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Disseminated oligodendroglial-like leptomeningeal tumor with anaplastic progression and presumed extraneural disease: case report

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ABSTRACT

We report the neuroimaging and histopathologic findings of a 12-year-old female patient with a disseminated oligodendroglial-like leptomeningeal tumor with anaplastic progression and presumed extraneural metastatic disease. These tumors may represent distinct pathology primarily seen in pediatric patients. Neuroimaging demonstrates diffuse, progressive enhancement of the leptomeninges often with interval development of intraparenchymal lesions on follow-up. Disease is typically confined to the central nervous system, though diffuse peritoneal disease was seen in our case, possibly through metastatic seeding of the abdomen via ventriculoperitoneal shunt.

1. Introduction

Disseminated oligodendroglial-like leptomeningeal tumors are rarely encountered neoplasms predominately reported in pediatric patients. Recent case series suggest that the overlapping features among similar low-grade neoplasms with leptomeningeal dissemination represent a distinct clinicopathologic entity [1,2]. Disease course is typically indolent, but aggressive progression of disease is reported in a subset of patients [1]. Extraneural disease is previously unreported. We describe the case of a 12-year-old female with a disseminated oligodendroglial-like leptomeningeal tumor and anaplastic progression who developed extensive intraparenchymal disease as well as presumed extraneural disease throughout the peritoneum thought to be associated with placement of a ventriculoperitoneal shunt.

2. Case report

A previously healthy 12-year-old female presented with a 3-week history of headaches associated with transient slurred speech and right-sided weakness. Multiple lumbar punctures revealed xanthochromatic cerebrospinal fluid (CSF) with consistently elevated opening pressures and protein and no neoplastic cells. Extensive viral workup was negative, as was testing for Rickettsial and Lyme disease. Magnetic resonance imaging (MRI) revealed leptomeningeal enhancement in the left frontal and bilateral parietal regions (Fig. 1A). Multiple levels throughout the spine demonstrated mild leptomeningeal enhancement as well (Fig. 1B). No discrete mass lesion was noted. The patient was treated with intravenous immunoglobulin and high-dose steroids for a possible infectious etiology. Her mental status markedly improved and she was discharged with an uncertain diagnosis on a dexamethasone taper.

The patient was readmitted 1 week later with severe headache and episodes concerning for seizure. MRI revealed marked interval enlargement of the ventricles and persistent leptomeningeal enhancement. A ventriculoperitoneal shunt was placed and biopsy of the left frontal cortex and dura was performed. Biopsy revealed a small number of atypical cells in the leptomeninges of unclear significance. Repeat MRI taken 2 weeks after her admission showed progressing disease with more pronounced leptomeningeal thickening and enhancement with involvement of cranial nerves (Fig. 2). Another biopsy was performed, this time of the lumbar spinal dura.

Abbreviations: CNS, Central nervous system; CSF, Cerebrospinal fluid; MRI, Magnetic resonance imaging.

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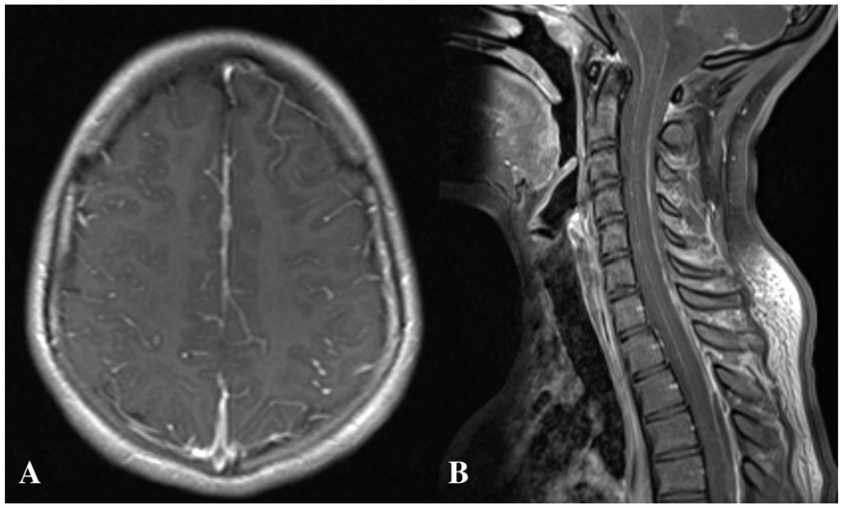

Fig. 1. MRI of brain and spine. (A) Axial postcontrast T1 imaging showing leptomeningeal enhancement of the bilateral parietal regions. (B) Sagittal postcontrast imaging showing mild enhancement at multiple levels throughout the cervical spine.

The second biopsy revealed small aggregates of monomorphic cells with round nuclei and clear cytoplasm involving the leptomeninges. The cells were positive for OLIG-2 and synaptophysin and were negative for NeuN and chromogranin. The histological features and immunohistochemical staining pattern were most consistent with a diagnosis of a low-grade oligodendroglial-like tumor. FISH analysis revealed neither 1p nor 19q deletion and immunohistochemistry for IDH1(R132H) was negative.

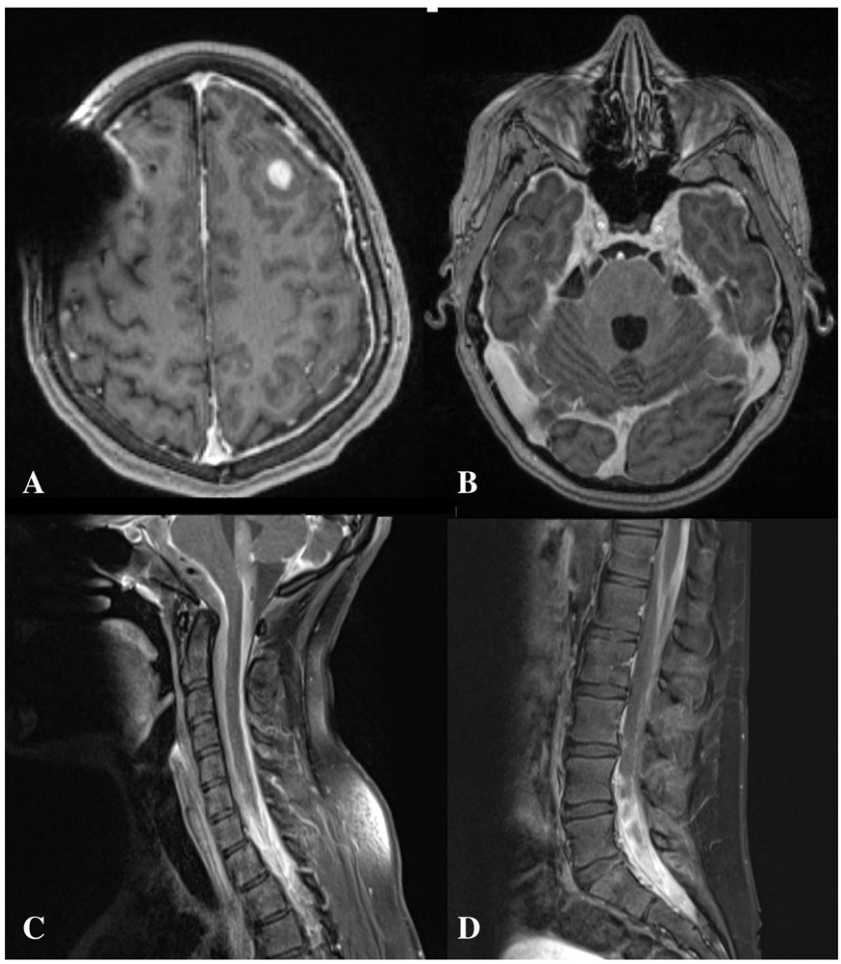

Fig. 2. MRI of brain and spine. (A) Axial postcontrast T1 imaging of the brain showing postoperative sequelae of right frontal ventriculoperitoneal shunt placement and left frontal brain biopsy with increased leptomeningeal enhancement along the left parietal and frontal region. (B) Axial postcontrast T1 imaging showing prominent leptomeningeal enhancement in the subarachnoid spaces of the middle cranial fossa medial to the temporal lobes extending along cranial nerves V and VI. (C and D) Postcontrast T1 imaging of the cervical and lumbar spine showing interval increase in leptomeningeal enhancement along the spine on multiple levels.