# Intraventricular dysembryoplastic neuroepithelial tumour: case report

## *Wewnątrzkomorowy dysembrioplastyczny guz neuroepitelialny – opis przypadku*

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## Abstract

Dysembryoplastic neuroepithelial tumour (DNT) is located in the cerebral cortex with very few exceptions. In this article, an extremely rare case of intraventricular DNT originating from the septum pellucidum is reported.
A 25-year-old woman presented with 5-month history of headache. Cranial magnetic resonance imaging (MRI) scans revealed a mass in the right lateral and third ventricle which was hypointense on T1-weighted image, and hyperintense on T2-weighted images. No contrast enhancement was detected. The lesion was excised totally using a transcallosal-transventricular approach. Immunohistochemical examination revealed DNT. The patient was discharged without any neurological deficits.
Intraventricular DNT presents with symptoms of increased intracranial pressure rather than seizures. Distinguishing DNT from other intraventricular tumours is essential as DNT is characterized by benign clinical course and does not require adjuvant therapy.

**Key words:** dysembryoplastic neuroepithelial tumour, cerebral ventricles, headache, septum pellucidum.

## Streszczenie

Dysembrioplastyczne guzy neuroepitelialne (DNT) są umiejscowione, z nielicznymi wyjątkami, w korze mózgowej. W bieżącej pracy autorzy przedstawiają wyjątkowo rzadki przypadek chorej z DNT położonym wewnątrzkomorowo, wychodzącym z przegrody przezroczystej.
Chora, 25 lat, zgłosiła się z powodu utrzymującego się od 5 miesięcy bólu głowy. W badaniu za pomocą rezonansu magnetycznego uwidoczniono guz położony w komorze bocznej prawej i w komorze trzeciej, hipointensywny w obrazach T1-zależnych, hiperintensywny w obrazach T2-zależnych i niewzmacniający się po podaniu środka kontrastowego. Guz wycięto w całości z dojścia przez ciało modzelowate i przez komorę. W badaniu immunohistochemicznym stwierdzono DNT. Chora została wypisana do domu bez ubytkowych objawów neurologicznych.
Wewnątrzkomorowe DNT przejawiają się raczej wystąpieniem objawów wzmożonego ciśnienia śródczaszkowego niż napadów padaczkowych. Odróżnianie DNT od innych guzów wewnątrzkomorowych jest nieodzowne ze względu na fakt, że guz przebiega łagodnie i nie wymaga leczenia wspomagającego.

**Słowa kluczowe:** dysembrioplastyczny guz neuroepitelialny, komory mózgu, ból głowy, przegroda przezroczysta.

## Introduction

Dysembryoplastic neuroepithelial tumour (DNT) was first defined as an oligodendroglioma-like tumour in temporal lobe specimens of epileptic patients by Cavanagh *et al.* in 1958 [1,2]. Daumas-Duport *et al.* in 1988 [1] improved our understanding of DNT by revealing that the tumour of that type is mostly seen in children and young adults and typically occurs in the supratentorial cortex. The tumour usually manifests with heada-

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che and intractable seizures. Although DNT behaves in a benign fashion, radiological features are similar to low-grade gliomas. To the best of our knowledge, previous studies have reported 22 intraventricular DNT cases including intraventricular, nucleus caudatus and septum pellucidum [2-13]. This report presents clinical, radiological and histological features of an intraventricular DNT in a young adult.

## Case report

A 25-year-old woman presented with progressive headache, nausea and vomiting for 5 months. There was no seizure in her medical history. No abnormal findings were present in general or in neurological examination. Cranial magnetic resonance imaging (MRI) scans revealed a non-enhanced mass lesion (19 × 15 × 12 mm) originating from the inferior septum pellucidum that extended to the third ventricle through the right foramen of Monro (Fig. 1). The tumour was hypointense on T1-weighted image and hyperintense on T2-weighted images. No contrast enhancement was observed. The tumour was removed totally using the transcallosal and transventricular approach (Fig. 2). The greyish and soft tumour originated from the septum pellucidum. The tumour extended to the third ventricle through the foramen of Monro and had very low vascularization. Immunohistochemical studies showed columnar nature, floating neuronal cells and synaptophysin reactivity that established the diagnosis of DNT (Fig. 3). The patient was discharged three days after the operation without any neurological deficit.

## Discussion

Dysembryoplastic neuroepithelial tumour is a benign, focal, intracortical tumour including a cystic component inside [10,11]. Dysembryoplastic neuroepithelial tumours present mostly with headache and intractable seizures. Nolan *et al.* [11] reported that DNT is involved in 14-18% of surgically treated epilepsy cases. Headache, which is the primary symptom

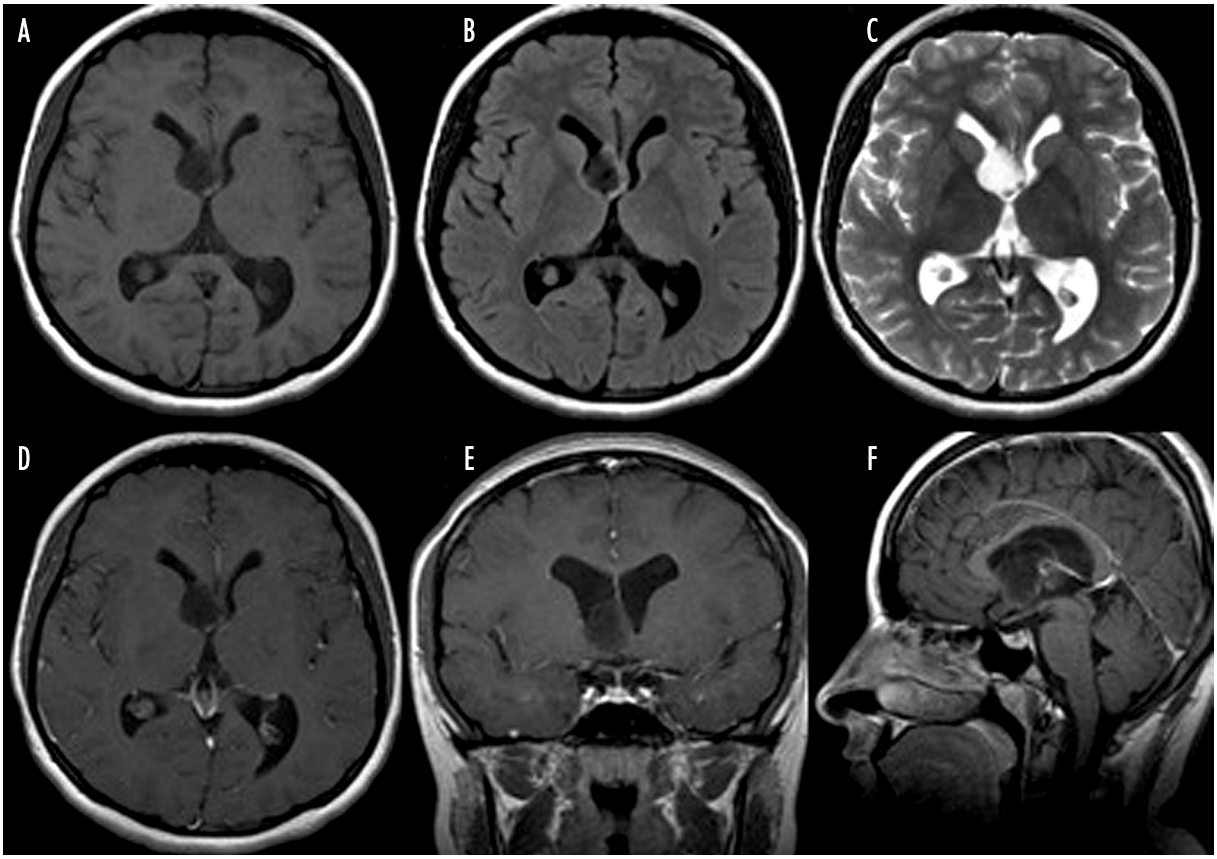


**Fig. 1.** A-C) The tumour adjacent to the inferior part of the septum pellucidum is hypointense on T1-weighted image and hyperintense on T2-weighted MR image. D-F) Non-enhancing tumour extending to third ventricle through foramen of Monro

in intraventricular DNTs, was also the dominant symptom in the presented case.

Although the most common location of DNT is the supratentorial cortex, often the temporal lobe, it is also possible to encounter DNT in various extracortical locations corresponding to the secondary germinal layers, including the dentate fascia, subpial germinal layer, subependymal germinal layer, and external granular layer in the cerebellum. So far, 22 DNT cases with extracortical locations have been reported. The most common extracortical locations are the septum pellucidum and nucleus caudatus. As in the present case, it is also possible to encounter cases with extension of the tumour from the foramen of Monro to the third ventricle [3,5,6].

The relation to the septum pellucidum, multilobulated appearance, T2-bright signal reflecting high mucin content, and a lack of contrast enhancement are the imaging features of intraventricular DNTs [5].

Although DNT shares some features with oligodendroglioma, it generally occurs intracortically and has distinctive multinodular form. Oligodendrocyte-like cells and floating neurons in a mucinous matrix which are the typical histological features of DNT were observed in the present case.

The pathophysiology is still controversial. Although malignant transformation of DNT has been reported, it is still unclear whether DNT is a neoplasm or a hamartoma [14]. Some factors, such as accompanying cortical dysplasia, young age and deformity of the overlying skull, connote dysembryogenetic origin [13]. The familial occurrence of intraventricular DNT was reported by Saito *et al*. [15], which suggests that these tumours may arise from germline mutation [15].

The largest series was reported by Baisden *et al.* in 2001 and included 3 children and 7 young adults [3]. Cervera-Pierot *et al.* [6] reported four patients with

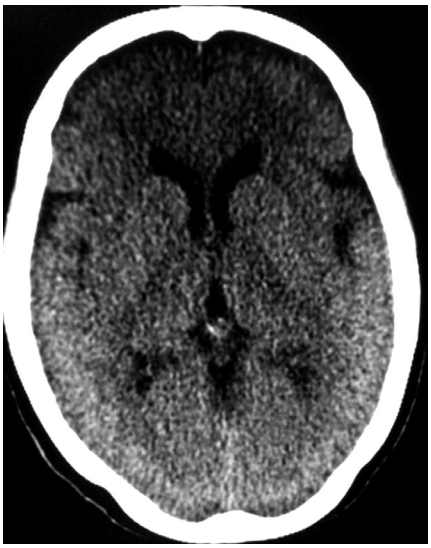

**Fig. 2.** Computed tomography performed 5 days after surgery shows the gross-total excision of the tumour

tumours that occurred in the nucleus caudatus and extended to the intraventricular area. Cataltepe *et al.* [5] recently reported a similar case, a 15-year-old boy presenting with epilepsy. Unlike the previous cases, the lesion extended from the corpus callosum to the septum pellucidum [3]. Bilginer *et al.* reported a very interesting intraventricular DNT in a 9-year-old boy with disseminated spinal tumour [4].

In the presented case and in the previous cases, the tumour was soft and sparsely vascular. Concerning these features, endoscopic removal of intraventricular DNT seems to be a safe and easy procedure. Harter *et al.* [8] reported a similar case which was successfully removed by endoscopy.

Intraventricular DNT is extremely rare but distinguishing this entity from more common intraventricu-

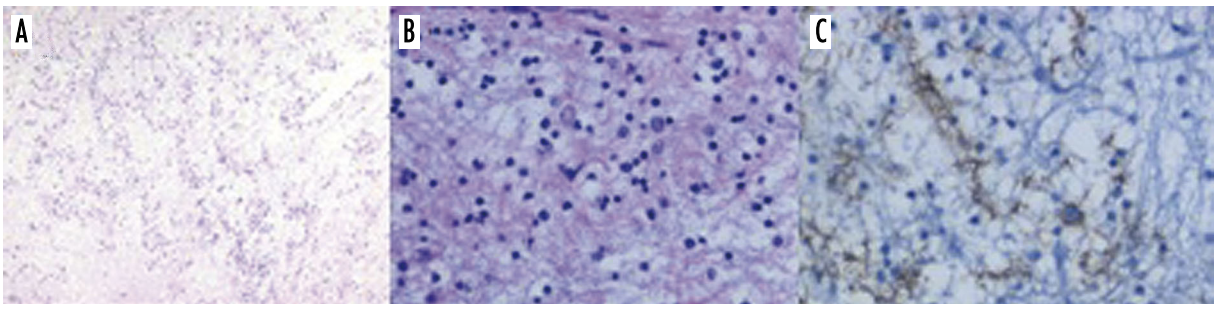


**Fig. 3.** A) Columns formed by bundles of axons oriented perpendicularly to the cortical surface. Neurons with normal cytology float between those columns on the pale eosinophilic background (H&E, ×40). B) Cytologically normal floating neurons with ganglion cells (H&E, ×200). C) Synaptophysin immunoreactivity of neuronal cell processes and ganglion cells (synaptophysin, ×200)

lar tumours, such as low-grade gliomas, ependymomas, or neurocytomas, is vital, since total removal provides a favourable prognosis and adjuvant therapy is not required. Rushing *et al.* [16] reported a patient with DNT that showed malignant transformation after radiation and chemotherapy [14].

## Conclusions

Intraventricular DNTs usually originate from the septum pellucidum and present with headache. They should be considered in the differential diagnosis of intraventricular tumours.

## Disclosure

Authors report no conflict of interest.

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