

# Dysembrioplastic Neuroepithelial Tumor: Cytological Diagnosis by Intraoperative Smear Preparation

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*The dysembrioplastic neuroepithelial tumor (DNT) is a benign and slow-growing temporal lobe cortical lesion that presents with clinically unresponsive partial and complex seizures, with onset before age 20 yr. DNT shows a multinodular architecture and the presence of mixed glioneuronal elements. The authors report on a case of DNT diagnosed by the intraoperative smear cytological technique. We describe the cytological characteristics of smear preparations of DNT, with emphasis on the differential diagnosis. The authors present a 5-yr-old male with partial seizures for the preceding 3 yr. The patient showed no other neurological deficits. Magnetic resonance identified a lesion in the cortical area of the right temporal lobe. The smear cytology showed small, round, oligodendroglial-like cells arranged in lobules, with microcystic spaces containing ganglion cells floating in a fine fibrillary background. Histology confirmed the diagnosis of DNT. This report shows that the smear preparation can be trustworthy for the intraoperative diagnosis of DNTs, helping to determine the appropriate neurosurgical procedure and thus avoiding extensive and unnecessary resections.* Diagn. Cytopathol. 2002;26:92-94; DOI 10.1002/dc.10042 © 2002 Wiley-Liss, Inc.

**Key Words:** dysembrioplastic neuroepithelial tumor; smear preparation; magnetic resonance

The dysembrioplastic neuroepithelial tumor (DNT) was described in 1988 by Daumas-Dupont et al.<sup>1</sup> and incorporated in the revised WHO classification in 1993<sup>2</sup> among the category of neuronal and mixed neuronoglia tumors. The clinical manifestations of DNT include partial and complexes seizures in a young population, refractory to the

usual drug therapy, with no further underlying neurological deficits. DNT is composed of small oligodendroglial-like cells aligned along the side of an eosinophilic, occasionally vascularized, core known as a specific glioneuronal element and immersed in an abundant mucinous background, with a somewhat microcystic appearance containing occasional floating nerve cells. Cortical dysplasia might also be seen in the adjacent cortex. The authors present a case report of DNT with intraoperative diagnoses made by cytological examination of a smear preparation.

## Case Report

A 5-yr-old male presented in September 1999 with partial seizures, with secondary generalization, for the preceding 3 yr. Physical examination showed a small reduction of the cognitive functions, but no other neurological abnormalities. Magnetic resonance showed a superficial right temporal lobe lesion compatible with a low-grade glioma. The patient underwent a craniotomy that demonstrated a well-delimited cortical temporal lobe lesion. Intraoperative diagnosis was made with 1 mm<sup>3</sup> of tumor tissue that was smeared between two glass slides and stained with 1% aqueous Toluidin blue. The cytological observation showed small, round, oligodendroglial-like cells arranged in lobules. Microcystic spaces containing large ganglion cells floating in a fine fibrillary background were present. Foci of perivascular distribution of tumor cells were also noted. Blood vessels were thin-walled, with no endothelial proliferation (Figs. 1-3). Occasional areas with astrocytes were also detected. No necrosis and no mitosis were noted. Following the smear cytology diagnosis of DNT, a complete surgical resection was performed. The histological sections confirmed the diagnosis (Figs. 4, 5). To date, the patient remains well and free of seizures.

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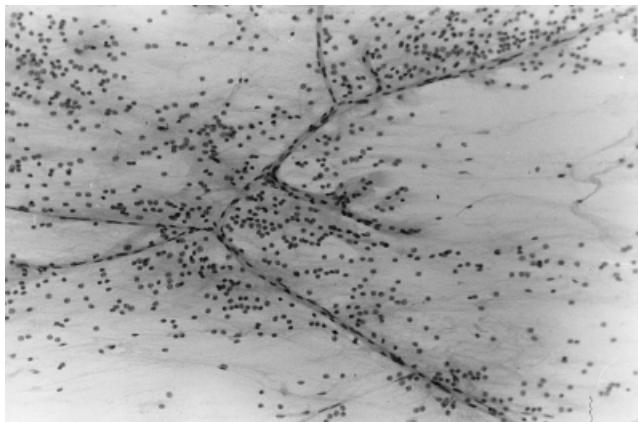
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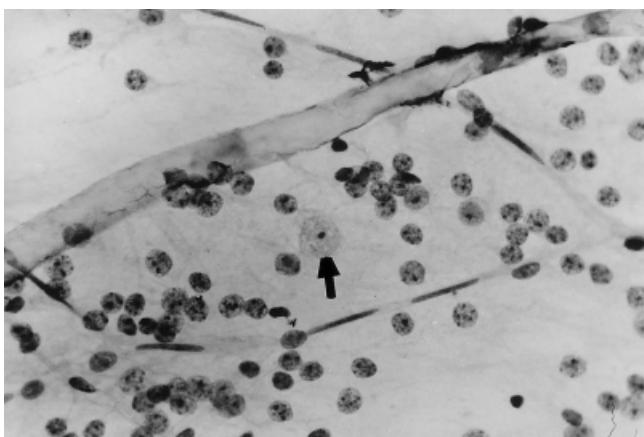
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**Fig. 1.** Smear preparation of DNT, showing small round oligodendroglial-like cells in a fibrillary background with thin-walled blood vessels (Toluidine blue,  $\times 100$ ).

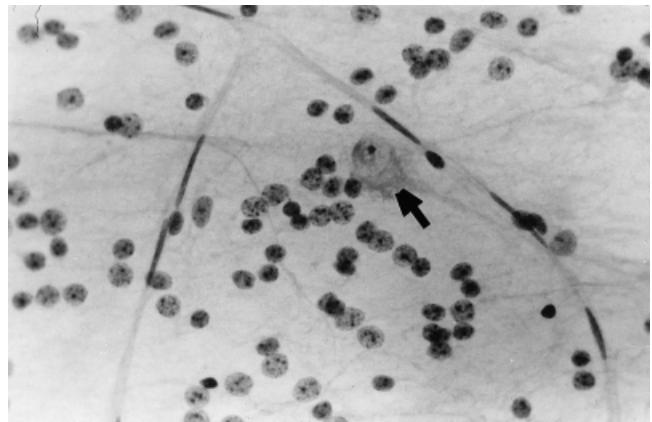


**Fig. 2.** High-power view of smear preparation of DNT, showing round oligodendroglial-like cells arranged in clusters with occasional large floating nerve cells (arrow) immersed in a fibrillary background (Toluidine blue,  $\times 400$ ).

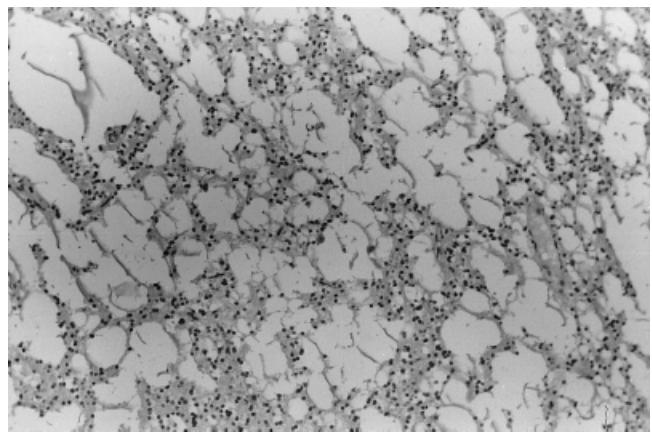
## Discussion

DNT is a superficial and intracortical lesion with multinodular architecture and a heterogeneous cell composition. The term “dysembrioplastic neuroepithelial tumor” is proposed for conditions that: 1) present with foci of cortical dysplasia that imply its appearance during cortical development; 2) are composed of different types of cells; 3) occur at a young age; and 4) accompany bone abnormalities near the tumor.<sup>1,3,4</sup> This is a benign, slow-growing lesion, surgically curable with no need of further radio- or chemotherapy. For this reason, early surgery can prevent other neurological damage caused by persistent and chronic epilepsy, improving the quality of life and prognosis of those patients.<sup>1,3,5</sup>

The smear technique has been widely used in the diagnosis of brain tumors.<sup>6,7</sup> The cytological presentation of DNT in a smear preparation is pleomorphic, depending on which component of the lesion is sampled. A mixture of



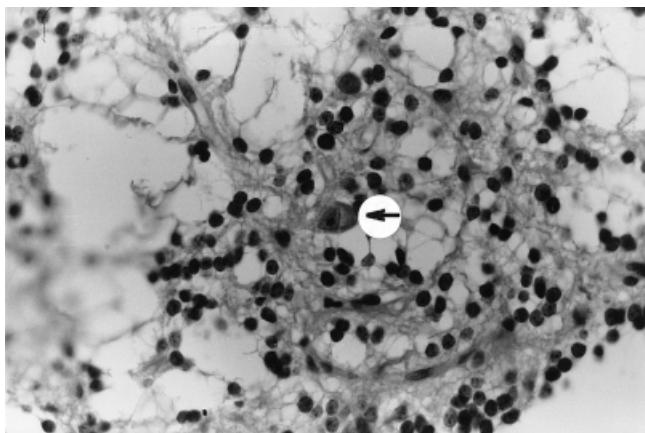
**Fig. 3.** High-power view of smear preparation of DNT. Arrow indicates large floating nerve cell (Toluidine blue,  $\times 400$ ).



**Fig. 4.** Histological section of DNT, showing a microcystic tumor composed of cords of oligodendroglial-like cells delimiting small spaces that contain floating nerve cells (hematoxylin-eosin,  $\times 100$ ).

ganglion cells, astrocytes, and cells resembling oligodendrocytes arranged in strains have been identified. DNT may show areas of ganglion cells set in a fine fibrillary background, resembling a neuronal tumor or even relatively unremarkable central nervous tissue. In other areas, oligodendroglial-like cells might predominate. These are frequently arranged in lobules closely related to thin-walled blood vessels. Other areas might resemble a central neurocytoma with scattered ganglion cells. Mitotic figures are not present. A characteristic feature of DNT is the cytological variability of appearances in different areas of the smear preparation, which confirms its variegated cellular composition.

The differential diagnosis of DNT on cytological grounds should be made with oligodendrogloma, low-grade astrocytoma, and ganglion-cell tumors. The smear preparation of oligodendrogloma is composed of similar cells and might form lobular structures. However, oligodendroglomas are uncommon in the temporal lobe of young patients and affect



**Fig. 5.** Histological section of DNT, showing nerve cell (arrow) immersed in a population of oligodendroglial-like cells (hematoxylin-eosin,  $\times 400$ ).

mainly the deep frontal lobes, with frequent foci of calcification detected by CT scan. Nevertheless, the final distinction between these two entities may require histology.

Low-grade astrocytomas might present with an abundant fibrillary matrix mimicking DNT. It is also important to consider this differential diagnosis, since most of the temporal lobe gliomas, particularly low-grade astrocytomas, might have unusual histological presentations with frequent dysplastic features. An important cytological point that might help this differential diagnosis is the presence of neurons floating among the more primitive-looking oligodendroglial cells, which tend to be arranged in lobules or clusters. Furthermore, surgical resection in astrocytic tumors, which are more infiltrative, tends to be more aggressive than in DNT, which is characteristically multinodular and well-delineated from the surrounding brain tissue.

Ganglion-cell tumors can be confused with DNT by showing a mixture of ganglionic and glial elements.<sup>3,4,8,9</sup> However, these lesions are also slow-growing and normally receive a neurosurgical treatment similar to that of DNT.

Knowledge of the entity, its clinical presentation, and its peculiar CT scan and magnetic resonance-imaging appearance help the intraoperative diagnosis. Accurate diagnosis of DNT and its distinction from ordinary gliomas have important therapeutic implications such as establishing the right margin at the surgery, and avoiding more aggressive surgical decisions.<sup>1,3,5,8</sup>

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