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Lethal disseminated dysembryoplastic neuroepithelial tumor following West Nile virus: Report of a very unusual combination

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ABSTRACT

Dysembryoplastic neuroepithelial tumors (DNETs) are typically benign World Health Organization (WHO) grade I tumors of the cortical or deep gray matter with a favorable prognosis. We encountered a patient with DNET who has been evaluated and treated for West Nile encephalitis 7 months before presentation. Over the course of 2 years, the patient developed diffuse leptomeningeal carcinomatosis. As the disease burden increased, the patient eventually became quadriparetic. The patient elected for hospice care and expired shortly thereafter. Autopsy revealed DNET (WHO grade I) with extensive involvement of the cervical, thoracic, and lumbar spinal cord, bilateral cerebellar hemispheres, brainstem, the cortex of the right frontal and temporal lobes, and meningeal carcinomatosis of the brain and spinal cord. Mortality from DNET is rare, and as per our extensive literature search, there has been only 1 case reported of death attributed to seizures from this diagnosis. To the best of our knowledge, this is the only case of disseminated DNET with meningeal infiltration or carcinomatosis resulting in mortality.

Case report

A 36-year-old man was transferred to our care for hydrocephalus and a T2-T3 spinal cord lesion. He had a known history of West Nile encephalitis, diagnosed 7 months before presentation by viral polymerase chain reaction on a cerebrospinal fluid (CSF) specimen. On presentation, he endorsed a 2-week history of migraine and blurred vision. His additional symptoms included weakness, numbness, urinary incontinence, and vomiting. On physical examination, he was noted to have decreased strength in his left lower extremity, clonus in his right foot, and a positive left Babinski sign. Imaging demonstrated enhancement of the filum terminale and of nerve roots in the

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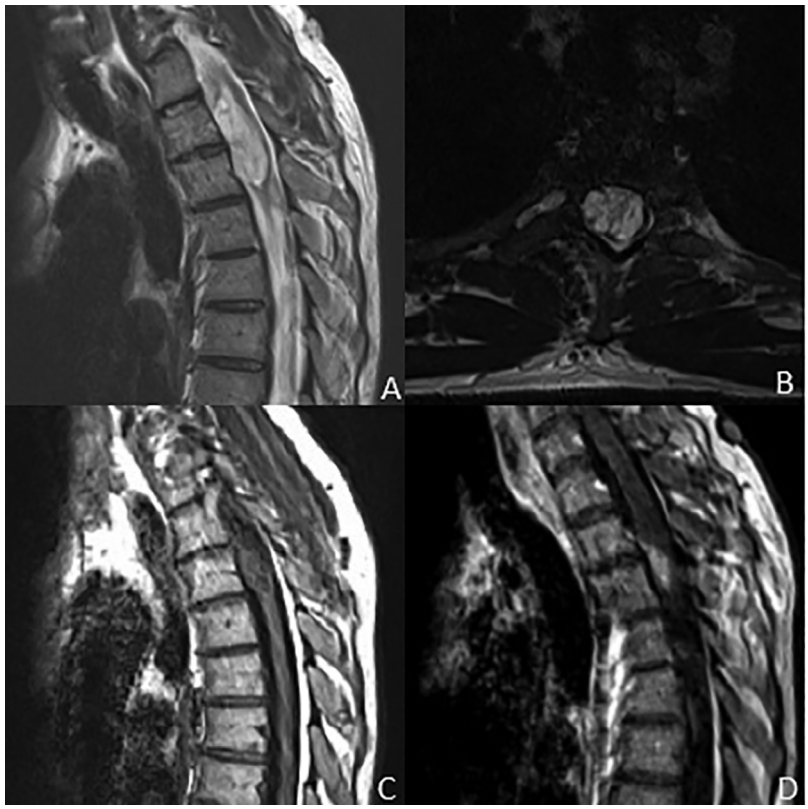

Fig. 1 – Sagittal (A) and axial (B) T2-weighted magnetic resonance imaging (MRI) of the thoracic spine on the patient's initial presentation demonstrates an expansile intramedullary mass from T1 to T3-4. Sagittal T1-weighted pre- and postcontrast MRI (C and D) demonstrate enhancement of the mass.

lumbar and sacral regions (Fig. 1). This is thought to represent leptomeningeal spread from the patient's known thoracic spinal cord mass.

An endoscopic third ventriculostomy was performed. CSF specimen was sent for cytology at the time of the procedure and was negative for malignancy. The patient's symptoms initially improved postoperatively; however, he then proceeded to present to the hospital multiple times in the following months with complaints of increasing headaches, enuresis, and decreased visual acuity.

Three months after the ventriculostomy, he was readmitted for intractable headache, pain in the neck, lower back, and left thigh, as well as worsening lower extremity weakness. Lumbar imaging showed nodular leptomeningeal disease with a prominent dural-based mass in the distal lumbar spinal canal. He underwent lumbar laminectomy and a biopsy of a lumbar subdural lesion. Pathology came back as dense fibrous connective tissue with chronic inflammation and myxoid-like foci, and was negative for malignancy. Imaging repeated 1 month after the procedure showed worsening of leptomeningeal enhancement.

The patient re-presented with continually worsening neurologic symptoms and underwent multiple shunt revisions. Throughout his clinical course, serial imaging demonstrated severe progression of a cervical-thoracic intramedullary spinal cord mass, a sacral tumor, tonsillar herniation, and diffuse carcinomatosis. He had repeat biopsy of an enhancing cerebellar mass, but again the sample showed no evidence of his malignancy. Additionally, he continued to show West Nile virus (WNV) activity, with persistent immunoglobulin M antibodies in the CSF.

Ultimately, the patient's symptoms worsened as he became quadriparetic. Two years after his initial presentation to our institution, magnetic resonance imaging (MRI) yet again showed worsening diffuse leptomeningeal carcinomatosis (Fig. 2). One month after the MRI confirming further advancement of his disease, the patient and his family elected for hospice care and he unfortunately expired shortly thereafter.

On autopsy, he was found to have dysembryoplastic neuroepithelial tumor (DNET) (World Health Organization grade I), with involvement of the cervical, thoracic, and lumbar spinal cord, bilateral cerebelli, brainstem, the cortex of the right frontal and temporal lobes, and meningeal carcinomatosis of the brain and spinal cord (Fig. 3).

Discussion

DNET is a typically benign superficial cortical neoplasm. For a long time, it was considered a hamartomatous lesion but molecular studies and clonal analysis confirmed its neoplastic nature. More than 60% of DNET are located in the temporal lobe. DNET is a slowly growing tumor, and the morbidity associated with this neoplasm consists virtually of only intractable, difficult to manage, seizures. Most patients