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Intraventricular Dysembryoplastic Neuroepithelial Tumor-Like Neoplasm with Disseminated Spinal Tumor

Yaygın Spinal Tümör ile Birlikte Ventrikül İçi Disembriyoplastik Nöroepitelyal Tümör-Benzeri Lezyon

Burçak BİLGİNER[1]
Figen SÖYLEMEZOĞLU[2]
Ayşenur CİLA[3]
Nejat AKALAN[4]

[1,4] Hacettepe University School of Medicine, Department of Neurosurgery, Ankara, Turkey
[2] Hacettepe University School of Medicine, Department of Pathology, Ankara, Turkey
[3] Hacettepe University School of Medicine, Department of Radiology, Ankara, Turkey

ABSTRACT

Dysembryoplastic neuroepithelial tumor (DNT)- like lesions arise in extracortical locations and behave in a benign fashion similar to that of cortical DNTs. They usually come to attention by symptoms and signs of increased intracranial pressure rather than focal neurological impairment. Here we report a case of 9-year-old boy with a complaint of headache and back pain. A third ventricular mass lesion with disseminated spinal tumor was detected on his magnetic resonance imaging. The presence of floating neurons in a mucinous matrix, oligodendrocyte-like cells (OLCs) aligning axonal columns and vessels, immunohistochemical profile of the neoplasm in addition to the clinical and radiological manifestations of the patient led to the diagnosis of "DNT-like neoplasm of the third ventricle".

KEYWORDS: Dysembryoplastic neuroepithelial tumor, Spinal seeding, Brain tumor

ÖZ

Disembriyoplastik nöroepitelyal tümör (DNT)-benzeri lezyonlar ekstrakortikal olarak yerleşirler ve kortikal yerleşimli disembriyoplastik nöroepitelyal tümör'lere benzer şekilde iyi huylu olarak davranırlar. Fokal nörolojik bozukluktan ziyade daha çok artmış kafa içi basıncı belirti ve bulguları ile dikkat çekerler. Biz burada baş ağrısı ve sırt ağrısı şikayeti olan 9 yaşında erkek hastayı sunduk. Hastanın manyetik rezonans görüntülemesinde (MRG) üçüncü ventrikül içi kitle ile birlikte yaygın spinal tümör bulundu. Müsinöz matriks içinde yüzen nöronların bulunması, aksonal sütun ve damarlarda dizili oligodendrosit benzeri hücreler (OLCs), hastanın klinik ve radyolojik bulgularının yanısıra tümörün immüno histokimyasal özellikleri hastada üçüncü ventrikülün disembriyoplastik nöroepitelyal tümör - benzeri lezyonu tanısını koydurmuştur.

ANAHTAR SÖZCÜKLER: Disembriyoplastik nöroepiteltal tümör, Spinal ekilim, Beyin tümörü

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Correspondence address:
Burçak BİLGİNER
E-mail: burcak@tr.net

INTRODUCTION

DNTs are seizure-producing and slow-growing neoplasms of children and young adults. They are generally cortical tumors and frequently associated with cortical dysplasia. DNTs can be located in any part of the supratentorial cortex with a special predilection for the temporal lobe. However there is an increasing number of publications and observations of extra-cortical DNTs. In 2001, Baisden et al reported 10 cases of DNT in the septum pellucidum and named this neoplasm as DNT-like neoplasm of the septum pellucidum due to the lack of classical clinical and morphological features of DNT (1).

Here we report a case of DNT-like neoplasm with disseminated spinal tumor.

CASE REPORT

A 9-year-old boy was admitted to our hospital with complaints of headache and back pain. He had headaches and nausea attacks for 15 days with progressive worsening. His neurological examination was normal except for bilateral papilledema.

Magnetic resonance (MR) images revealed a third ventricular mass lesion with obstructive hydrocephalus. The lesion was mildly hyperintense relative to CSF on T1-weighted images and hyperintense on FLAIR images relative to brain tissue. No contrast enhancement was observed (Figure 1A,B,C). The tumor was best delineated in TRACE images of DWI. Spinal MR images showed diffuse leptomeningeal contrast enhancement, regarded as seeding, at the cervicothoracal and lumbal regions (Figure 2 A,B,C,D). The spinal lesion showed moderate enhancement, contrary to the primary tumor.

An anterior interhemisferic transcallosal approach was performed and the surgery was subtotal due to the lack of demarcation between the tumor and hypothalamus. The histopathological examination of the tumor revealed a neoplasm with microcysts and mucinous matrix. The neoplasm was composed of cells with uniformly round and hyperchromatic nuclei and prominent perinuclear clearing, resembling oligodendrocytes. These oligodendrocyte-like cells were clustered tightly around perivascular spaces and were arranged in parallel to form a well-defined linear array, an architectural pattern described in cortical DNT as a

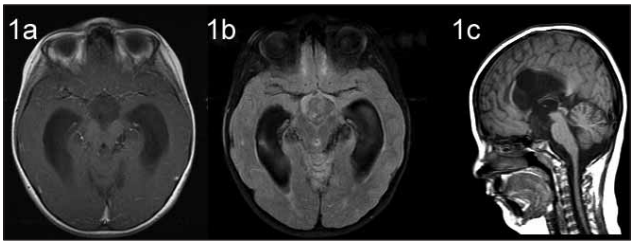

Figure 1. A*) Preoperative T1-weighted axial postcontrast MRI, a mild hyperintense mass is seen at the third ventricle with no contrast enhancement.*
B*) Preoperative axial FLAIR MRI, heterogenous hyperintense lesion is seen in the anterior recesses of the third ventricle.*
C*) Postoperative T1-weighted sagittal postcontrast MRI, the mass has been resected partially. The tumor at anterior recesses of the third ventricle is clearly seen.*

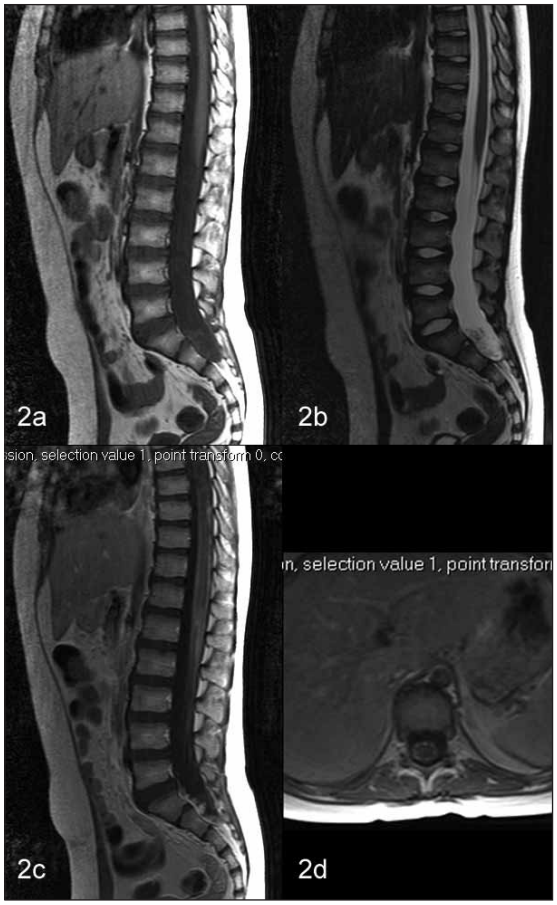

Figure 2. A*) Preoperative sagittal T1-weighted MRI of the thoracolumbal region, only minimal hyperintence mass is seen in the cul-de-sac.*
B*) Preoperative sagittal T2-weighted MRI of the thoracolumbal region, only hypointence mass is seen in the cul-de-sac.*
C*) Preoperative sagittal T1-weighted postcontrast MRI, diffuse leptomeningeal contrast enhancement around the spinal cord and nodular enhancement in the cul-de-sac drop metastasis is seen.*
D*) Preoperative axial T1-weighted postcontrast MRI, shows pial nodular enhancement of the metastasis.*

specific glioneuronal element. Easily identifiable neurons seemed to float within a transudate between these columns (Figure 3 A,B,C).

Cranial and spinal MR images were obtained at follow-up one year after the surgery. Cranial MRI showed diffuse leptomeningeal contrast enhancement compatible with leptomeningeal dissemination (Figure 4A). His one-year follow-up spinal MR images were stable when compared with the initial images (Figure 4B). Regardless of the subtotal resection and the spinal lesions, the patient is still doing well three years after the operation.

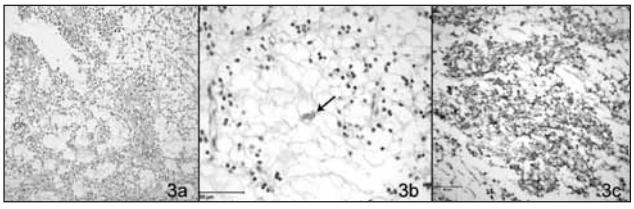

Figure 3. A) *A neoplasm characterized by microcysts lined by oligodendrocyte-like cells (HE).*
B) *A floating neuron in mucinous matrix (HE) (Arrow).*
C) *Oligodendrocyte-like cells are aligning NFP expressing axons (NFP).*

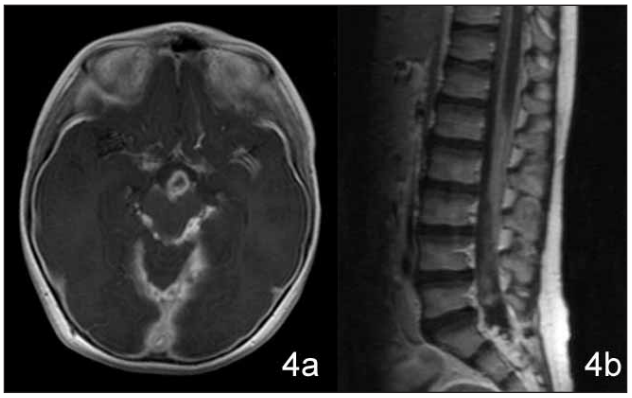

Figure 4. A) *Cranial,* ***B)*** *Spinal, one year control MRI showing massive enhancement in the subarachnoidal space in the brain and spine compatable with leptomeningeal dissemination.*

DISCUSSION

Many authors report extracortical localization of DNTs such as the basal ganglia, pons, thalamus, cerebellum and the third ventricle, but caudate nucleus and septum pellucidum have been the most common sites (1,3,4). The presenting symptoms of those lesions were due to increased intracranial pressure and they were radiologically hypointense on T1-weighted MR images, hyperintense on T2 images and typically nonenhancing. As Baisden et al. reported previously, DNT-like lesions must be considered in the differential diagnosis of intraventricular tumors such as, oligodendroglioma, central neurocytoma and subependymal giant cell astrocytoma (7).

In this report we describe a case of DNT-like tumor which was located primarily in the third ventricle with disseminated spinal tumor.

Histologically the current case is characterized by oligodendrocyte-like cells (OLCs) and floating neurons in a mucinous matrix, the so called "special glioneuronal element", which is accepted as typical for DNT. However, the diagnosis of a third ventricular DNT can be challenging. In general, the principal differential diagnosis of DNT is with oligodendroglioma, while this possibility is low at a ventricular location. Ventricular DNT has to be distinguished from a group of low-grade neoplasms such as central neurocytoma, ependymoma, pilomyxoid astrocytoma, and pilocytic astrocytoma. The absence of neuronal marker expression by OLCs and lack of neuropil-like islands discourage the diagnosis of central neurocytoma. Though the perivascular collection of tumor cells superficially resembles ependymoma, the absence of GFAP and EMA immunoreactivity speaks against ependymoma and subependymoma. The presence of neurons, the absence of intratumoral Rosenthal fibers and eosinophilic granular bodies, and the lack of GFAP immunoreactivity differentiates this lesion from pilocytic astrocytoma. Again, the lack of GFAP immunoreactivity of OLCs along with the presence of neurons, despite perivascular orientation of tumor cells in a myxoid background, are against pilomyxoid astrocytoma.

Regarding the spinal lesion there are several possibilities such as leptomeningeal dissemination, multifocality or another primary spinal tumor. The spinal lesion observed during the presentation of the patient was not confirmed by biopsy, so it is difficult to rule out multifocality or probability of another primary neoplasm. Though controversial, two cases of malignant transformation of DNT has been described (6,10), however there was no sign of anaplasia in the ventricular neoplasm. Given the reported leptomeningeal dissemination in other children with other low-grade neuroepithelial tumors, the leptomeningeal dissemination scenario is the most probable. However spinal leptomeningeal seeding has never been reported in DNTs.

Leptomeningeal dissemination (LMD) of low grade gliomas (LGG) is reported in clinical neurooncology practice with a frequency of 5-10% (2,5,12). This is a relatively low estimate since patients with LGG were not routinely staged. LMD has been reported in association with almost all known subtypes of low grade neuroepithelal neoplasms. The evidence of LMD in children with LGGs does not seem to have a negative impact on patients' long term outcome. Based on the data available, it seems reasonable to be conservative. Chemotherapy has been the most commonly recommended tool for treatment prior to craniospinal RT (8,9,11).

From the neurooncological point of view, the recently described pathological entities such as DNT-like tumors cause inconvenience. Since the pathological findings indicate a rather benign tumor, there is not enough follow-up data to determine the necessity of adjuvant therapy following surgery, especially in those cases with subtotal resection. Our case is an example of such a challenge, where we decided to follow-up with close radiological surveillance.

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