



Rare CNS Presentations of Multiple Myeloma with Intraparenchymal Metastasis

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Abstract

The purpose of this report is to present two rare cases of multiple myeloma that demonstrated diffuse intraparenchymal metastasis. Recognition of rare CNS parenchymal involvement by multiple myeloma is critical with regards to treatment options.

Case Presentation

Case 1

A 69 year old male presented with de novo IgG multiple myeloma diagnosed initially in 2007. The patient's initial treatment lasted from 2008 to 2009 when the patient was in complete remission requiring only maintenance therapy. However, the patient had a relapse in 2014. During this workup, the patient obtained an MR of the brain with contrast demonstrating leptomeningeal disease. The patient was treated a second time, however an MR of the brain with contrast in 2017 demonstrated a third relapse, and in 2018 an MR of the brain with contrast now demonstrated new enhancing intraparenchymal lesions.

Case 2

A 79 year old female presented with status epilepticus in the setting of multiple myeloma, breast cancer, and cholangiocarcinoma. MR imaging of the brain demonstrated diffuse hemorrhagic metastasis to the brain. A lumbar puncture was performed with the sample sent to cytopathology for analysis, demonstrating abundant single cells with plasmacytoid features, including mononuclear and binucleate cells and morphologically favoring multiple myeloma. Although the patient does have both breast cancer and cholangiocarcinoma, the CSF sample sent for cytology proved to have malignant cells consistent with multiple myeloma. An MRI of the brain demonstrated diffuse intraparenchymal metastasis throughout the cerebral hemispheres as well as diffuse leptomeningeal enhancement along the cerebellar folia. In this rare case, the patient demonstrated diffuse intraparenchymal metastasis in the setting of multiple myeloma.

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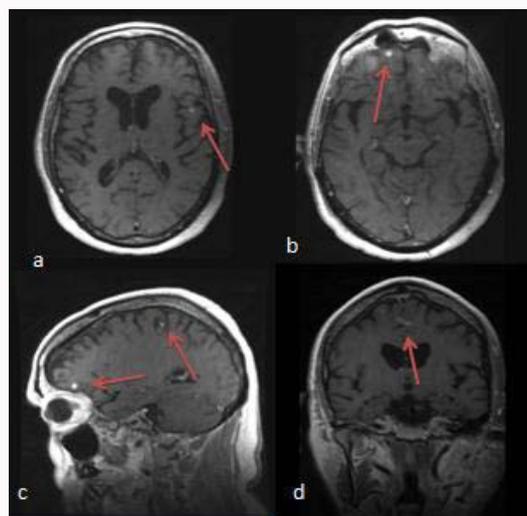


Figure 1: Imaging from Case Report #1: Axial (a, b), sagittal (c), and coronal (d) post-contrast MR imaging of the brain demonstrates multiple enhancing intraparenchymal lesions (arrows, a-c). There is also leptomeningeal disease (arrow, d).

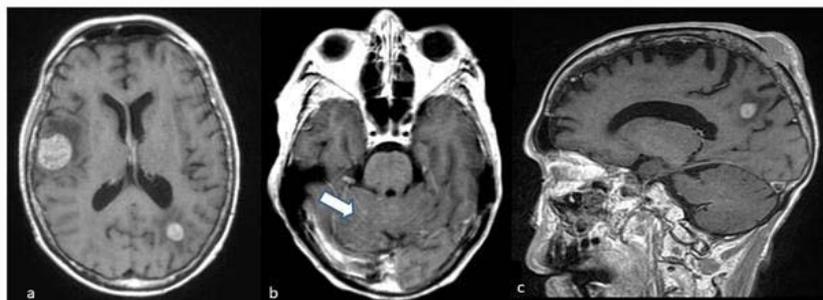


Figure 2: Imaging from Case Report #2: Axial post contrast (a, b) and sagittal post contrast (c) MR images of the brain demonstrate multiple enhancing parenchymal lesions as well as diffuse calvarial soft tissue masses. There is diffuse leptomeningeal enhancement noted along the superior cerebellum (arrow).

Description

Central nervous system involvement is a rare complication of multiple myeloma. Patients may present with either an intraparenchymal lesion or with a leptomeningeal lesion [1]. It is important to identify intracerebral metastasis from multiple myeloma because the tumor is radiosensitive and thus radiotherapy may play an important role in therapy [2]. The mechanism for the spread of plasma cells to the leptomeninges is not clear, however, it is thought that the plasma cells may extend to the central nervous system through microscopic veins in the arachnoid membrane [1].

Summary

Multiple myeloma is a neoplastic disorder of plasma cells that secrete a monoclonal immunoglobulin. The disorder represents approximately 1% of all cancers, approximately 10% of all hematologic malignancies, and is the most common primary osseous malignancy in adults [3,4]. Risk factors include exposure to ionizing radiation, as well as toxic exposures such as to benzenes, herbicides, and insecticides [5,6]. Clinical presentation can include bone pain, fatigue, and anemia with lethargy [3]. The highest incident of cases is from ages 50 to 70, with incidence increasing with age, and it is more common in men than women [7,8]. Multiple myeloma can affect multiple organs and organ systems. Extramedullary plasmacytoma is overall a less common manifestation of multiple myeloma, with 6% to 8% of patients newly diagnosed with multiple myeloma presenting with extramedullary manifestations, and with an estimated 10% to 30% of patients with relapsed or refractory multiple myeloma presenting with extramedullary manifestations [8,9]. Improvements in multiple myeloma therapy have also likely lead to an increase incidence of patients who are now able to present with extramedullary myeloma due to increased survival [3,8,10]. Typical locations of extramedullary plasmacytomas include lymph nodes, spleen, and liver, though there are reports of involvement of nearly every organ system [3].

The presence of extramedullary manifestations of diagnosed multiple myeloma is relevant to patient care as there is an association with higher relapse rates and with a genomically higher risk disease, as well as with shorter overall and disease-free survival [3,8,11-13]. In addition, any relapse of multiple myeloma is more likely to include extramedullary manifestations if they were present on initial diagnosis [14].

A retrospective review in 1963 by Silverstein et al. [15] demonstrated a 3% rate of intracranial metastases in 273 patients with multiple myeloma. More recently, the reported incidence rate for central nervous system involvement of multiple myeloma is 0.7% to 1% [16-19]. Most often the disease manifests as leptomeningeal

carcinomatosis, with intracranial plasmacytoma being a rarer manifestation [1,19,20]. There have been case reports discussing the occurrence of plasmacytoma in the sella, involving the temporal bone, and involving the intraocular muscles [4,21-24]. Typically myelomatous involvement of the central nervous system occurs in relapsing patients, rather than as the primary presentation. Diagnostic imaging, including contrast-enhanced MRI, has become a useful aid in diagnosis, staging, and post-treatment surveillance of multiple myeloma with possible extramedullary manifestations.

In the first case report, a patient with multiple relapses to multiple myeloma presents with imaging evidence of both leptomeningeal and intraparenchymal disease. In the second case report, a patient with known multiple myeloma, breast cancer, and cholangiocarcinoma, presents with diffuse hemorrhagic metastases resulting in a clinical presentation of status epilepticus. A subsequent lumbar puncture and CSF analysis including cytology demonstrates evidence of multiple myeloma. Although rare, central nervous system myelomatous involvement can cause many complications for the patient. While intracranial plasmacytomas are radiosensitive, there is an overall shortened survival in patients with multiple myeloma and concurrent extramedullary disease, including CNS involvement. Thus it is important for the radiologist to be aware of such presentation.

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