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Meningeal Myelomatosis: CT and MR Appearances

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Summary: Meningeal myelomatosis is a rare feature of multiple myeloma. We report a case of IgG-κ myeloma presenting as bilateral intracranial extraaxial masses.

Index terms: Multiple myeloma; Meninges, neoplasms; Brain, neoplasms

Meningeal myelomatosis is a rare finding; at least 40 cases have been reported (1, 2). Men are affected more often than women by approximately 2:1, and those affected are 31 to 76 years old (1). In approximately one third of patients with meningeal myelomatosis, changes in mental status are the initial neurologic manifestation (1). Other reported clinical manifestations are lower extremity weakness, cranial nerve palsies, gait disturbance, headache, speech disorder, meningismus, seizure, vertigo, hemisensory loss, diffuse weakness, hemiparesis, muscle wasting, limb pain, and dysmetria (1). The diagnosis is usually made by finding myeloma cells in the cerebrospinal fluid. Previous reports show a disproportionate number of IgA (23%) and IgD (13%) tumors in patients with meningeal myelomatosis, which might indicate a propensity for these two rare subtypes to spread to the meninges (1). The remainder are IgG (23%), IgM (3%), light chain only (8%), and unspecified (33%). The median survival is 1 month in patients who had no initial response to therapy and 5 months in those who had an initial response (1).

Case Report

A 78-year-old woman with a history of colon carcinoma had a 2-week history of inappropriate behavior and urinary incontinence. The symptoms rapidly progressed to a decreased level of consciousness. Physical examination revealed normal reflexes, normal strength, and intact cranial nerve function. The patient was disoriented, dyspraxic, and remembered none of three items at 1 and 5 minutes. She made inappropriate statements when interviewed. Laboratory data were unremarkable. CT (Fig 1) and MR imaging (Fig 2) were performed. Our working diagnosis was meningeal carcinomatosis versus lymphoma. The patient underwent craniotomy, biopsy, and partial resection. The tumor involved dura mater, arachnoid, and pia mater. Histopathology showed diffuse sheets of plasma cells. Many cells were immature with enlarged vesicular nuclei and prominent eosinophilic nucleoli. Serum protein electrophoresis showed a monoclonal spike, and immunoelectrophoresis revealed an IgG-κ gammopathy. The patient was treated with whole-brain irradiation but died 3 weeks after presentation.

Discussion

Meningeal myelomatosis is a rare finding; at least 40 cases have been reported (1, 2). Men are affected more often than women by approximately 2:1, and those affected are 31 to 76 years old (1). In approximately one third of patients with meningeal myelomatosis, changes in mental status are the initial neurologic manifestation (1). Other reported clinical manifestations are lower extremity weakness, cranial nerve palsies, gait disturbance, headache, speech disorder, meningismus, seizure, vertigo, hemisensory loss, diffuse weakness, hemiparesis, muscle wasting, limb pain, and dysmetria (1). The diagnosis is usually made by finding myeloma cells in the cerebrospinal fluid. Previous reports show a disproportionate number of IgA (23%) and IgD (13%) tumors in patients with meningeal myelomatosis, which might indicate a propensity for these two rare subtypes to spread to the meninges (1). The remainder are IgG (23%), IgM (3%), light chain only (8%), and unspecified (33%). The median survival is 1 month in patients who had no initial response to therapy and 5 months in those who had an initial response (1).

CT findings of meningeal myelomatosis have been reported as peripheral, markedly hyperdense masses with associated parenchymal edema and mass effect (3). MR findings were reported for a case similar to ours in which a solitary extraaxial parietal mass was the presenting feature (2). The extraaxial masses of our patient were homogeneous on T2-weighted images, in contrast to the previously reported case, in which the extraaxial mass has heterogeneous signal (2). In our patient, diffuse homogeneous enhancement of the masses oc-
A central nonenhancing focus (2).

Meningeal myelomatosis has also been reported to show abnormal leptomeningeal enhancement in the cerebellar sulci (2) and can show abnormal enhancement of the cranial nerves (1). Other extraaxial mass lesions that can be isointense with gray matter on long-repetition-time and short-repetition-time images and can enhance homogeneously with intravenous contrast administration include meningioma, lymphoma, and carcinomatous meningitis.

In conclusion, extraosseous myelomatosis is an uncommon clinical form of multiple myeloma that is more aggressive and resistant to therapy. Meningeal involvement, reported in approximately 40 cases, demonstrates rapid progression. The intracranial finding of an extraaxial mass has been reported in two cases that had characteristics distinctly different from...
our case on CT and MR imaging. Meningeal myelomatosis should be considered in the differential diagnosis when slightly hyperdense subdural masses are seen on CT and when homogeneous lobulated meningeal masses that are isointense with gray matter on long-repetition-time and short-repetition-time images show diffuse homogeneous enhancement on MR imaging.

References

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