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Craniocerebral Plasmacytoma: MR Features

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Summary: We report the MR imaging findings in two patients with solitary craniocerebral plasmacytoma, a benign plasma cell tumor that can arise from the skull, the dura, or, rarely, the brain. In both patients, the lesion was extraaxial and nearly isointense with gray matter on T2-weighted MR images, and diffusely enhanced after administration of contrast material, bearing some similarities to meningioma. A diagnosis of solitary craniocerebral plasmacytoma should be considered when a mass with these imaging features is seen, because total excision may not be necessary for this radiosensitive tumor.

Index terms: Brain, neoplasms; Skull, neoplasms

The skull and central nervous system are rarely involved by plasma cell tumors without evidence of a plasma cell dyscrasia at another site (1). Solitary craniocerebral plasmacytomas are uncommon plasma cell tumors that produce monoclonal immunoglobulin and occur in the absence of other clinical, radiologic, or laboratory evidence of a plasma cell dyscrasia (1). There are two forms of solitary craniocerebral plasmacytoma: primary plasmacytomas arising from the skull and intracranial extramedullary plasmacytomas arising from the dura or, rarely, the brain (2). The radiologic appearance of solitary craniocerebral plasmacytoma can simulate that of more common neoplasms. Magnetic resonance (MR) imaging findings of craniocerebral plasmacytoma have rarely been reported (3). We describe the MR imaging characteristics of solitary craniocerebral plasmacytomas in order to show typical findings and to emphasize the similarity to meningiomas.

Case Reports

Case 1

A 42 year-old woman was examined for a palpable softening in her occiput. MR imaging showed a homoge-

neously enhancing mass arising within the occipital bone at the level of the torcular, displacing the dural sinus anteriorly (Fig 1). The lesion eroded both the inner and outer tables of the skull. A biopsy specimen of the lesion showed a diffuse infiltrate of atypical plasmacytoid cells that expressed IgG- κ , consistent with plasmacytoma of the skull. Results of subsequent urine protein electrophoresis, peripheral blood evaluation, bone marrow biopsy and aspirate, 24-hour urine sample for proteinuria, bone scan, and metastatic bone survey were negative. The patient was treated with radiation therapy.

Case 2

A 67-year-old woman was found to have a left-sided inferior homonymous hemianopsia during an examination for glaucoma. MR imaging showed large extraaxial mass lesions involving the tentorium cerebelli, falx cerebri, and extending through the dura over the convexities (Fig 2). At one point, the lesions appeared to be connected by a slightly thickened segment of dura. The preoperative diagnosis was meningioma. At surgery, a reddish avascular mass, which was densely adherent to adjacent dura and brain, was found along the falx cerebri and tentorium cerebelli. A biopsy specimen revealed a highly cellular tumor in which there was monotypic IgG- κ in most of the plasma cells, consistent with the diagnosis of plasmacytoma arising from dura. Following subtotal resection of the mass, the patient was treated with radiation therapy. A small IgG- κ M-component was noted on immunoelectrophoresis, but no evidence of myeloma was found on complete blood count, skeletal survey, or bone marrow aspirate.

Discussion

Solitary craniocerebral plasmacytomas are lesions that, by definition, arise from the skull or its contents, are not the result of extension from extracranial skeletal sites of myeloma, and are not associated with plasmacytosis within bone

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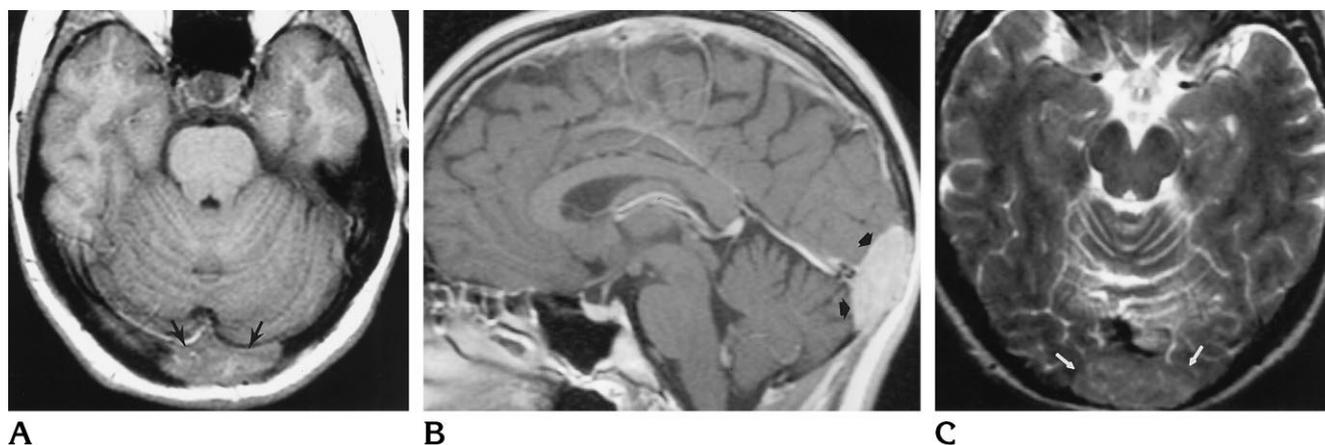
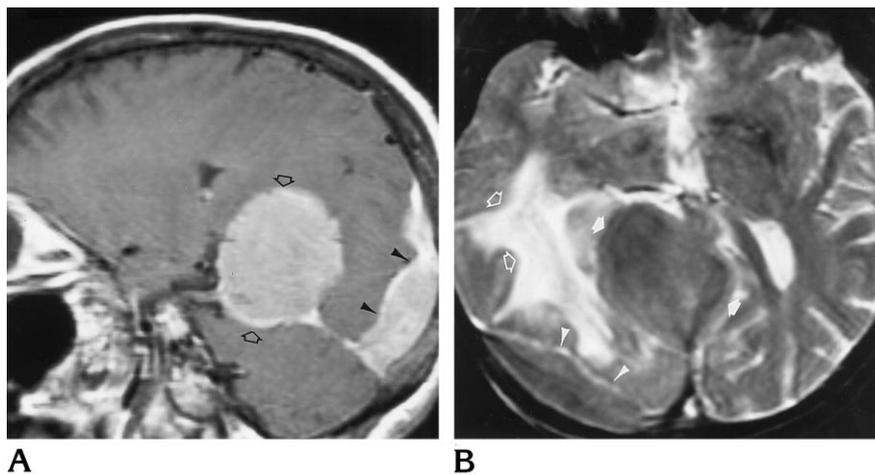


Fig 1. A 42-year-old woman with a palpable softening in the occiput.
 A, Noncontrast T1-weighted axial MR image (500/11/2 [repetition time/echo time/excitations]) shows a slightly inhomogeneous mass (*arrows*) that is nearly isointense with gray matter arising within the occiput.
 B, Contrast-enhanced T1-weighted (450/11) sagittal MR image shows a homogeneously enhancing mass centered at the level of the torcular, displacing adjacent brain.
 C, T2-weighted axial MR image (5500/108 effective) shows a mass (*arrows*) that erodes bone, is mildly inhomogeneous, and slightly hyperintense relative to gray matter.

Fig 2. A 67-year-old woman in whom left homonymous hemianopsia developed as a result of multifocal plasmacytoma arising from dura.

A, Contrast-enhanced T1-weighted (500/11) right parasagittal MR image shows a multifocal mass with the larger portion (*open arrows*) located at the anterior portion of the tentorium cerebelli and the smaller portion (*arrowheads*) arising along the dura overlying the convexity. On a more midline image (not shown), the two lesions were connected by a slightly thickened segment of dura.

B, T2-weighted (2250/80) axial MR image shows both the anterior component (*solid arrows*) and the posterior component (*arrowheads*) of the tumor. Most of the tumor is isointense with gray matter, but portions are slightly hypointense. A large amount of vasogenic edema (*open arrows*) extends into the right temporal lobe.



marrow or extracranial myeloma (4). Solitary craniocerebral plasmacytoma is a separate entity from the far more common plasma cell tumor that arises within the cranium as a result of disseminated multiple myeloma (1). The distinction between the two entities is important because solitary craniocerebral plasmacytoma is a relatively benign entity that is potentially curable, whereas multiple myeloma generally has a poor prognosis (1, 5). Solitary craniocerebral plasmacytoma is also a distinct entity from plasma cell granuloma, which is an inflam-

matory pseudotumor characterized by a benign proliferation of polyclonal plasma cells (unlike the monoclonal population seen in plasmacytoma), lymphocytes, histiocytes, and many small blood vessels (6). Although solitary craniocerebral plasmacytoma is not associated with extracranial disease, an abnormal monoclonal immunoglobulin can occasionally be detected within serum or urine (7).

Once the histologic diagnosis of plasmacytoma is reached, a systemic plasma cell dyscrasia must be excluded by bone marrow eval-

uation, serum protein electrophoresis for presence of a monoclonal gammopathy, and bone scan or skeletal survey. A monoclonal population of cells excludes both plasma cell granuloma and meningioma with conspicuous plasma cell components (5).

Solitary craniocerebral plasmacytoma is the least common form of extramedullary solitary plasmacytoma (2). The most common sites of solitary plasmacytomas arising from the skull are the parietal bone and the bones of the skull base (2, 7). The dura is the most common site of craniocerebral plasmacytoma not originating in the skull, occasionally secondarily involving the calvaria and brain parenchyma (4). Rarely, solitary plasmacytoma can arise within brain tissue (1).

Computed tomographic (CT) findings in solitary plasmacytoma of the skull are those of an expansile, lytic lesion involving the diploic space and the inner and outer tables of the skull, typically compressing adjacent brain (2). Diffuse, homogeneous enhancement after contrast administration is generally seen (2). Intratumoral calcifications due to bone destruction are often present, simulating calcifications seen in meningiomas (2). The extraaxial location, sharp margins between tumor and brain, signal characteristics, and enhancement pattern of the lesion in our first case bore some similarities to meningioma. However, erosion of the full thickness of the skull made meningioma an unlikely diagnosis. The findings in a previous report of MR imaging of dural plasmacytoma bore similarities to those in our second patient, including large size and a large amount of associated edema (3). Whereas the lesion in that report was hypointense relative to gray matter on T2-weighted images (3), the lesions in our patients were isointense or slightly hyperintense. Previous reports of the MR imaging appearance of solitary plasmacytoma of the skull have described a slightly inhomogeneous, expansile mass eroding bone that is isointense with brain on noncontrast T1-weighted images and isointense or slightly hyperintense on T2-weighted images (8, 9), similar to the findings in our first patient. Solitary plasmacytoma of the skull must be distinguished from multiple myeloma, which can produce an extradural mass (10). However, multiple lytic lesions are typically present in multiple myeloma of the skull (unlike solitary plasmacytoma) and, on MR imaging, the associated extradural mass has signal fea-

tures that are more inhomogeneous than in our first patient (10).

The lesions in our second patient consisted of dural masses with well-defined, smooth margins, near-isointensity with brain on both T1-weighted and T2-weighted images, and a homogeneous contrast enhancement pattern. These features resulted in the mistaken preoperative diagnosis of meningioma. In fact, dural plasmacytomas share a number of features with meningiomas. Both lesions usually occur in middle or late life, are more common in women, are sometimes many centimeters in size, have a predilection for similar sites within the intracranial vault (ie, the sphenoid ridge, cerebral convexities, falx cerebri, and tentorium cerebelli), and, upon gross inspection, are rounded masses of variable consistency and vascularity with a well-defined interface between brain and tumor (4, 7, 11). In a review of 12 intracranial plasmacytomas reported as of 1982, it was noted that the preoperative diagnosis was meningioma in all five cases in which a preoperative diagnosis was given (11), as was the case in a report of MR findings of dural plasmacytoma (3). Reports of the CT findings of dural plasmacytoma note that these lesions are slightly hyperdense, rounded or lobulated extraaxial masses that homogeneously enhance after contrast administration (4, 11), analogous to the MR imaging findings in our second patient. Dural plasmacytoma also has MR imaging features similar to plasma cell granuloma, which has been reported to appear as a homogeneously enhancing dural mass compressing brain and also simulating meningioma (6). The distinction between plasmacytoma and plasma cell granuloma is made by immunohistochemical analysis; the majority of plasma cells in plasmacytoma are monoclonal in nature while plasma cell granulomas are characterized by a polyclonal distribution of plasma cells (6).

Solitary craniocerebral plasmacytoma is a radiosensitive tumor that can be treated by radiation therapy alone, surgery alone, or a combination of the two (1, 2). When increased serum immunoglobulin is present, levels can be followed as a measure of therapeutic success. Generally, patients are maintained on long-term follow-up to monitor for development of a generalized plasma cell dyscrasia, which is an uncommon event, especially in the late postoperative period (5).

In summary, solitary craniocerebral plasma-

cytoma is a tumor that can closely simulate meningioma on MR imaging studies. On the basis of our cases and a review of the radiologic literature, it appears that in many instances the diagnosis will only be reached after biopsy. The diagnosis of plasmacytoma should be borne in mind when a large extraaxial lesion is seen, because total excision may not be necessary in this radiosensitive tumor.

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