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Case Report

Purely Intraparenchymal Plasmacytoma: A Diagnostic Conundrum. To Operate or Radiate?

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ABSTRACT

Intracranial plasmacytomas are rare entities that are thought to spread to the intraparenchymal space via pre-existing lesions from the dura or skull. Purely intraparenchymal plasmacytomas, without cranial or dural involvement, are therefore, rarer and seem to contradict the mechanism of spread that have been attributed to these lesions. Only a few cases of purely intraparenchymal lesions have been described in the literature, and they represent diagnostic conundrums when identified as they mimic the radiographic appearance of other parenchymal lesions, such as metastatic tumors or infectious collections. The authors report an unusual case of a purely intraparenchymal plasmacytoma in the right frontal lobe of a 68-year-old female with a known history of multiple myeloma. No evidence of extension from a cranial or dural-based lesion was found radiographically or intraoperatively and given the non-diagnostic radiographic features of this plasmacytoma, the diagnosis was made upon pathological examination after gross total resection. A review of the literature on purely intraparenchymal plasmacytomas is provided and the management and prognosis of these lesions are addressed.

Introduction

Malignant plasma cell neoplasms encompass a range of disorders in which there is an aberrant monoclonal proliferation of bone marrow plasma cells and include multiple myeloma, isolated solitary bone plasmacytoma, and extramedullary plasmacytomas. Intracranial plasmacytomas are particularly uncommon but are still frequently found at the skull base and the dura mater [1-15]. Far fewer descriptions exist of purely intraparenchymal plasmacytomas without cranial or dural attachment, thus making these lesions true clinical rarities [16-19]. Reported here is a case of a purely intracranial plasmacytoma without any dural or cranial involvement that was discovered in the right frontal lobe of a middle-aged female with multiple myeloma.

Case Presentation

A 68-year-old female with the diagnosis of multiple myeloma presented to our institution with recurrent headaches and a worsening left hemiparesis. Six months prior she was treated for acute renal failure secondary to IgA kappa and kappa-free light chain nephropathy. At that time a bone marrow aspirate demonstrated hypercellular marrow with atypical plasmacytosis, consistent with the diagnosis of multiple myeloma. Her current examination was non-revealing except for a graded 3/5 strength in her left arm, and 4/5 strength in her left leg. She reported a progressively worsening of her left hemiparesis over the previous three weeks. MRI demonstrated a solitary, avidly enhancing right frontal mass anterior to the motor cortex with surrounding vasogenic edema, restricted diffusion, and marked elevation of the choline/creatine ratio on spectroscopy (Figures 1 and 2). Given the appearance of the lesion, uncertainty in diagnosis, and the patient's neurological decline, she was taken to the operating room for resection and biopsy was deferred given the accessibility of the lesion. Frozen sectioning demonstrated a hypercellular inflammatory lesion suggestive of plasmacytoma and final pathology demonstrated sheets of atypical cells expressing CD138 with MIB1/Ki-67 proliferation index greater than 90%, consistent with plasmacytoma (Figure 3). Post-operative MRI demonstrated gross-total resection of the lesion and the patient recovered

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well, with her left-sided strength returning to baseline by discharge from the hospital (Figure 4).

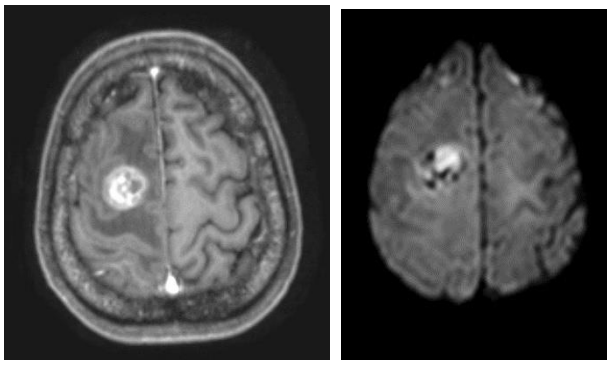

Figure 1: Contrast-enhanced and diffusion-weighted MRI demonstrating an avidly enhancing right frontal mass with surrounding vasogenic edema and restricted diffusion within the lesion, respectively.

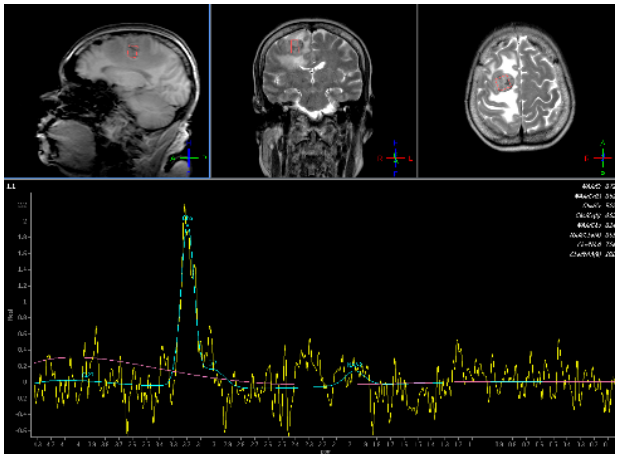

Figure 2: MR Spectroscopy demonstrating a choline peak centered within the lesion

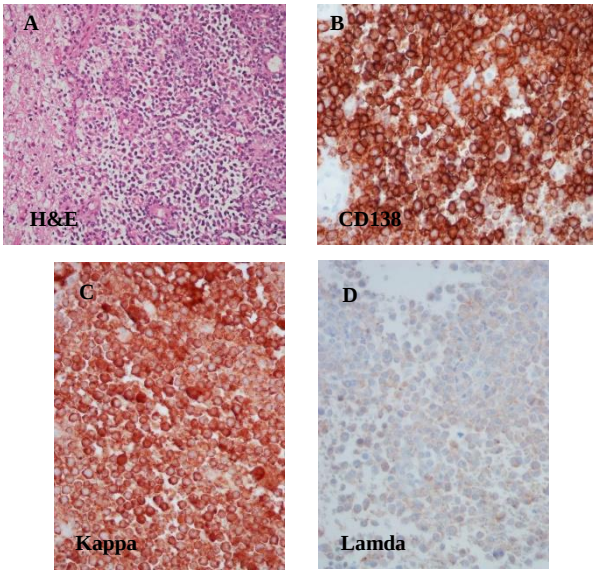

Figure 3: Pathology sections demonstrating hypercellular sheets of atypical cells (A) with avid CD138 staining (B), consistent with plasmacytoma. Kappa staining was much more pronounced (C) than Lambda staining (D).

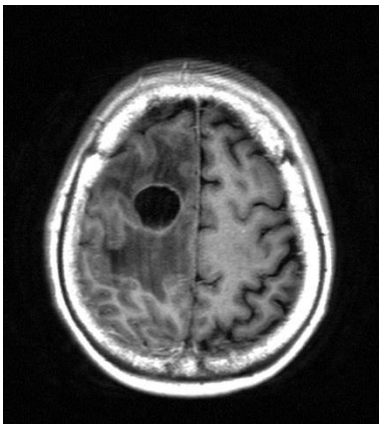

Figure 4: Postoperative contrast-enhanced MRI demonstrating gross total resection of the lesions with improved mass effect despite persistent edema.

Discussion

To the best of our knowledge, we have presented the first documented case of a purely intracranial plasmacytoma without evidence of cranial or dural involvement found in the right frontal lobe of a patient with pre-existing multiple myeloma. Given the rarity of a presentation of a purely intraparenchymal plasmacytoma, and its ill-defined radiographic features, we initially assumed the lesion was a benign primary tumor, possibly a metastatic lesion, and unlikely an infectious process. However, with our diagnosis questionable, and the patient presenting with progressive motor deficit, we opted for early surgical intervention and a definitive treatment with surgical total resection. The diagnosis of purely intraparenchymal plasmacytoma was made after pathological examination of the tumor.

Purely intracranial plasmacytomas are rare and poorly described in the literature. These intraparenchymal lesions have been reported in four other cases, in the lateral ventricle, temporal-insular lobe, occipital lobe, and cerebellum [16-20]. Since plasma cells are not naturally found in brain tissue, an isolated intraparenchymal lesion is challenging to explain without any evidence of direct extension from dural disease. Therefore, it has been suggested that parenchymal involvement may occur through hematogenous dissemination of progenitor plasma cells or through plasma cells acquiring cytogenetic characteristics, such as 17p13 deletions [12, 20-22].

Multiple myeloma patients with central nervous system involvement usually have a p53 deletion, elevated LDH levels, and more frequently had either IgA or IgD subtypes of myeloma. They also have a younger average age of onset at 53 years [23]. Central nervous system involvement confers an aggressive profile and an even worse prognosis than traditional multiple myeloma [24]. Once the central nervous system is involved, the median overall survival is from one year to twenty-five months [4]. Treatment protocols for multiple myeloma with extramedullary central nervous system involvement have not been well established [25].

Since solitary intracranial plasmacytomas carry a particular treatment regimen and are highly radiosensitive, efforts have been made to identify

its features radiographically. Magnetic resonance imaging of an intracranial plasmacytoma is usually homogeneous and hypointense compared with skeletal muscle on T1-weighted images and hyperintense on T2-weighted images and exhibits mild to intense enhancement [26]. Still, radiographic characteristics are not satisfactory for the confirmation of a plasmacytoma and other pathologies must be identified or excluded. Diagnosing these lesions remains a diagnostic challenge and it necessitates histopathological results and a comprehensive evaluation to rule out the potential presence of alternative neoplasms with metastases to the brain, among other pathologies.

Although a challenging diagnosis to make, atypical intracranial lesions found in patients with multiple myeloma should raise suspicion of an intraparenchymal plasmacytoma. The present case is unique in that the reported patient had a right frontal plasmacytoma without skull base or dural attachment who had complete cure and resolution of her neurological symptoms. We believe a viable treatment option for these lesions includes total resection, when easily accessible, and a biopsy and standard radiation when resection is more complex. Immediate pathological examination is critical in these cases intraoperatively.

Conclusion

A purely intraparenchymal plasmacytoma without existing dural attachment or cranial involvement is extremely uncommon. Since these entities present without distinguishing clinical or radiographic features, they are a particularly difficult preoperative diagnosis to make. Even so, the imaging for our patient did not take on the characteristic appearance of a plasmacytoma and diagnosis was only made postoperatively with pathology. Biopsy versus gross total resection is a decision made based on the location of the lesions, the patient's neurological examination, and the surgeon's and patient's preference.

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