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Atypical central neurocytoma with metastatic craniospinal dissemination: a case report

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ABSTRACT

Central neurocytomas comprise nearly half of adult intraventricular neoplasms. The median age of onset is 34 years. It is typically a low-grade neoplasm (World Health Organization grade II), although some cases of malignant neurocytomas have been described. We present a rare case of an atypical central neurocytoma with craniospinal dissemination, including both imaging and pathologic findings.

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A 57-year-old Hispanic male presented with a two-month history of symptoms of progressive obstructive hydrocephalus. Prior to admission, he had experienced headache, ataxia, and disorientation. On admission to the hospital, he progressed to coma. Initial magnetic resonance imaging (MRI) of the brain showed a tumor with widespread cerebrospinal fluid dissemination of multiple intraventricular masses in both frontal horns of the lateral ventricles with involvement of the septum pellucidum. Tumor deposits were also present in the third ventricle and in the superior portion of the fourth ventricle. There was mild right-to-left midline shift, hydrocephalus, and transependymal edema. In addition, there was evidence of multiple lesions in the quadrigeminal plate cistern, cerebellar fissures, and along the surface of bilateral cerebellar flocculi. These masses demonstrated hyperintensity on T1-weighted images (Fig. 1) and heterogeneous hypointensity on T2-weighted images (Fig. 2). Only mild peripheral enhancement was noted with absence of diffusion restriction (Figs. 1D and 2C).

The patient underwent a frontal interhemispheric transcallosal approach surgical resection for debulking of the lateral and third ventricular masses. Intraoperatively, the tumor was gray, soft, and easily distinguished from the brain. The patient tolerated the procedure well and eventually improved to his premorbid baseline.

Pathologic analysis of the resected tumor showed high cellularity and a monomorphic population of cells with uniform, round nuclei evenly distributed in a relatively scant, granular eosinophilic stroma. No calcifications were found in the tissue submitted to pathology. Scattered rosette-like structures suggested neurocytic differentiation, and the tumor showed diffuse immunohistochemical positivity for the neuronal markers, synaptophysin, chromogranin, and neuronal nuclear antigen (NeuN). The tumor was also positive for S-100, and scattered cells were positive for the astrocytic marker, glial fibrillary acidic protein (GFAP). Stains for neurofilament, keratin, and common leukocyte antigen (CD45) were negative. Although these findings were most compatible with the diagnosis of central neurocytoma (Fig. 3), the tumor had several unusual features that are generally associated with aggressive behavior in brain tumors. There were apoptotic cells indicating high cellular turnover and up to 5 mitotic figures per 10 high-power fields. An immunohistochemical stain for the proliferation-associated antigen, Ki-67 (MIB-1), showed an elevated proliferation index of up to 15% in some areas. Necrosis and endothelial proliferation were suspected but not definitive.

One month after surgery, spinal dissemination of the tumor was identified on MRI. Lumbar spine MRI showed two small intradural enhancing nodules consistent with spinal leptomeningeal spread involving the cauda eqina at the L4–L5 level and involving the left L4 nerve root (Fig. 4). The patient received Gamma Knife sterotactic radiosurgical treatment for ablation of the intracranial masses. He was followed for tumor progression as an outpatient.

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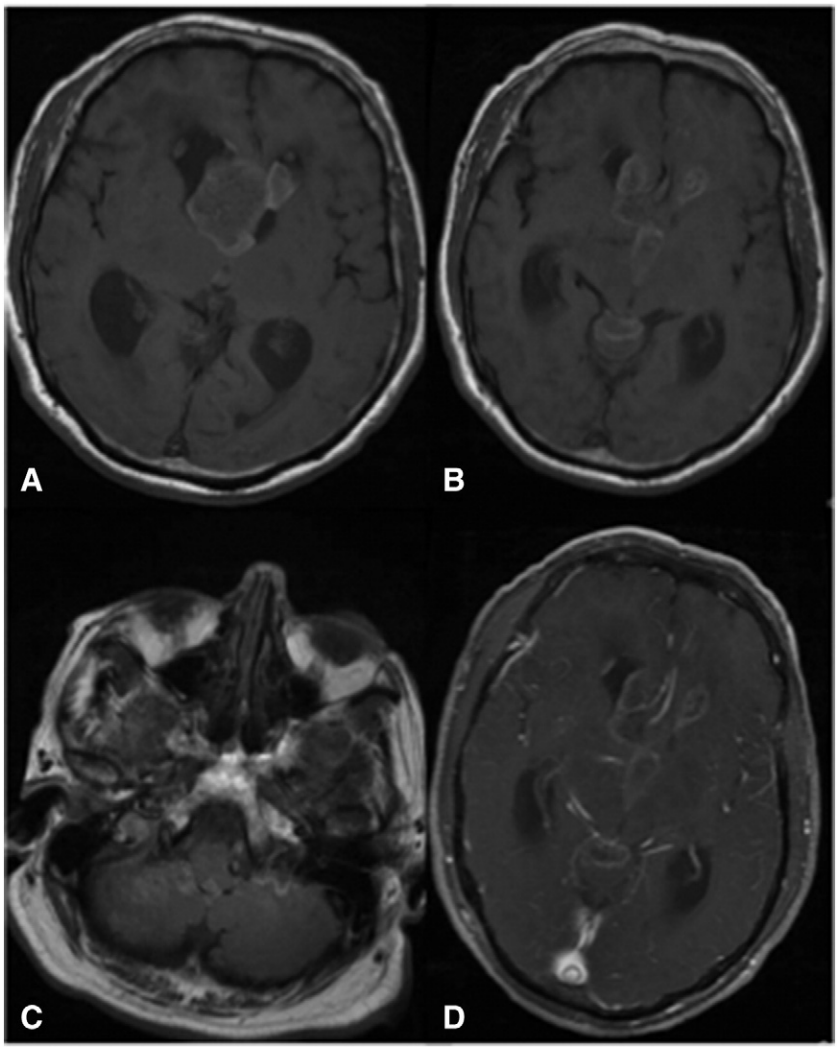

Fig. 1. MRI preoperative examination. (A–C) Axial T1-weighted images reveal multiple intraventricular masses involving the lateral, third, and fourth ventricles. Multiple leptomeningeal lesions are also noted in the fissures of the cerebellum and along the flocculi. All the masses appear hyperintense on T1-weighted images. (D) Axial postcontrast T1-weighted image demonstrates only mild peripheral enhancement.

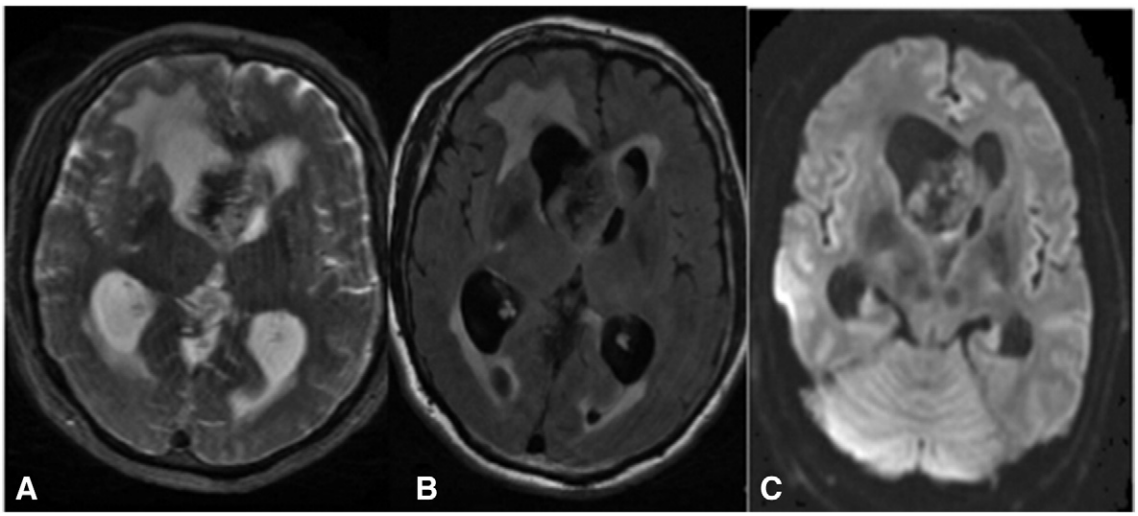

Fig. 2. MRI preoperative examination. (A) Axial T1-weighted image demonstrates heterogeneously hypointense signal within the intraventricular mass. (B) Axial T2 FLAIR image shows hyperintensity in the periventricular white matter, consistent with transependymal edema and hydrocephalus. (C) Axial diffusion-weighted image reveals no significant diffusion restriction within the mass.