

Dysembryoplastic neuroepithelial tumor in the third ventricle with mesial temporal sclerosis: An unusual association

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Abstract

We report a patient with medically refractory complex partial seizures in whom neuroimaging revealed a dual pathology, an intraventricular lesion which was histopathologically proved to be a dysembryoplastic neuroepithelial tumor (DNET) with a co-existing right mesial temporal sclerosis (MTS). Occurrence of this tumor in the ventricle is rare. This is the first case reported from India to the best of our knowledge. The co-existence of intraventricular DNET and MTS also has not been previously reported. The co-existence may be another evidence that some cases of MTS are developmental and have a similar origin as DNET.

CASE REPORT

A 21-year-old male presented to us with refractory complex partial seizures of temporal lobe semiology since eight years of age. General physical and neurological examinations were normal. He underwent a presurgical evaluation for the seizures with a scalp-video electroencephalographic (S-VEEG) monitoring and a magnetic resonance imaging (MRI 1.5T). MRI revealed a well-defined mass in the anterior third ventricle and the foramen of Munroe with an associated MTS (Figure 1a,b). S-VEEG showed a right temporal ictal onset corresponding to the MTS.

The third ventricular tumor was resected through an interhemispheric, transcallosal approach. Histopathological examination of the resected tumor was diagnostic of DNET. (Figure 2a,b). The seizures persisted in the same frequency and semiology postoperatively. The patient was later subjected to right anterior temporal lobectomy with amygdalohippocampectomy. The histopathology showed hippocampal sclerosis without any associated dysplasia. The patient was seizure free at six months follow-up.

DISCUSSION

DNETs are a pathologically distinct subset of tumors commonly associated with medically refractory epilepsy when it is located in the

supratentorial cortex. They usually occur within dysplastic cortex and tend to affect the temporal lobes most commonly. However extracortical location of this tumor is also rarely seen. Baisden *et al*¹ reported a series of 10 low-grade neoplasms arising in the midline anteriorly in the region of the septum pellucidum with histological features of DNET. These tumors extended into the lateral ventricle from the septal region obstructing the foramen of Munroe with varying degrees of hydrocephalus. Onguru *et al*² reported a single DNET in the lateral ventricle, which was removed through a similar trans-callosal approach as in our case. Komori *et al*³ described a rosette forming glioneural tumor of the fourth ventricle and Guesmi *et al*⁴ reported a DNET, which expanded in the lateral ventricular septum and fornix. The occurrence of DNET in the ventricle has not been previously reported from the Indian subcontinent. The recognition of DNET with an unusual location like in the third or lateral ventricle has therapeutic and prognostic significance, as they are benign and one can avoid unwarranted and even deleterious treatments such as radiotherapy misdiagnosing them as gliomas. An origin from secondary germinal layers as previously suggested can explain these extra cortical locations.¹

Although, there have been no previous reports of the association of intraventricular DNET and MTS, several authors have reported the association of MTS with cortical dysplasia or dysgenesis.^{5,6}

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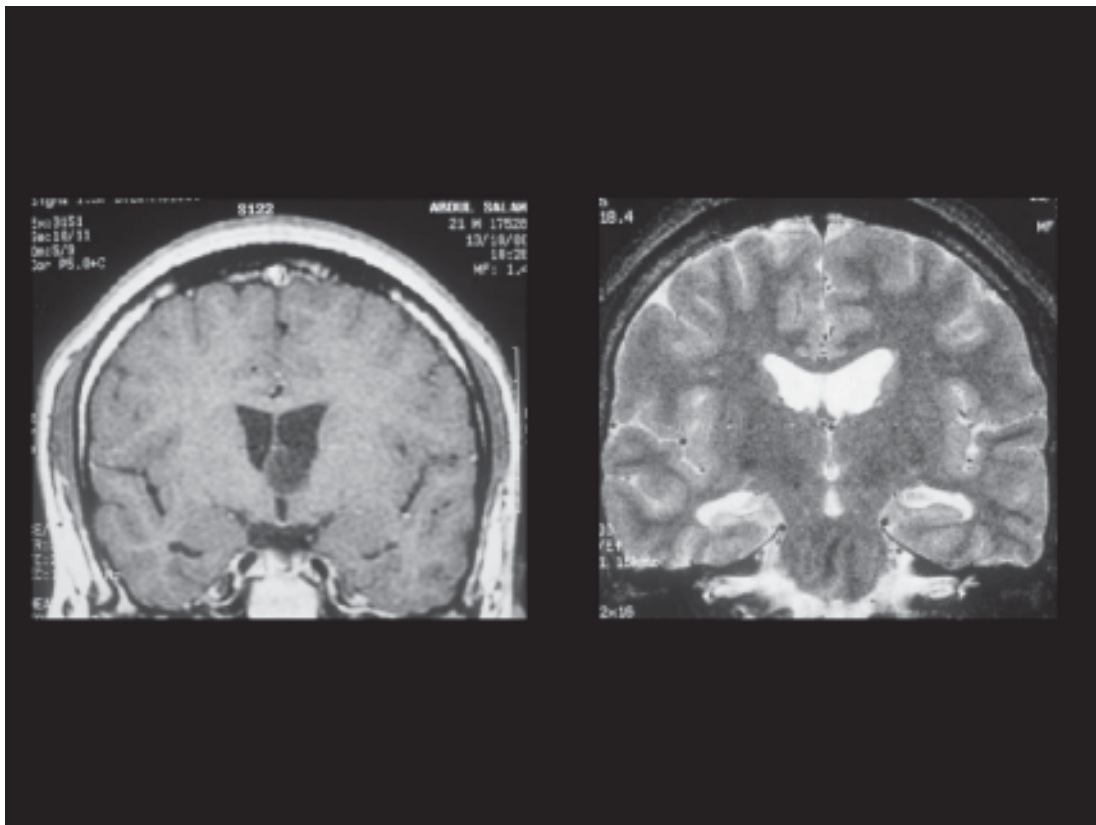


Figure 1a: Contrast enhanced T1 weighted coronal MRI shows a non enhancing hypo intense mass lesion occupying the region of anterior third ventricle and the foramen of Monroe. 1b Coronal T2 weighted spin echo sequence at the level of interpeduncular cistern shows hyper intensity and volume loss of right hippocampus suggestive of hippocampal sclerosis

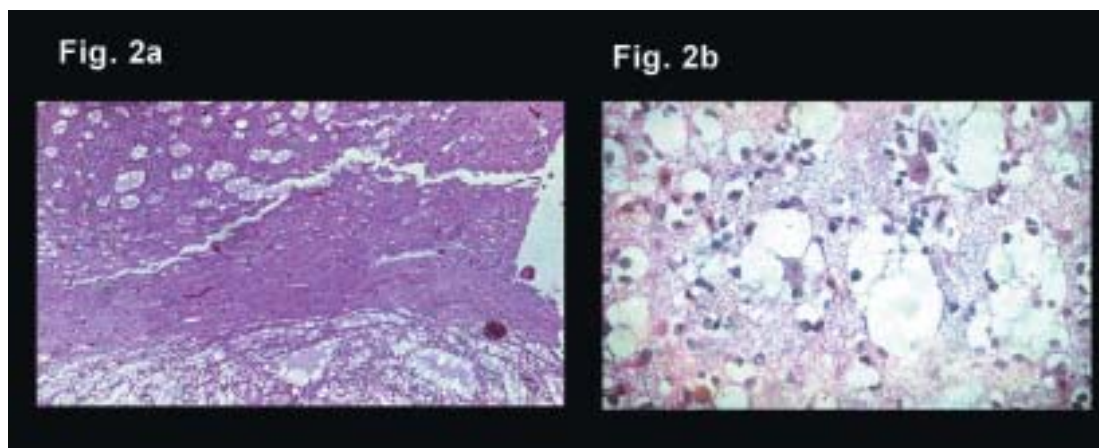


Figure 2a and b: Photomicrographs showing characteristic micro cystic spaces containing pale staining material. Some of these micro cystic spaces contain well-formed neuronal cells-floating neurons. In between these sheets glial cells are also seen. (Haematoxylin and eosin stain X200).

Others have also postulated that some cases of MTS may be developmental i.e. a form of “cortical dysgenesis”, as exhibited by the duplication of the dentate gyrus and abnormal orientation and shape of the pyramidal cells in some cases of MTS.^{7,8} DNET frequently occur in “dysplastic cortex” and the possibility that DNET themselves are a form of cortical dysgenesis has been previously reported.⁹ Therefore an association between MTS and this dysgenetic tumor is plausible, rather than a chance finding.

This case further broadens our understanding of dual pathology and is yet another piece of evidence that some cases of MTS may be developmental and have a similar origin as DNET.

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