

Dysembryoplastic Neuroepithelial Tumor : CT and MR Findings A Case Report¹

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Dysembryoplastic neuroepithelial tumor(DNET) is a recently described rare tumor that occurs most frequently in the temporal lobe of the brain and is characterized by long-standing, intractable complex partial seizures in children. The authors experienced one case of DNET occurring in a 13-year old boy, who had refractory complex partial seizure for 7 years. CT scan revealed nonenhancing low density mass in the left temporal lobe. MR images demonstrated a well-marginated cortical mass with very low signal intensity on T1WI and multinodular appearance of high signal intensity on T2WI. A few small enhancing foci within the mass were noted on contrast enhanced MR images. DNET, a rare tumor, should be considered in the differential diagnosis of neoplasm which causes seizure and is distinguished from other tumors because of its benign course. Differentiation between DNET and other tumors by CT and MR findings is very difficult. But, our case showed the multinodular pattern on T2W image, which may be helpful feature in the differential diagnosis.

Index Words : Brain neoplasms, CT
Brain neoplasms, MR
Children, central nervous system

INTRODUCTION

The authors report a case of dysembryoplastic neuroepithelial tumor(DNET) occurring in a child with chronic seizure disorders. To date, only a few studies concerning DNET have been published(1-4).

DNET is a new entity of glial tumor proposed by Daumas-Duport et al(1) and has been described as a benign neoplasm usually occurring in young male associated with intractable complex partial seizure(CPS) and arising within the cortical regions of the brain, typically in the temporal lobes. Since DNET is a new classification and curable by excision, correct diagnosis of this tumor is important. There may be con-

siderable difficulty in distinguishing this neoplasm from other neoplasms, especially low-grade astrocytomas and ganglioglioma.

We describe the characteristic radiologic, pathologic, and clinical features of DNET and discuss the difficulties in differential diagnosis.

CASE REPORT

A 13-year-old boy was admitted to our hospital with 7 years history of complex partial seizure that was not controlled by medical therapy. The initial work-up included a CT scan that was obtained with 9800 Hilight (GE, Milwaukee, WI, USA) and followed by MR scan which was performed with 1.5T(GE Signa, Milwaukee, WI, USA) using 256 × 192 matrix. MR sequences included T1-weighted(T1W) axial, sagittal, and coronal images (433/11/2, TR/TE/excitation) and fast spin echo (FSE) T2-weight(T2W) images (3500/85/2). Following gadolinium(Gd)-DTPA(Magnevist, Shering, USA) injection, axial, coronal, and sagittal T1W SE images were obtained. The CT scan revealed a nonenhancing hypodense mass in the left temporal lobe(Fig. 1a). MR images showed a well-marginated mass in left temporal lobe with low signal intensity on T1W image and

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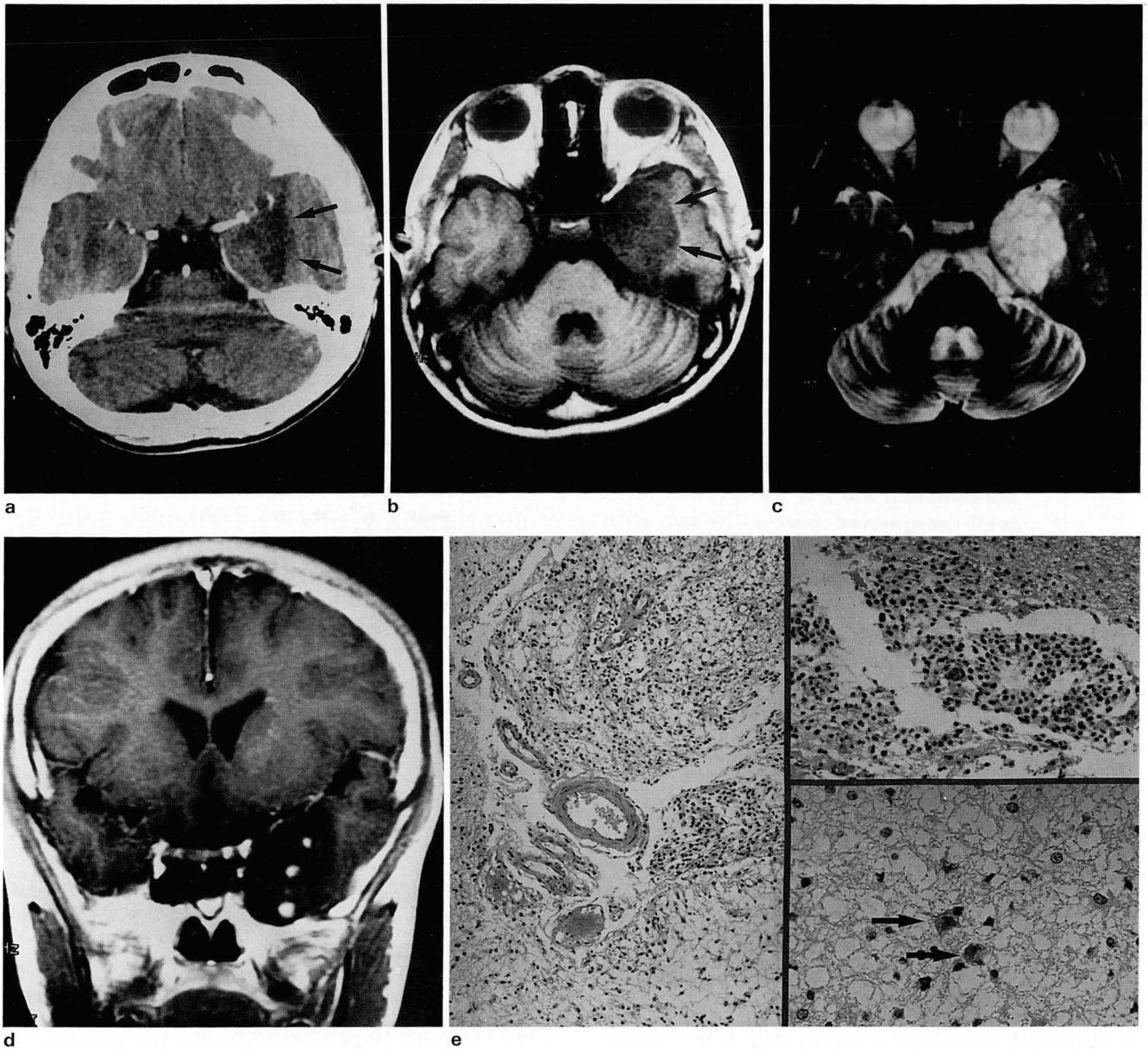


Fig. 1. Dysembryoplastic neuroepithelial tumor in 13-year-old boy

- a. Post-enhanced axial CT image shows a ill-defined nonenhancing hypodense mass in the left temporal lobe(arrows).
- b. T1 weighted axial MR image reveals a well-margined diffuse low signal intensity mass in the left medial temporal lobe(arrows).
- c. T2 weighted axial MR scan demonstrates a well-defined and diffuse high signal intensity mass and shows fine mutiseptated multinodular appearance within the mass at the left medial temporal lobe.
- d. Contrast enhanced coronal MR image shows small punctate enhancing foci within the well margined diffuse low signal mass. Smooth margined bony erosion is seen at the greater wing of left sphenoid bone.
- e. Microphotograph(H&E) demonstrated lobulated pattern of tumor with variable glial cellularity(left, $\times 33$), aggregation of small round oligodendrocytes(right upper, $\times 50$), and two abnormal neurons (arrows)(right lower, $\times 100$).

multinodular appearance of high signal intensity on T2W image(Fig. 1b & c). On coronal T1W enhanced image, small punctate enhancing foci were identified within the diffuse hypointense mass and smooth margined bony erosion was well demonstrated at the greater wing of left sphenoid bone (Fig. 1d). The patient underwent subtotal resection of the left temporal lobe. Histopathologic findings of the surgical specimen were characterized by intracortical location of the tumor with

multinodular architecture, and heterogeneity in cellular composition, astrocytes, oligodendrocytes, and neurons. The final pathologic diagnosis was DNET based on the presence of a unique specific glioneuronal elements(Fig. 1e).

DISCUSSION

Epilepsy occurs in approximately 1% of the general

population. One third of these patients have epileptogenic foci within the temporal lobes and, about half of these are medically refractory(5).

Pathologic study has revealed that mesial temporal sclerosis is the most common abnormality in patients with medically refractory temporal lobe epilepsy(6). Focal lesions of the temporal lobe occurs in about 24% of cases. These include gliomas(52%), nonspecific cerebral injury secondary to infection or trauma(24%), vascular malformations(9%), hamartomas(7%), non-glial tumors(5%), and tuberous sclerosis(6). Dumas-Duport *et al* (1) reviewed the pathologic diagnosis of over 265 patients, all of whom had operation for medically refractory complex partial seizures, and reclassified 39 cases as DNET.

DNET is a benign tumor frequently associated with medically refractory CPS. Males were more commonly affected than females. The lesions were usually located in the supratentorial cortex, the temporal lobe (62%) and the frontal lobe(31%) but rarely in the parietal and occipital lobes(1). Our case showed the mass at the left temporal lobe. It is hypothesized that these tumors may arise from the secondary germinal layer of the central nervous system, particularly the subpial granular layer. Remnants of the subpial granular layer have been found in the temporal and frontal lobe regions of normal infants, which correlates well with the most frequent locations of these tumors(3).

The term "dysembryoplastic neuroepithelial tumor" was proposed for these neoplasms because of the presence of multiple and distinct cell lineages, the early onset of clinical symptoms, and the associated presence of cortical dysplasia in most cases(3).

Dumas-Duport *et al*(1) reported that CT images showed "psuedocystic" well-demarcated, low density mass and some cases were associated with focal contrast enhancement(18%) or calcific hyperdensity(23%) but it may in fact be normal in 10% of cases. In our patient, CT scan revealed a homogenous low density lesion, however, it could not demonstrate the true extent, margin, and enhancing foci within the mass.

On MRI, the lesion had prolonged T1 and T2 relaxation times and demonstrated well-marginated low signal intensity on T1W images and high signal intensity on T2W images. The majority of these lesions showed minimal or no enhancement with Gd-DTPA. Koeller *et al*(2) reported that two of six cases of DNET showed the minimal enhancement. Our case showed small multiple enhancing foci within the hypointense mass. However, we found multinodular internal pattern on T2W images, which is not described as an usual pattern.

The MR findings of DNET may demonstrate a similar appearance to ganglioglioma or low-grade astrocytoma with a focal mass that is almost always hypointense on T1W images and hyperintense on T2W images. Especially, ganglioglioma is very similar both clinically and radiologically. Ganglioglioma occurs in children with intractable seizure and usually arise in the temporal lobe. The classic appearance on CT is a cyst with an hypodense mural nodule which is often calcified. MR findings of ganglioglioma are also non-specific, a well-delineated mass with hypointense on T1W images, hyperintense on T2W images, and variable enhancement pattern(7, 8). Histology of DNET show more useful features in distinguishing it from the other two conditions(3). DNET exhibit a high degree of cellular polymorphism and show at least one of three characteristics: a specific glioneuronal element displaying minimal cytologic atypia, a multinodular and multicystic component and an association with cortical dysplasia(1).

In summary, DNET is a pathologically benign neoplasm which is usually located in temporal lobe. The MR appearance may mimic that of ganglioglioma or low-grade astrocytoma with the presence of focal cortical lesion, hypointensity on T1W images, and hyperintensity on T2W images in patients who have a history of CPS with onset prior to young adult. However multinodular pattern within the mass on T2W MR images may suggest the diagnosis of DNET.

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태생기 발육부전 신경상피종: 전산화 단층촬영 및 자기공명영상소견 1예 보고¹

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태생기 발육부전 신경상피종 (Dysembryoplastic neuroepithelial tumor, DNET)은 최근에 소개된 드문 종양으로 소아에서 대뇌의 측두엽에 잘 발생하며, 난치성의 장기간 지속되는 복잡부분발작이 특징인 질환이다. 본 저자들은 13세 남아가 내과적으로 치유되지않는 복잡 부분발작이 7년간 지속되어 전산화 단층촬영(CT)과 자기공명영상(MRI)을 시행하고 수술하여 DNET 로 진단 받은 1예를 보고하고자 한다. 본 예에서 이 종양의 CT 소견은 조영증강되지않는 저음영의 병변으로 보였고 MRI 에서는 경계가 좋은 종양이 좌측 측두엽에 있었으며 T1강조영상에서 저신호 강도로 T2 강조영상에서는 소결절 다발모양의 고신호강도로 보였으며 조영 증강 후에는 조영증강이 된 몇개의 작은 점들이 종양내에서 보였다. 이 DNET는 드문 종양이기는 하나 발작을 일으킬수있는 다른 종양과 감별되어야 하는데 그 이유는 이 종양이 양성과정을 취하기 때문이다. DNET는 영상 진단기기인 CT와 MRI상 비슷한 소견을 보이는 다른종양과 감별이 잘되지 않는 것으로 보고되어 있으나 저자들의 예에서 다른 보고들의 예들에서는 없었던 multinodular pattern을 T2 강조영상에서 보여주고 있으므로 차후에 감별진단하는데 도움을 줄수있으리라고 생각된다.