Solitary Plasmacytoma of the Petrous Apex: A Case Report and Review of Literature

Tyler S. Quist, BS1; Cheryl A. Palmer, MD2; Joel D. MacDonald, MD3; Richard K. Gurgel, MD1

1University of Utah Division of Otolaryngology, 2University of Utah Department of Pathology, 3University of Utah Department of Neurosurgery

Abstract

OBJECTIVES: To describe the clinical presentation, evaluation, and treatment of solitary plasmacytoma of the bone (SPB).

STUDY DESIGN: Retrospective case review.

RESULTS: A 60-year-old man presented with sudden right sensorineural hearing loss, tinnitus, loss of balance, and jaw pain. Computed tomography and magnetic resonance imaging revealed a possible meningioma and the mass was resected. Pathology revealed solitary plasmacytoma. The patient was evaluated to rule out multiple myeloma and underwent localized radiotherapy for treatment.

CONCLUSIONS: Patients with SPB may present with cranial neuropathy. SPB can be mistaken for meningioma when it involves the petrous apex. Pathological examination is required for diagnosis.

Introduction

Plasmacytomas are neoplasms that arise from a monoclonal proliferation of plasma cells. These tumors develop most commonly within soft tissue of the head and neck or the axial skeleton (vertebra or skull) and are classified as either extramedullary plasmacytomas (EMPs) or solitary plasmacytomas of the bone (SPBs), respectively. The clinical presentation of skull base plasmacytomas often involves frontal headaches and cranial neuropathies. The diagnosis of solitary plasmacytoma requires exclusion of systemic multiple myeloma. SPBs involving the skull in the absence of systemic signs are very rare and these tumors often develop into multiple myeloma. The prognosis is often worse for patients with a SPB when compared to EMPs because SPBs more frequently progress to multiple myeloma. In this study, we report a case of an isolated, unilateral osseous plasmacytoma of the petrous apex that presented with profound unilateral sensorineural hearing loss in the absence of systemic signs or symptoms.

Case Presentation

A 60-year-old man presented with a several month history of sudden right hearing loss. His symptoms also included tinnitus, loss of balance, and jaw pain. Physical exam was remarkable for hearing loss in the right ear only. Audiometry showed a pure tone average (PTA) and speech discrimination score (SDS) of 70 dB and 0% in his right ear, respectively. In his left ear, PTA and SDS scores were 25 dB and 88%, respectively. Computed tomography (CT) showed cortical thinning and irregularity involving the right petrous apex with involvement of the carotid canal and the superior aspect of the basal turn of the cochlea (Fig. 1A). Magnetic resonance imaging (MRI) revealed a homogeneously enhancing mass that appeared to arise from the dura of the middle cranial fossa and extend into the petrous apex and measured 2.1 cm anterior-posterior x 2.3 cm transverse x 1.3 cm craniocaudal (Fig. 1B). The lesion did not extend beyond the petrous bone nor involve the cavernous sinus. Meningioma was suspected and surgical resection was performed via a middle fossa approach which was tolerated well without complications. Intraoperative examination revealed a bony erosive lesion extending from the dura down into the petrous apex.

Pathological evaluation of the mass revealed a plasma cell tumor rather than meningioma. Microscopic evaluation revealed sheets of small round, monomorphic collections of plasma cells (Fig 2A). Mitoses or necrosis was not present and the tumor infiltrated bone areas. Immunohistochemistry was diffusely positive for syndecan-1 (CD138); a plasma cell marker (Fig 2B) and epithelial membrane antigen. The mass was negative for broad-spectrum cytokeratins and glial fibrillary acidic protein. Lambda and kappa in situ hybridization studies showed a lambda light chain restriction in the plasma cell tumor (Fig 2C).

Once the proper diagnosis was established, the patient underwent an extensive workup to rule out multiple myeloma. Multiple myeloma was excluded and the patient underwent localized radiotherapy, receiving a total dose of 45 Gy in 25 fractions using Intensity-Modulated Radiation Therapy (IMRT).

Discussion

Clinical features of SPBs involving the skull base may include frontal headaches and cranial neuropathies. SPBs occurring in the vertebral column, sternum, or ribs most commonly present with local pain and or swelling secondary to bone destruction. By definition, these patients lack systemic involvement and thus do not have CRAB (hypercalcemia, Renal insufficiency, Anemia, multiple Bone lesions) features associated with myeloma. These tumors present in men more than women and most commonly presents in the fifth decade of life. Our patient’s clinical symptoms, gender, and age are consistent with the previously published literature regarding SPBs.

Diagnostic criteria for SPBs requires a lack of CRAB features associated with multiple myeloma, biopsy proving plasma cell infiltration, bone marrow biopsy showing normal bone marrow, and skeletal survey showing a single bone lesion. A lack of CRAB features includes normal calcium level, normal kidney function, low M-component (IgG < 5 g/dl and IgA < 3 g/dl, and urine light chains < 4 g/24 h), and hemoglobin > 10 g/dl. Microscopic examination of the biopsy should reveal sheets of monoclonal plasma cells defined by CD138 surface marker, which are either kappa or lambda restricted. Bone marrow biopsy must show less than 10% plasma cells. Our patient meets criteria for SPB and does not have any laboratory, pathology, or findings on imaging concerning for multiple myeloma.

Management options for solitary bone plasmacytomas include radiotherapy, surgical excision, chemotherapy or a combination of these options. Currently, radiotherapy (RT) is the standard of care and provides local control that can result in long remission or even cure. Thus, radiation therapy is used following gross excision of plasmacytomas to eliminate microscopic residual disease. Our patient underwent surgical resection initially as the preoperative diagnosis was meningioma. Once the diagnosis of SPB was established, our patient underwent radiotherapy, receiving a total dose of 45 Gy in 25 fractions using IMRT.

In terms of prognosis, patients with SPB may develop multiple myeloma (MM), local recurrence, or new bone lesions in the absence of MM. The rate of progression is much higher for SPB compared to EMP and patients who progressed from SPB to MM have a worse prognosis than patients with MM who progressed from EMP. Frassica et al reported that of 46 cases of SPBs, 54% developed MM, 2% failed with new lesions without MM, and 11% developed local recurrences with a median time to progression of 18 months. Given the high rates of progression, our patient require continual follow up.

Conclusions

Solitary plasmacytomas of the bone are rare, especially in the absence of systemic involvement or CRAB symptoms associated with multiple myeloma. Patients with a SPB may present with cranial neuropathy, such as unilateral sensorineural hearing loss, or other neurological deficits. SPB can be mistaken on CT scan or MRI for meningioma when it involves the petrous apex. Thus, following CT and MRI evaluation, pathological examination is crucial for proper diagnosis of solitary plasmacytomas of the bone.

References: