapy. The characteristic clinical features of DNT include intractable partial complex seizures with onset before 20 years of age, lack of significant interictal neurological deficit and no stigmata of DNT(5). Patients with DNT have an excellent prognosis with surgical resection. In the study of Daumas-Duport, no recurrences were identified in 37 patients in follow up ranging from 1 to 18 years. Even if only partial resection was obtained, all the 39 patients in the previous study experienced at least a significant reduction in seizure frequency and usually were symptom free. Our patient also underwent total resection of the tumor and is seizure free since operation.

In conclusion, although DNT are rare tumors of the pediatric population, they should be considered in the differential diagnosis of neoplasms causing seizures. The successful histological identification of DNT, a surgically curable tumor is important because it precludes unnecessary radio and/or chemotherapy in these young patients. The lack of recognition of DNT as a clinicopathological tumor entity may be explained by the failure to detect them before the advent of modern imaging techniques and by the reluctance to consider surgery as an option for patients with medically refractory partial complex seizures.

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REFERENCES

1 Koeller K, Dillon WP. Dysembryoplastic neuroepithelial tumors. MR appearance. AJNR 1992,13:1319-1325.

2 Prayson RA, Estes ML. Dysembryoplastic neuroepithetial tumor. Am J Clin Pathol 1992,97:398-401.

3 Duport CD. Dysembryoplastic neuroepithelial tumor. Brain Pathology 1993, 3: 282-295.

4 Duport CD, Scheithauer BW, Chodkiemicz JP, Laws ER, Vedrenne C. Dysembryoplastic neuroepithelial tumor: A surgically curable tumor of young patients with intractable partial seizures. Neurosurgery 1988, 23: 545-556.

5 Vali AM, Clarke MA, Kelsey A. Dysembryoplastic neuroepithetial tumor as a potentially treatable cause of intractable epilepsy in children. Clinical Radiology 1993, 47: 255-258.

6 Keliheces P, Burger PC, Scheithauer BW. The new WHO classification of brain tumors. BrauvPathology 1993, 3: 255-268.

7 Leung SY, Gui E, Ng HK, *et al.* Dysembryoplastic neuroepithetial tumor- A tumor with small neuronal cells resembling oligodendroglionia. Am J Surg Pathol-1994,18: 604-614.

8 Raymond AA, Halpin SFS, Alsanjari N. *et al.* Dysembryoplastic neuroepithelial tumor: Features in 16 patients. Brain 1994, 117: 461-475.