Dysembryoplastic Neuroepithelial Tumor : A Case Report

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Abstract : **We report here a classic case of dysembryoplastic neuroepithelial tumor (DNT) located in the right temporal lobe of a 12-year-old Thai boy presenting with visual, auditory and gustatory hallucinations for 4 years. DNT is an uncommon low-grade neuronal and mixed neuronal-glial tumor which has a characteristic clinical presentation, as well as characteristic radiological and histopathological findings. DNT has excellent prognosis and surgery is mainly curative treatment. Hence, it is important to recognize this entity in order to avoid unwarranted radio- or chemotherapy.**

Key words : Dysembryoplastic neuroepithelial tumor

เรื่องย่อ : **Dysembryoplastic Neuroepithelial Tumor : รายงานผู้ป่วย 1 ราย**

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รายงานผู้ป่วยเนื้องอกของสมองในผู้ป่วยเด็กชายอายุ 12 ปี ที่มาพบแพทย์ด้วยอาการชักมานานสี่ปี ผลการตรวจทางรังสีวินิจฉัยพบว่ามีก้อนเนื้องอกของสมองบริเวณกลีบสมอง temporal ด้านขวา ก้อนเนื้องอกได้รับการผ่าตัดออกและส่งตรวจทางพยาธิวิทยา การตรวจชิ้นเนื้อสมองพบว่าเป็นเนื้องอกชนิด dysembroyoplastic neuroepithelial tumor เนื้องอกชนิดนี้พบได้น้อย การวินิจฉัยโรคจำเป็นต้องอาศัยอาการแสดงทางคลินิก การตรวจพบทางรังสีวิทยา ร่วมกับลักษณะทางพยาธิวิทยา การให้การวินิจฉัยที่ถูกต้องมีผลต่อการรักษาและการพยากรณ์โรค ซึ่งได้ผลดีด้วยการผ่าตัด

INTRODUCTION

Dysembryoplastic neuroepithelial tumor (DNT) was first described in 1988 by Daumas-Duport et al. in a report of 39 cases[1]. In that series, all tumors were cortical lesions of mixed cellular composition. These included neurons, astrocytes and oligodendroglia-like cells (OLCs) with the so-called "specific glioneuronal element" (SGE). In the revised World Health Organization (WHO) histological tumor classification[2], this entity has been placed in the category of grade I neuronal and mixed neuronal-glial tumors. Unique presentations are children or young adults with a history of long-standing drug-resistant partial seizures, usually beginning before age 20, without neurological or stable congenital deficit. It is characterized by cortical lesions mostly occurring in the temporal and frontal lobes. DNT has excellent prognosis. Surgery is recommended therapy. Long-term clinical follow-up reveals no evidence of recurrence, even for patients with partial surgical removal. We report the following case:

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CASE REPORT

A 12-year-old Thai boy presented with visual, auditory and gustatory hallucinations for 4 years. The symptoms gradually increased in frequency despite full medication. Other neurological examinations were normal. The MRI demonstrated a right temporal lobe mass with hypointense signal on T1-weighted and hyperintense signal on T2-weighted images. (Figure) Gadolinium enhancement was almost absent. No peritumoral edema or mass effect was seen. There was no deformity of the overlying cavarium. A temporal lobe lobectomy was performed. Gross examination of the resected temporal lobe exhibited focal widening of the cerebral cortex by viscous tissue forming megagyri. (Fig) Serial sectioning disclosed a well-demarcated gelatinous appearance of the involved cortex and minute petechiae. Histopathologically, the tumor was made up of different cell lineages consisting of neurons, astrocytes and oligodendroglia-like cells (OLCs) embeded in microcystic mucinous matrix. OLCs hanging along axonal processes with floating neurons, characteristic of SGE, which is composed of bundle of axons perpendicular to cortical surface lined by OLCs with accompanying floating neuron in mucin-rich matrix, were observed. Multiple vague nodules composed of mainly OLCs and scattered astrocytes were also found. Neither necrosis nor mitosis was seen. The adjacent cortical dysplasia was not identified. In the immunohistochemistry study, the OLCs expressed for synaptophysin, neurofilament and S-100 protein. GFAP was positive on scattering astrocytes. After three months follow-up, the patient had obviously improved with regard to seizure control.

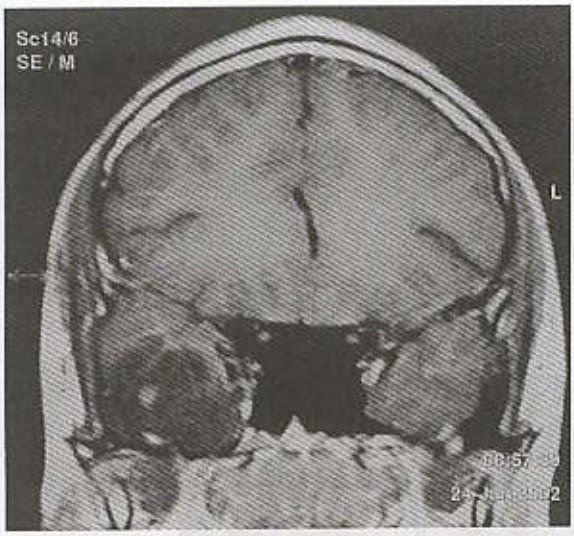

Figure 1. T2-weighted MR image showed a hyperintense lesion in the right temporal lobe. There is no peritumoral edema or mass effect.

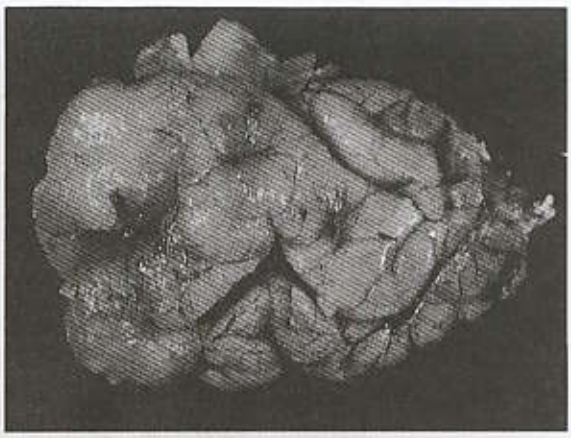

Figure 2. Grossly, the tumor presented as megagyri and gelatinous surface.

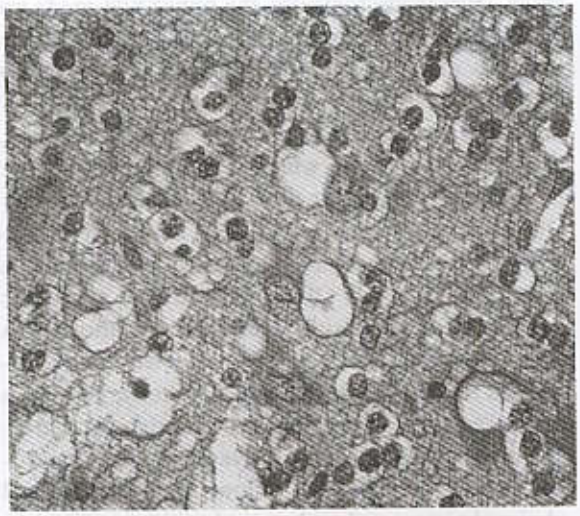

Figure 3. Oligodendroglia-like cells exhibiting a "fried egg" appearance and scattered neurons embedding in micromucinous matrix.

DISCUSSION

DNT is a benign glial-neuronal neoplasm occurring in children or young adults with a long-standing history of intractable partial seizures. All patients have normal intelligence quotients (IQ). The tumor is typically a supratentorial intracortical lesion, and is nearly always located in the temporal and frontal lobes. It can be found in the caudate, cerebellum and brainstem in rare case reports[3-5]. Computer topography (CT) demonstrates hypodense mass without evidence of mass effect or peritumoral edema. Deformities of the overlying bone or cranial fossa enlargement are often seen, and this finding further supports the diagnosis of DNT. MRI displays hypointense mass on T1-weighted image and hyperintense on T2-weighted image[6]. On pathological finding, the tumor expands the effected cortex, resulting in megagyri. Microscopically, it is usually made up mainly of OLCs and, to lesser extent, neu-

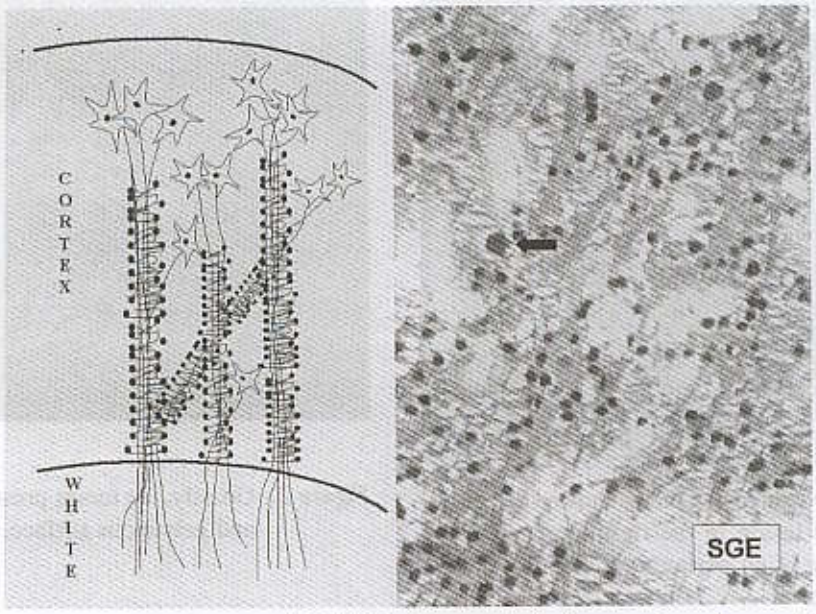

Figure 4. Specific glioneuronal element of DNT: the characteristic element is made up of OLCs attached to bundles of axons and neurons (arrow) floating in the interstitial matrix.

rons and scattered astrocytes. The presence of so-called "specific glioneuronal element" (SGE) is a hallmark. Two histologic variants of DNT have been recognized: a simple form, which displays only SGE, and a complex form (previously described in the original report) consisting of SGE, glial nodules and cortical dysplasia. The glial nodules comprise a heterogeneous appearance of oligodendrocytic, astrocytic and neuronal components forming nodular architecture. Morphologic atypia and vascular proliferation may be encounterd: Wide variation in neuronal population, from normal to dysplastic morphology, is also seen, though this does not indicate malignant potential[7].

Recently, a non-specific variant of DNT, the entity that has a similar clinical presentation, neuroradiological profile and failure to show growth in long-term follow-up by CT or MRI, has been proposed. However, since this lesion lacks glioneuronal component and multinodular architecture and is histologically indistinguishable from pilocytic astrocytoma, oligodendroglioma or oligoastrocytoma, the concept of a non-specific variant of DNT remains controversial[2].

OLCs have small round nuclei with perinuclear halos. The exact nature of these cells remains uncertain[8]. Many neuronal markers, including neuronal nuclei antigen (NeuN) and N-methyl-D-aspate-recepter subunit 1 (NR1), are demonstrated in these cells. This indicates neuronal differentiation, whereas GFAP-positive oligodendroglia-like cells in most DNT suggest astrocyte differentiation[9,10]. Even the immunoreactivity for myelin oligodendrocyte glycoprotein found in many OLCs[11], supporting oligodendrocyte differentiation, has been reported.

The differential diagnoses of DNT are other glial and neuronal tumors[12]. Generally, a large resected specimen of DNT, as presented in this report, has unique gross findings. The diagnosis is straightforward. In small biopsy, it is a diagnostic problem, since SGE is seldom recognized and OLCs may share a number of histopathological similarities with oligodendroglioma, low-grade astrocytoma, ganglioglioma, and ependymoma. Ganglioglioma is another low-grade neuronal-glial tumor that mimicks DNT in terms of clinical presentations and cortical location. Unlike DNT, it displays dense connective tissue and lymphocytic infiltration within the tumor and usually produces mass effect.

The management of DNT is surgery, and the indication for therapy is related to seizures. Long-term clinical follow-up reveals neither clinical nor radiological evidence of recurrence in the patients with complete or incomplete surgical removal. Radiotherapy and chemotherapy have no role[13]. Even in rare cases where malignant transformation is reported[14], DNT still has an excellent prognosis.

Here we report a classic case of dysembryoplastic neuroepithelial tumor (DNT). This entity should be recognized and distinguished from other aggressive glial and neuronal tumors. Clinically, the diagnosis of DNT should be considered whenever all the following criteria are present: 1) partial seizures with or without secondary generalization, usually beginning before age 20, 2) no progressive neurological deficit, 3) predominantly cortical topography of a supratentorial lesion, best demonstrated on MRI, and 4) no mass effect on CT or MRI, except if related to a cyst, and no peritumoral edema. On histopathological findings, the tumor displays heterogenous cellular composition including neurons, astrocytes and oligodendroglia-like cells and typical component "specific glioneuronal element" (SGE).

REFERENCES

1. Daumas-Duport C SB, Chowkiewicz JP, Laws ER, Vedrenne C. Dysembryoplatic neuroepithelial tumor: a surgically curable partial seizures. Neurosurgery 1988; **23:** 545-56.
2. Paul Kleihues WKC. Pathology and genetics of tumours of the nervous system. Lyon: IARCP Press, 2000.
3. K Kuchelmeister TD, E Schlore, M Bergmann, F Gullotta. Dysembryoplastic neuroepithelial tumour of the cerebellum. Acta Neuropathol (Berl) 1995; **81:** 418-27.
4. Pascale Cervera-Pierot PV, Jean-Paul Chodkiewicz, Catherine Daumas-Duport. Dysembryoplastic neuroepithelial tumors located in the caudate nucleus area: report of four cases. Neurosurg 1997; **40:** 1065-70.
5. Fugimoto K, Ohnishi H, Tsujimoto M, Hoshida T, Nakazato Y. Dysembryoplastic neuroepithelial tumor

of the cerebellum and brainstem. Case report. J Neurosurg 2000; **93:** 487-89.
6. Lee DY, Chung CK, Hwang YS, Choe G, Chi JG, Kim HJ, et al. Dysembryoplastic neuroepithelial tumor: radiological findings (including PET, SPECT, and MRS) and surgical strategy. J Neurooncol 2000; **47:** 167-74.
7. Daumas-Duport C. Dysembryoplastic neuroepithelial tumours. Brain Pathol 1993; **3:** 283-95.
8. Peter C.Burger BWS. Atlas of Tumor Pathology Tumors of The Central Nervous System. Washington, D.C.: Armed Forces Instituite Of Pathology, 1994.
9. Wolf HK, Buslei R, Schmidt-Kastner R, Schmidt-Kastner PK, Pietsch T, Wiestler OD, et al. NeuN: a useful neuronal marker for diagnostic histopathology. J Histochem Cytochem 1996; **44:** 1167-71.
10. Taratuto AL PH, Sevlever G, Gallo G, Monges G. Dysembryoplastic neuroepithelial tumour: morphological, immunocytochemical, and deoxyribonucleic acid analyses in a pediatric series. Neurosurgery 1995; **36:** 474-81.
11. Gyure KA, Sandberg GD, Prayson RA, Morrison AL, Armstrong RC, Wong K. Dysembryoplastic neuroepithelial tumor: an immunohistochemical study with myelin oligodendrocyte glycoprotein. Arch Pathol Lab Med 2000; **124:** 123-26.
12. Honavar M, Janota I, Polkey CE. Histological heterogeneity of dysembryoplastic neuroepithelial tumour: identification and differential diagnosis in a series of 74 cases. Histopathology 1999; **34:** 342-56.
13. Cabiol J, Acebes JJ, Isamat F. Dysembryoplastic neuroepithelial tumor. Critical Reviews in Neurosurgery 1999; **9:** 116-25.
14. Hammond R, Duggal N, Woulfe J, Girvin J. Malignant transformation of dysembryoplastic neuroepithelial tumor. Case report. J Neurosurg 2000; **92:** 722-25.