Case Report

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Diffuse Ependymal Dysembryoplastic Neuroepithelial Tumor Causing Spinal Drop Metastases: A Case Report

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Dysembryoplastic neuroepithelial tumors (DNETs) arise mostly in the supratentorial cerebral cortex. A very rare case of intraventricular DNET with diffuse ependymal involvement, which causes spinal drop metastasis, is presented.
Index terms: *Brain tumor; Dysembryoplastic neuroepithelial tumor*

INTRODUCTION

A dysembryoplastic neuroepithelial tumor (DNET) is a benign, mixed neuronal-glial neoplasm associated with a medically intractable seizure in children and young adults (1). DNETs are typically located in the supratentorial cortex, mostly in the temporal lobe. There have been few reports regarding extracortical locations of DNET, such as the cerebellum, brainstem, pericallosal region, septum pellucidum, caudate nucleus, and intraventricular region (2-5). Intraventricular DNET is extremely rare and manifested as a well circumscribed focal mass with no contrast enhancement (2, 6-9). Herein, we present a case of DNET that had widespread, diffuse ventricular ependymal involvement, which has not previously been described in the literature.

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CASE REPORT

A 29-year-old female patient presented with a 2-month history of decreased visual acuity and hand tremors. In the 4 days prior to presentation, she had gait disturbance. Neurological examination at the presentation revealed a drowsy mental status, diplopia, and gait disturbance. MR imaging demonstrated diffuse, thick nodular ependymal masses throughout the entire ventricles. The lesion was hypointense on T1-weighted images and markedly hyperintense on T2-weighted images (Fig. 1A, B). Fluid attenuated inversion recovery (FLAIR) images showed a well defined hyperintense thin rim surrounding the tumor (Fig. 1C). In addition, peripheral rim like enhancing portions within the mass in the right lateral ventricle were noted on postcontrast T1-weighted imaging (Fig. 1D). Obstructive hydrocephalus was also noted. There was no diffusion restriction on the diffusion-weighted images (Fig. 1E). The preoperative diagnosis was intraventricular astrocytoma. Neuronavigation-assisted excisional biopsy was performed on the mass in the temporal horn of right lateral ventricle.

Korean Journal of Radiology
KJR

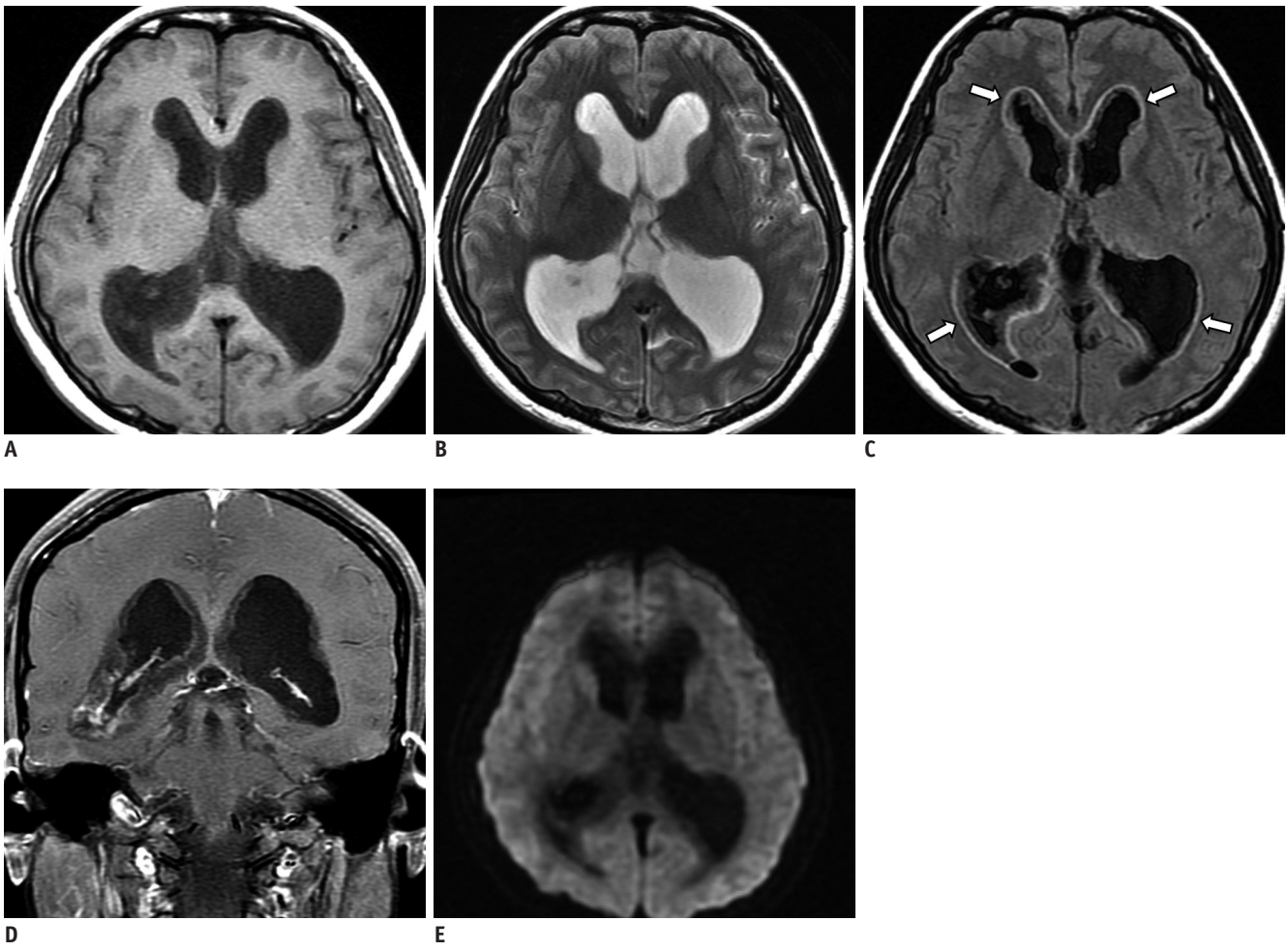

Fig. 1. Diffuse ependymal dysembryoplastic neuroepithelial tumor with spinal drop metastasis in 29-year-old female patient.
A. Axial T1-weighted image shows diffuse and nodular low signal intensity lesions along ependymal surface of lateral ventricles and third ventricle. **B.** Lesions show bright signal intensity on axial T2-weighted image. **C.** Axial fluid attenuated inversion recovery image shows hyperintense rim (arrows) along wall of ventricles between mass lesions and underlying periventricular white matter. **D.** Coronal, contrast-enhanced T1-weighted image shows peripheral rim like or nodular enhancement along surface of mass lesions. **E.** Axial diffusion-weighted image shows no diffusion restriction within tumor.

Pathological evaluation revealed oligodendrocyte-like tumor cells with uniformly round and hyperchromatic nuclei and a perinuclear halo in a myxoid background (Fig. 1F); immunohistochemical staining showed floating neurons with a positive synaptophysin reaction (Fig. 1G). The histopathologic diagnosis was determined to be a dysembryoplastic neuroepithelial tumor. The patient underwent a ventriculoperitoneal shunt for treatment of hydrocephalus and was managed conservatively. 29 months after her initial presentation, she developed lumbar pain and motor weakness in both legs. Lumbar spine MR imaging demonstrated multiple well defined enhancing lesions surrounding cauda equina and S1 nerve roots, which is suggestive of leptomeningeal drop metastases (Fig. 1H, I). The patient consequently underwent radiation therapy to the lumbosacral region.

DISCUSSION

DNET is a benign tumor of neuroepithelial origin that occurs in children and young adults and is usually located in the cerebral cortex. These tumors frequently cause medically intractable partial seizures. The histopathological hallmark of DNETs is the glioneuronal element, which contains oligodendrocyte-like cells attached to bundles of axons and neurons floating in a myxoid interstitial fluid (10).

DNETs are typically an intracortically located supratentorial tumor. The temporal lobe is the most common site, followed by the frontal lobe, parietal lobe, and occipital lobe (10). Only a few intraventricular DNET

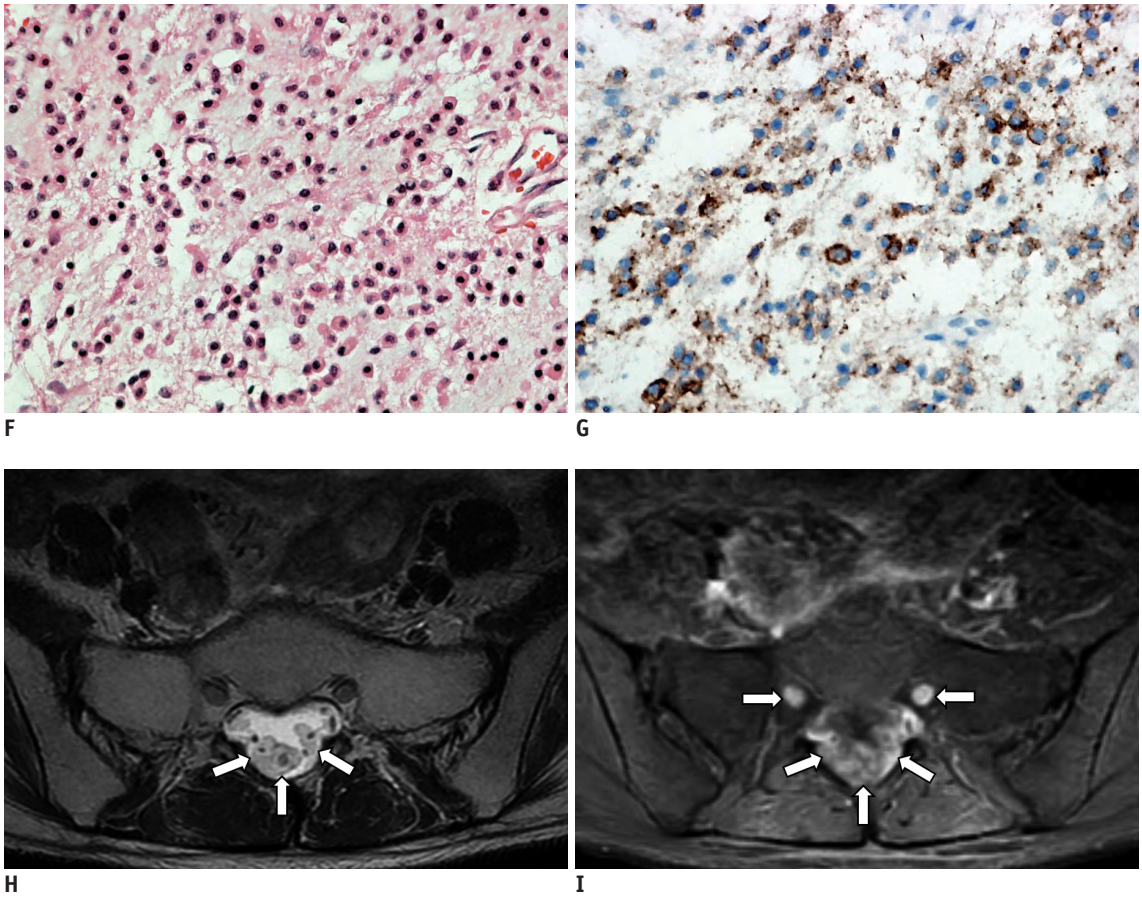

Fig. 1. Diffuse ependymal dysembryoplastic neuroepithelial tumor with spinal drop metastasis in 29-year-old female patient.
F. Photomicrograph shows oligodendrocyte-like cells in mucinous matrix (H&E, x 400). **G.** Immunohistochemical staining shows that synaptophysin is expressed in larger tumor cells, which are supposed to be neuronal component (IHC, x 400). **H.** Axial, T2-weighted lumbar spine magnetic resonance (MR) image obtained 29 months after initial presentation shows well defined, multiple intradural masses with high signal intensity surrounding cauda equina (arrows). **I.** Axial, contrast enhanced T1-weighted lumbar spine MR image shows extensive enhancement in intradural masses and bilateral S1 nerve roots (arrows).

cases have been reported in the literature to date (2, 6-9). It has been suggested that a intraventricular DNET derives from the subependymal secondary germinal layer (5). Intraventricular DNET has been described as a well circumscribed, localized mass located in the lateral ventricle or third ventricle with associated obstructive hydrocephalus in the literature. In contrast to previous reports, our case has a unique morphological feature; widespread, diffuse nodular masses along walls of entire ventricles. To the best of our knowledge, this "diffuse ependymal involvement type" of DNET has not been previously reported.

The MRI signal characteristics of intraventricular DNETs are similar to those of ordinary DNETs located in the cortical region. At MR imaging, DNETs are hypointense on T1-weighted images and hyperintense on T2-weighted images without mass effect or surrounding vasogenic edema, regardless of their location. Calcifications and hemorrhage are rare (10). The other signal characteristics included a relatively low incidence of contrast enhancement and pseudocystic appearance because of abundant myxoid interstitial matrix of the tumor. Around 20% of DNETs show a nodular, ring-like, or heterogeneous contrast enhancement (10, 11). Recently, Parmar et al. (12) reported that the finding of hyperintense rim surrounding the tumor on FLAIR images is sensitive and specific for DNETs. They speculated that the hyperintense rim on FLAIR images may represent loose glioneuronal elements at the periphery of the tumors. Our case also demonstrated uniform, thin hyperintense rim between nodular tumors and underlying normal-appearing white matter on FLAIR images, which has not been described in previously reported cases of intraventricular DNETs.

Our patient developed multiple intradural masses of the cauda equina and sacral nerve roots 2.4 years after initial presentation, which is suggestive of spinal drop metastasis due to leptomeningeal seeding of the tumor. We found

Korean Journal of Radiology
KJR

only one case report of intraventricular DNET with spinal leptomeningeal seeding in the literature. Bilginer et al. (9) reported a case of 9-year-old boy with a localized DNET in the third ventricle and leptomeningeal dissemination. Leptomeningeal dissemination of low grade glioma has been reported with a frequency of 5-10% (9). As shown in our case and previous case reports, there is a likelihood of spinal dissemination in cases of intraventricular DNETs. Thus, we recommend that the patients with intraventricular DNETs should be routinely screened and followed up with spine MR imaging.

In conclusion, we present the first case of intraventricular DNET with widespread, diffuse ependymal involvement that caused spinal drop metastases. Intraventricular DNETs may also show the hyperintense rim sign on FLAIR images similar to those with cortical location. In addition, routine spine MR imaging should be recommended for the early diagnosis of spinal drop metastasis in patients with intraventricular DNETs.

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