Neuroimaging Highlight

Editors: William Hu, Mark Hudon

Glioblastoma Multiforme and Ascending Weakness

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An 18-year-old man with a known glioblastoma multiforme, previously treated with de-bulking, chemotherapy, radiation and steroids, underwent a repeat craniotomy after presenting with several days of intractable vomiting and headache. Post-operatively, he developed progressive delirium, bilateral sixth and seventh nerve palsies, dysphagia and ascending weakness of the upper and lower limbs. Within five postoperative days, he was quadraparetic, areflexic and had marked extraocular muscle and facial weakness, dysphagia, dysphonia and respiratory muscle weakness. The patient was started empirically on intravenous immunoglobulin for suspected Guillain-Barré syndrome (GBS). The following day, lumbar puncture demonstrated elevated protein (1.96g/l; normal <0.45), and a reduced glucose (0.5 mmol/l) and elevated red ($4125x10^6$ per litre) and white cell ($214x10^6$ per litre) counts in the cerebrospinal fluid (the red cells were felt to be related to surgery). The cell differential included 69% neutrophils, 3% lymphocytes, 15% monocytes and 13% blast-like cells. Gram stains and cultures of the cerebrospinal fluid were negative.

Nerve conduction studies were normal with no evidence of primary demyelination. Needle electromyography identified diffuse lower limb denervation. The studies suggested the possibility of multiple root damage rather than GBS. Cerebrospinal fluid cytopathology revealed atypical cells, suspicious for malignancy.

Magnetic resonance imaging (MRI) demonstrated multiple enhancing and nonenhancing leptomeningeal nodules in the cervical, thoracic and lumbar spine, as well as the nerve roots of the cauda equina. There was no clear enhancement of the intracranial meninges. A diagnosis of spinal meningeal gliomatosis was made and the patient died two days later.

This patient had an unusual, fulminating presentation of spinal meningeal gliomatosis; a rare complication of glioblastoma multiforme. Meningeal gliomatosis may present with a variety of findings, including altered mentation, papilledema, cranial nerve palsies, motor weakness, ataxia and headache, but a presentation resembeling acute GBS has not been reported.[1-5]

Estimates of survival in cases of meningeal gliomatosis have

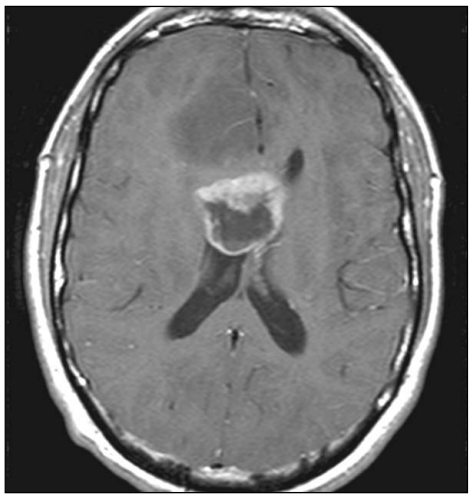

Figure 1: *Gadolinium-enhanced, T1-weighted, axial MRI showing the tumor at the time of initial presentation.*

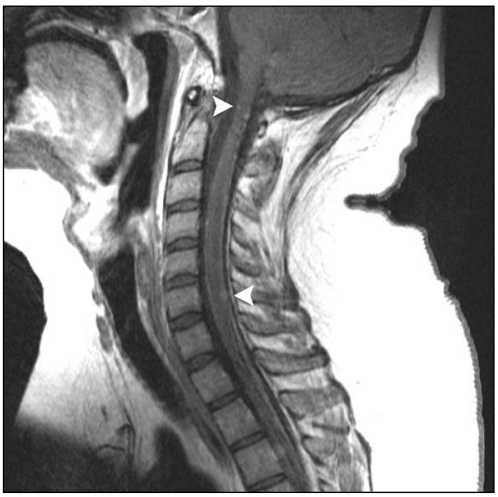

Figure 2: *Gadolinium-enhanced, T1-weighted, sagittal MRI of the cervical spine, demonstrating nodular enhancement of the leptomeninges.*

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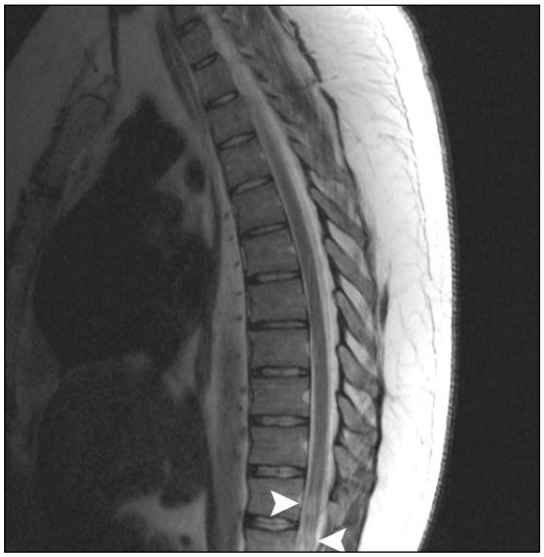


Figure 3: *T2-weighted sagittal MRI of the thoracic spine showing nodular leptomeningeal hyperintensity inferiorly.*

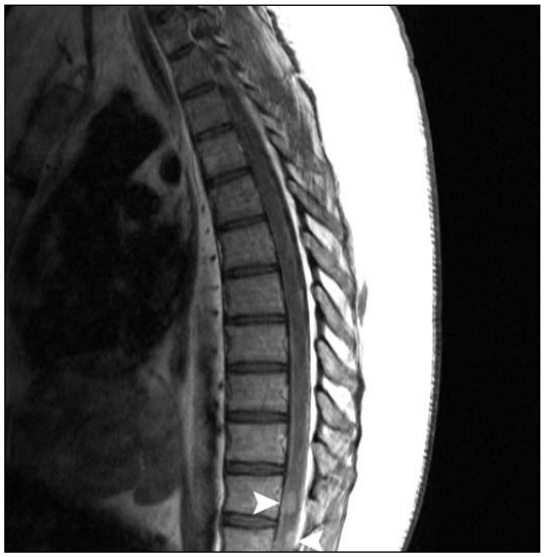
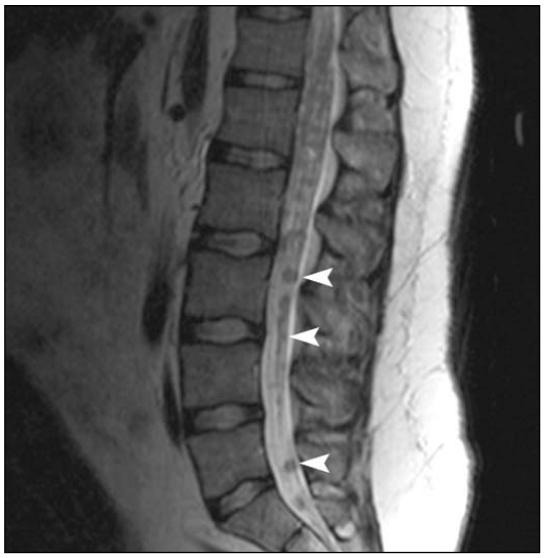


Figure 4: *Gadolinium-enhanced sagittal MRI of the thoracic spine showing nodular leptomeningeal enhancement.*



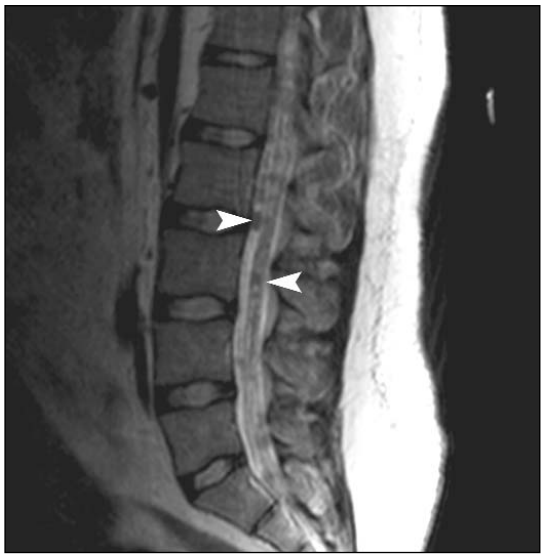


Figure 5 and 6: *Sagittal T2-weighted MRI of the lumbar spine. There is evidence of thickening and nodules over the nerve roots of the cauda equina.*

previously been reported to range from eight weeks to 10 months.[2,6] There is some evidence to suggest that younger patients and patients with tumors with PTEN mutations are more likely to develop meningeal gliomatosis.[3,4,7] Spinal meningeal gliomatosis may also masquerade as infection, stroke, transverse myelitis, motor neuron disease, or encephalitis.[8-10] One study suggests that it may, in fact, be more common than expected, with up to 20% of glioblastoma patients showing spinal leptomeningeal metastases at autopsy.[5]

Diagnosis is made on the basis of nodular enhancement along the meninges on gadolinium-enhanced MRIs. Elevated protein, pleocytosis with abnormal cells (75% of cases) and normal to low glucose in the cerebrospinal fluid may lend support to the diagnosis.[4,9] Cerebrospinal fluid histochemistry may also be useful.

When the diagnosis of GBS is suspected in a patient with glioblastoma multiforme, spinal meningeal gliomatosis must be considered and lumbar puncture and MRI must be performed urgently to differentiate between these two conditions. One should recall Occam's razor, first expressed by William of Occam in 1320: "what can be done with fewer assumptions is done in vain with more".[11]

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