CASE REPORT

Solitary Intramedullary Plasmacytoma of the Skull Base Mimicking Aggressive Meningioma

Joel R. Meyer, M.D., Sudipta Roychowdhury, M.D.,
George Cybulski, M.D., and
Eric J. Russell, M.D. FACR

ABSTRACT—The authors present an unusual case of solitary intramedullary plasmacytoma. Awareness of this entity can facilitate appropriate surgical planning, which may include a limited biopsy prior to considering more extensive skull base surgery.

Solitary intramedullary plasmacytoma is an uncommon neoplasm that can involve the skull base.¹⁻⁴ A significant number of patients with solitary plasmacytoma, up to 85%, will eventually develop multiple myeloma.^{2,5,6} We report one such case where the preoperative imaging diagnosis favored aggressive meningioma or hemangiopericytoma. An intraoperative biopsy and frozen section were obtained prior to more extensive resection, thereby establishing the diagnosis of plasmacytoma. The patient was treated with radiation therapy, with a rapid effective response, thereby sparing the patient more extensive skull base surgery. Recognition of the invasive nature of this neoplasm is important to suggest its diagnosis preoperatively.

CASE REPORT

A 54-year-old white female with no significant past medical history presented with a 2-week history of left-sided exophthalmos following several months of antibiotics for presumed sinus inflammatory disease. Her neurologic examination was otherwise normal.

A screening sinus CT (Fig. 1) revealed a destructive mass lesion along the floor of the left middle cranial fossa associated with lytic destruction of the greater wing of the sphenoid and the left pterygoid plate. The mass extended inferiorly into the infratemporal fossa and medially into the sphenoid sinus.

MRI revealed a $5 \times 5 \times 7$ cm mass involving the lesser and greater sphenoid wings extending into the left middle and anterior cranial fossae extra-axially (Fig. 2). The tumor also extended within the left infratemporal fossa, sphenoid sinus, and the lateral extraconal compartment of the left orbit. The epicenter of the lesion was within the diploic space of the floor of the middle cranial fossa, indicating the site of probable origin. Prominent fronds of soft tissue extension were thought to represent "mushrooming" of the mass such as may be seen with aggressive meningioma or a hemangiopericytoma. The mass was isointense to brain on T_1 -weighted images and demonstrated homogenous enhancement following the administration of intravenous contrast agent (gadopentate dimeglumine, 0.1 mmol/kg). The lesion was heterogeneous in signal intensity on T_2 -weighted images, with the bulk of the extra-axial tumor isointense to gray

Figure 1. Screening coronal sinus CT reveals a mass (arrow) along the floor of the left middle cranial fossa with lytic destruction of the left greater sphenoid wing and pterygoid plate

matter. Distinct flow voids were demonstrated along the periphery of the lesion suggesting capsular hypervascularity. Preoperative differential diagnosis included aggressive meningioma, hemangiopericytoma, and metastatic disease. Other neoplasms and inflammatory disease were less likely differential diagnostic considerations.

The patient underwent preoperative angiography, which demonstrated a hypervascular mass supplied by the distal left internal maxillary branches and the left middle meningeal artery (Fig. 3). Late arterial and capillary phase images showed prolonged vascular blush similar to the angiographic appearance of meningioma. Preoperative embolization of the external carotid supply to the lesion was successfully performed with 300–500 μ polyvinyl alcohol.

A bifrontal craniotomy revealed extensive extradural tumor that infiltrated the temporalis muscle and the calvarium. A biopsy and frozen section were performed intraoperatively that demonstrated a highly cellular lymphocytic lesion suggestive of a plasma cell tumor. No further resection was performed, as plasmacytoma can be treated with radiation therapy alone.^{2,6} Serum protein electrophoresis demonstrated paraproteinemia, and bone marrow aspirate performed after the surgery was diagnostic for multiple myeloma. Plasma cells accounted for 6% of the marrow aspirate with predominately lambda staining cells. The patient was subsequently treated with local radiation therapy to the left

skull base. Several months later, the patient developed advanced systemic multiple myeloma.

DISCUSSION

Solitary intramedullary plasmacytoma (SIP), a subtype of plasma cell tumors, is a malignant neoplasm that may occur as a primary focal entity or as the first presentation of systemic multiple myeloma.^{2,5,6} Plasma cell tumors are classified as either multiple myeloma (systemic), intramedullary plasmacytoma (involving bone), or extramedullary plasmacytoma (involving soft tissues).⁵ There is considerable controversy whether plasmacytomas are distinct entities or represent an early form of systemic multiple myeloma.^{4,5} A significant percentage (up to 85%) of patients with solitary plasmacytoma will eventually develop disseminated multiple myeloma, if followed for a sufficient period of time.^{2,5,6}

The distinction between SIP and multiple myeloma is important because the primary treatment modality for SIP is radiotherapy in contrast to systemic multiple myeloma, which is treated with chemotherapy.^{2,5,6} Fine-needle aspiration may not provide an adequate tissue sample for making the diagnosis of plasmacytoma, and a core biopsy of the mass is usually necessary.^{2,5} Traditional surgical approach to plasmacytoma of the skull base involves a limited biopsy without radical resection, as these tumors are highly radiosensitive.² Others have suggested more radical approaches to accomplish complete surgical resection followed by adjuvant radiation therapy.⁷

Plasma cell tumors involving the cranium are most commonly a manifestation of systemic multiple myeloma; however, solitary plasmacytoma also rarely occurs.^{2–4,8} Solitary intracranial plasmacytoma is a rare neoplasm that has a mean age of 47 years and a female predominance similar to meningioma.^{8,9} A monoclonal cell line, as evidenced by a predominant population of either kappa or lambda chains, confirms the diagnosis of plasma cell tumor.^{4,5} Histologically, plasma cell neoplasms must be differentiated from plasma cell granuloma and meningioma with plasma cell infiltration.⁴ The prognosis for SIP is better than disseminated multiple myeloma.¹⁰ Intracranial plasmacytomas are associated with cranial nerve palsies and clinical features of other large space-occupying lesions such as headaches, visual problems, or seizures.¹⁰

The typical radiologic feature of plasmacytoma is osteolytic involvement of the skull base with variable aggressive bone destruction.⁴ Classically, calvarial lesions in multiple myeloma are multiple and characterized by punched out osteolytic lesions confined to the skull. On the other hand, plasmacytomas often present as solitary intra- and extracranial masses that bridge the diploic site of origin. They are often well advanced at

Figure 2. Axial precontrast T₁-weighted MRI shows a 5 × 5 × 7 cm extra-axial mass (arrow) in the left middle cranial fossa with extension into the sphenoid sinus (curved arrow). **B.** There is homogenous enhancement of the mass (arrows) on axial post-contrast T₁ weighted images. **C.** Coronal postcontrast T₁-weighted sequence reveals that the mass is centered within the diploic space of the floor of the middle cranial fossa with extension into the middle cranial fossa (curved arrow), sphenoid sinus (open arrow), and infratemporal fossa (arrow). **D.** Axial T₂-weighted image demonstrates a heterogeneous extra-axial mass (arrow) with peripheral flow voids, suggesting hypervascularity (curved arrow).

Figure 3. Selective lateral left external carotid digital subtraction angiogram shows a hypervascular mass supplied by the left middle meningeal artery and the distal internal maxillary arteries with a prolonged vascular blush (arrows).

the time of diagnosis and typically present as a large mass.⁴ SIP may extend from the skull, as in our case, to involve the suprasellar cistern, cavernous sinus, or infratemporal fossa.³⁻⁵ However, imaging findings are often nonspecific, and there may be considerable overlap with a variety of neoplasms and inflammatory processes affecting the skull base.

On CT, SIPs have been reported as slightly high attenuation extra-axial masses that often demonstrate homogenous contrast enhancement, very similar to meningioma.^{5,9,10} On MRI, plasmacytomas are often indistinguishable from meningioma, with intermediate signal on T_1 -weighted sequences and isointense signal to gray matter on proton density and T_2 -weighted sequences. With contrast, they frequently demonstrate homogenous enhancement, similar to our case.⁸ On angiography, SIPs are commonly hypervascular masses supplied by external carotid artery branches with prolonged capillary tumor stain that may also resemble meningioma.^{10,11}

Differential diagnostic considerations in this case included aggressive meningioma or hemangiopericytoma. On noncontrast CT, meningiomas are typically sharply circumscribed high attenuation dural based extra-axial masses that may or may not have distinct calcification.¹ On MRI, meningiomas are typically hypointense to isointense on T_1 -weighted images and isointense to hyperintense on T_2 -weighted images.^{1,12} The hypervascularity of meningiomas is associated with homogenous enhancement on both CT and MRI. Flow voids at the periphery of the lesion also support the hypervascular nature of these lesions. Imaging features of meningioma that may overlap with other skull base neoplasms such as plasmacytoma include marked peritumoral edema, little or no calcification, necrosis, irregular tumor margins, and "mushrooming"

(prominent peripheral tumor pannus).^{1,13,14} The bony change most commonly associated with meningioma is focal hyperostosis at the site of dural attachment, but bone destruction by transosseous growth of tumor can also occur.¹ However, the occurrence of lytic bone destruction with prominent extra-cranial involvement is atypical of meningioma and should suggest aggressive meningioma, hemangiopericytoma, or metastatic disease.

Hemangiopericytoma is a highly vascular neoplasm that arises from vascular pericytes and is commonly grouped with aggressive meningiomas with regard to radiographic appearance.¹ Imaging features suggestive of a hemangiopericytoma include a multiloculated contour, a narrow dural base or mushroom shape, large intratumoral flow voids, and bone erosion.¹⁵

The features of this case that suggest the diagnosis of SIP are the epicenter of the lesion within the diploic space of the floor of the middle cranial fossa and the lytic bone destruction of the calvarium. While meningiomas and hemangiopericytomas may resemble the imaging characteristics of this case, the location of the tumor and the aggressive bone changes should raise other possibilities such as plasmacytoma or metastatic disease.

With recent advances in imaging and operative technique, surgical approaches to the skull base have significantly evolved, resulting in improved resectability of large transcranial lesions. Since skull base surgery may require extensive disassembly of the craniofacial skeleton to facilitate tumor resection, preoperative or intraoperative recognition that the lesion may be a radiosensitive tumor, such as plasmacytoma, may spare a patient extensive surgery. Although there are some surgeons who advocate combined surgery and radiation for

the treatment of SIP,^{7,8} in this case it was thought that if the SIP responded well to local radiation therapy, the patient could avoid extensive craniofacial dissection and tumor resection. While fine-needle aspiration rarely yields an adequate tissue sample to make a definitive diagnosis of plasmacytoma, a limited open biopsy or an image-guided core biopsy may help establish the diagnosis and facilitate appropriate oncologic management.

In conclusion, solitary intramedullary plasmacytoma should be considered in the differential diagnosis of aggressive skull base neoplasms. In fact, SIP can mimic meningioma or hemangiopericytoma; however, the epicenter of the lesion and lytic bone destruction should raise the possibility of plasmacytoma. Awareness of this entity can facilitate appropriate surgical planning, which may include a limited biopsy prior to considering more extensive skull base surgery.

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