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Case Reports

Primary leptomeningeal glioblastoma with systemic metastases—case report and review of literature ☆,☆☆,★

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ABSTRACT

We present a case of a rare meningeal glioblastoma with extensive systemic metastases, along with a review of the existing literature. The patient presented with headache and visual and sensory symptoms and had a rapidly progressive clinical course despite therapy. The patient went on to develop spinal, orbital, muscular, and cutaneous metastases over a short period. Review of the literature reveals 13 similar previously reported cases, none with a similar presentation.

1. Introduction

Primary leptomeningeal astrocytomas (PLAs) are rare glial tumors of meningeal origin that may occur anywhere along the craniospinal axis. More commonly, these present as diffuse meningeal enhancement and mimic metastases or infectious or inflammatory meningitis. The focal form, on the other hand, can mimic a meningioma or a dural metastatic lesion [1]. Systemic metastases are rare, both with intraaxial glial tumors or PLAs [2–4]. We report a case of a primary meningeal glioblastoma (GBM) with extensive systemic metastases, along with a review of imaging findings.

2. Case report

A 51-year-old Hispanic male presented to our hospital emergency room with complaints of headache and blurred vision for 3 weeks associated with right-sided parasthesias. There were no associated motor, cranial nerve, or cognitive deficits. There was no history of any fever, trauma, or drug abuse leading up to the symptoms. Past medical history and family history were noncontributory.

Noncontrast computed tomographic (CT) study revealed presence of a mixed-density extraaxial lesion in the left parafalcine region, minimally extending across the midline. There was no apparent overlying bony destruction. Subsequent magnetic resonance imaging (MRI) redemonstrated a mixed solid–cystic extraaxial left parafalcine lesion with extension across the midline and through the overlying bone into the subcutaneous tissue. Short segment superior sagittal sinus involvement was also seen (Fig. 1a–e). The lesion was felt to represent a metastatic lesion versus an aggressive meningioma. CT of the chest, abdomen, and pelvis was however negative for any primary tumor.

The lesion was noted to be extraaxial at surgery, and a gross total resection was performed. Histopathology revealed a moderately pleomorphic meningeal neoplasm with superficial cortical invasion (Fig. 2). Perivascular rosetting, primitive gland formation, and vascular invasion were also noted (Figs. 3 and 4). The tumor was positive for vimentin; weakly positive for glial fibrillary acidic protein (GFAP); and negative for TTF1, CK7, CK20, pankeratin, synaptophysin, chromogranin, myogenin, progesterone receptor, MART1, and CD99. Further staining was positive for SOX2, SOX10, and OLIG-2, supporting a glial origin. A final diagnosis of glioblastoma, adenoid variant, was therefore made.

The patient was started on temozolamide and radiation therapy but was subsequently admitted about 6 weeks later with complaints of fever and diffuse back pain. MRI revealed extensive vertebral marrow signal abnormality and enhancement (Fig. 5). Additionally, there were multiple enhancing nodules involving the paraspinal muscles (Fig. 6). Subsequent bone biopsy revealed diffuse marrow tumor infiltration, histopathologically similar to the original glioblastoma (Fig. 7). Extensive

Abbreviations: PLA, primary leptomeningeal astrocytoma; GFAP, glial fibrillary acidic protein; GBM, glioblastoma.

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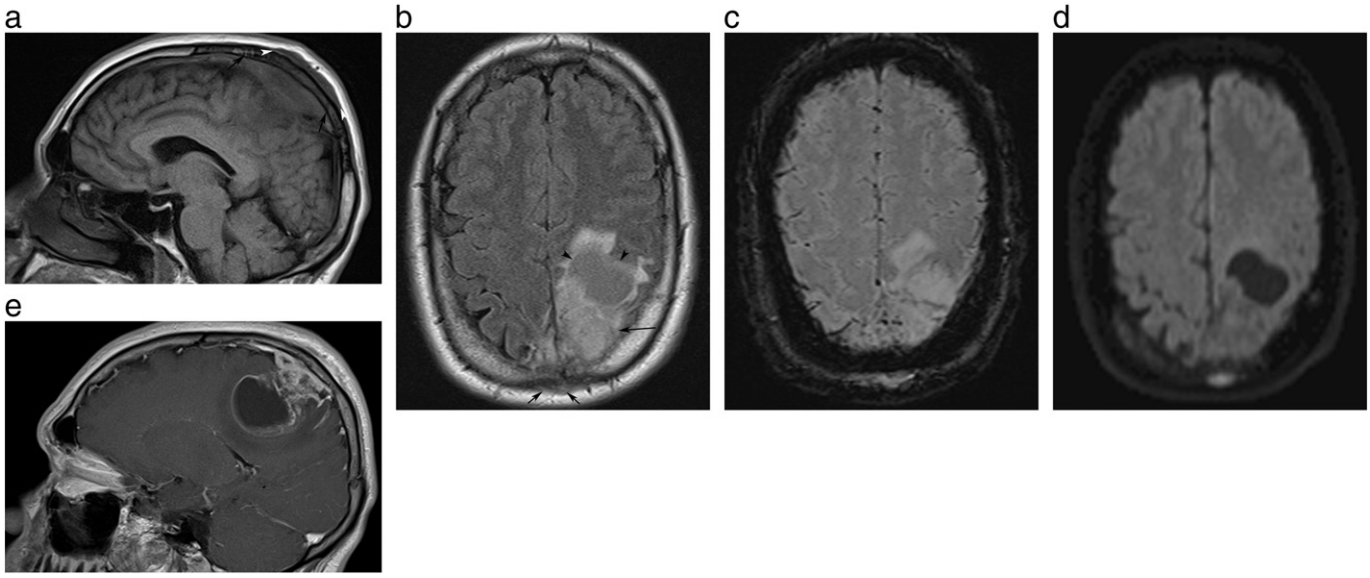

Fig. 1. (a–e) Sagittal precontrast image (a) reveals dural-based lesion in the left parafalcine region with extension through the overlying bone (black arrows) and a small subcutaneous component (white arrowheads). Axial fluid-attenuated inversion recovery image (b) again reveals left parafalcine tumor (long arrow) with a cystic component (arrowheads) and transcalvarial extension (small arrows). The lesion shows few microhemorrhages on susceptibility-weighted image (c) and foci of restricted diffusion on DW image (d). Postcontrast image (e) demonstrates the heterogeneous enhancement and unilocular cystic component.

infectious workup for the fever was negative, and it was eventually attributed to the tumor. During the same admission, the patient also developed acute left eye chemosis and proptosis. MRI revealed metastatic involvement of the extraocular muscles and left lateral orbital wall, not seen on the initial imaging studies (Fig. 8). The patient was also noted to have a chest wall skin nodule, which again proved to be metastatic on cytological examination.

Given the overall poor functional status, rapid tumor progression, and thrombocytopenia from ongoing chemoradiation and marrow infiltration, the patient was felt to be a poor candidate for more aggressive therapy. The patient was offered palliative therapy and chose hospice. The patient subsequently died 3 weeks later.

3. Discussion

Primary leptomeningeal astrocytomas, by definition, arise within the meninges, without any attachment to the neural tissue or evidence of primary neoplasm elsewhere within the neuraxis [2,5]. They are thought to arise from heterotropic nests of glial tissue within the meninges, originally described by Wolbach [1,2,6,7]. These may be seen in up to 1% of normal population, but have been noted in up to 25% of those with coexisting central nervous system (CNS) malformations, and often occur over medulla oblongata (57%), spinal cord (20%), pons (15%), midbrain (10.5%), cerebellum (8%), or the cerebral cortex (4%) [2,3,7]. Since these nests may contain oligodendrocytes, ependymal cells,

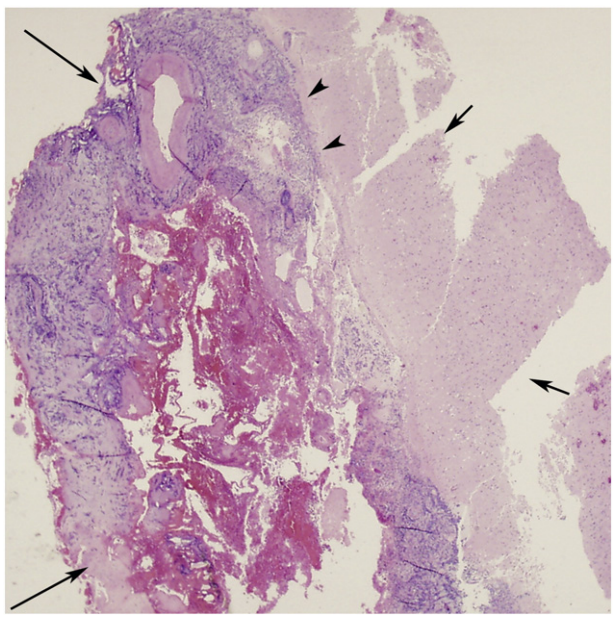

Fig. 2. Hematoxylin and eosin (H&E) 2× showing meningeal-based tumor (long arrows) with superficial cortical invasion (arrowheads) associated with reactive gliosis in the cortex (short arrows).

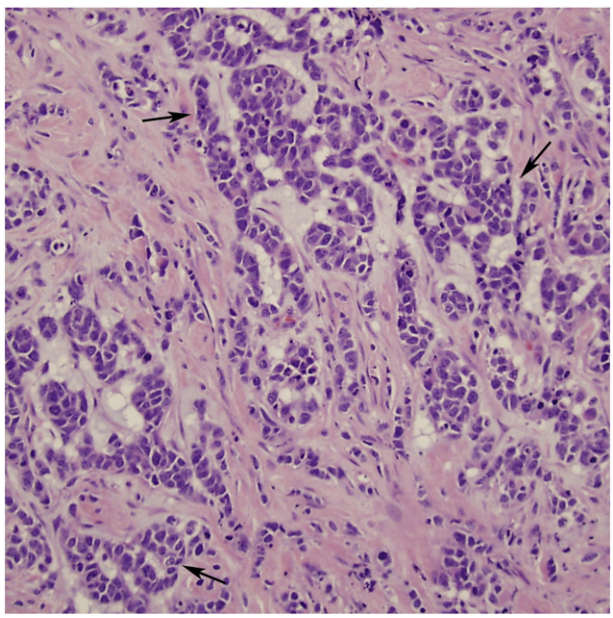

Fig. 3. H&E 40× illustrating tumor arranged in cords and trabeculae with focal gland formation (arrows). The neoplastic cells show moderate pleomorphism with high nuclear/cytoplasmic ratio, stippled nuclear chromatin, and occasional central nucleoli.