Falcotentorial plasmacytoma

Case report

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Intracranial solitary plasmacytomas are extremely rare tumors and are often misdiagnosed preoperatively. The authors report the successful treatment of a patient who harbored such a tumor involving both the falk and tentorium; this is the second case reported. A 59-year-old woman suffered from a seizure disorder due to a falcotentorial lesion, which had been identified 3 years earlier and was thought at the time to be an en plaque meningioma. Most recently, the patient presented with symptoms of increased intracranial pressure and hemiparesis. Computerized tomography and magnetic resonance imaging of her head revealed progressive growth of the tumor. The patient underwent partial resection of the tumor and chemo- and radiation therapies. Intracranial plasmacytomas must always be included in a differential diagnosis because potential complete cure can be achieved using fairly conservative treatment modalities.

KEY WORDS • atypical monoclonal plasma cell hyperplasia • central nervous system • falx cerebri • plasma cell granuloma • solitary plasmacytoma • tentorium

Plasma cell tumors may manifest as one of three pathological entities: 1) multiple myeloma; 2) solitary bone plasmacytoma; or 3) extramedullary plasmacytomas. Extramedullary plasmacytomas represent 3% of plasma cell neoplasms. Intracranial dural plasmacytomas, particularly those involving the falk and/or tentorium of the brain, are exceedingly rare, and descriptions in the world literature consist only of case reports. To the best of our knowledge, we present the ninth reported case of a plasmacytoma involving the falk or tentorium and the second involving these structures simultaneously. Despite the anatomical rarity, we report complete resolution of the tumor following partial resection and radiation and chemotherapy.

Case Report

History. This 59-year-old woman experienced seizures for the first time 3 years before the current admission. The diagnostic workup, including computerized tomography (CT) scanning obtained at that time, revealed an “en plaque” tumor involving the falk and tentorium that was strongly suggestive of a meningioma. Follow-up CT and magnetic resonance (MR) imaging performed 2 years later revealed progressive growth of the neoplasm.

The patient was admitted to our institution because she suffered from progressive headaches, left-sided weakness, and focal seizures of 1-month duration.

Examination. Neurological examination revealed bilateral papilledema, vertical and horizontal nystagmus, and left-sided hemiparesis with a motor strength of 3/5 in both extremities. Corticosteroid therapy was initiated.

Magnetic resonance imaging revealed a large mass involving both sides of the falk cerebri (Fig. 1). The mass appeared minimally hyperintense with respect to the white matter on proton density- and T2-weighted images, findings typical for a meningioma or small cell tumor. There was extensive surrounding vasogenic edema as well as intense contrast enhancement of the tumor. At the margin of the lesion, there were smooth meningeal enhancement and a “dural tail,” a feature characteristically associated with meningiomas. The process extended to involve the tentorium bilaterally. On the left side, there was a small attachment to the falcal mass, but on the right side, a discrete tentorial mass was seen.

Operation. The patient underwent a right parietal parasagittal craniotomy. The dura mater appeared unremarkable. The brain was swollen and the lesion was observed to be extraaxial, gray, soft, and hypovascular. The results
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Fig. 1. Contrast-enhanced coronal (left) and axial (right) T1-weighted MR images obtained during the patient’s initial presentation revealing a large mass involving both sides of the falk cerebri. Extensive vasogenic edema is noted bilaterally. The mass demonstrates intense contrast enhancement. A “dural tail,” which classically is associated with meningiomas, is noted (arrow), and large tentorial components are seen. The left tentorial mass appears to be connected to the falk by a thin neck, but the right mass appears separate.

Fig. 2. Photomicrograph showing the tumor in which virtually all of the cells are stained, some much more intensely than others. The eccentric position of the nuclei and the plasmacytoid configuration of the cells is accentuated by the intracytoplasmic localization of antibody to γ heavy chains. (Immunoperoxidase stain with anti-γ antibody, original magnification × 250.) There was no cellular staining for δ and µ heavy chains. Antibodies to κ light chain stained some cells and there was complete absence of staining for λ light chains.

of a frozen section were suggestive of plasmacytoma. In light of this, a partial resection of the tumor was performed. The postoperative period was uncomplicated.

Pathological Findings. Histopathological examination revealed that the tumor was composed of a relatively uniform, diffusely infiltrating population of plasmacytoid cells of variable differentiation within meningeal tissue (Figs. 2 and 3). The most differentiated cells had round, eccentrically placed nuclei with clumped, clock face–like chromatin and moderate amphophilic cytoplasm with a clear paranuclear zone. The most undifferentiated cells retained amphophilic cytoplasm; however, the nuclei were larger, more centrally placed, and vesicular, with one large or a few smaller nucleoli. Most of the cells appeared to fall between these two morphological extremes with a prevalence of less differentiated forms. Mitoses were numerous and many pyknotic nuclei were present. Immunohistological staining showed restrictive reactivity with the immunoglobulin κ light chain and γ heavy chain (immunoglobulin G, κ monoclonality). There were essentially no other cell types in the tumor apart from a few scattered lymphocytes; that is, there were no histiocytes, eosinophils, or fully mature plasma cells. The lack of heterogeneity in the cell population, combined with its high proliferative rate, cellular anaplasia, and immunophenotypical monoclonality, provided strong evidence favoring the diagnosis of a malignant plasma cellular disorder rather than a benign or reactive plasma cell granuloma. Bone marrow biopsy was performed postoperatively. There was no morphological evidence of myeloma, and immunohistological staining failed to define a clonal population of plasma cells.

Postoperative Treatment and Course. Repeated MR imaging performed 1 month later revealed shrinkage of the mass resulting from steroid therapy. An Ommaya reservoir was inserted into the right frontal horn of the lateral ventricle. Cerebrospinal fluid (CSF) and a small fragment of bone from the skull were submitted for microscopic examination; they showed no evidence of malignancy. Five courses of intrathecal methotrexate (12 mg) were administered. One-and-one-half months postoperatively, the patient received whole-brain radiation therapy with 6-MV photons at 200 cGy per day, in 20 fractions. The final 1000-cGy boost was given to a small area for a total dose of 5000 cGy. After completion of the radiation therapy, the patient was discharged with marked neurological improvement and was able to ambulate with a walker.

Follow-up MR imaging performed 6 months postoper-
Discussion

There are three variations of plasma cell tumors: multiple myeloma; solitary bone plasmacytoma; and solitary extramedullary plasmacytoma. The relationship of these tumors to each other is unclear. Extramedullary plasmacytoma as a separate clinical pathological entity represents 3% of plasma cell neoplasms. This tumor does not necessarily progress to multiple myeloma. Nevertheless, multiple myeloma should be ruled out in every patient presenting with a solitary, predominantly plasmacytic, intracranial lesion; these patients require lifelong follow-up monitoring.

The literature contains only a few descriptions of falci- cene or tentorial solitary intracranial plasmacytomas, and there is a striking preponderance of female patients (Table 1). Clinically, the tumor most often manifests during the patient’s fifth decade of life and presents with intracranial hypertension and focal neurological signs.

Plasmacytomas are seen on CT and MR images as iso- to hyperdense and iso- to hyperintense lesions, respectively, and are homogeneously enhancing after administration of contrast agent. On cerebral angiograms, they are characterized as avascular and space-occupying lesions.

Preoperative diagnosis most often favors meningioma because of the symptoms of intracranial hypertension and focal neurological signs appearing in middle-aged women with a mass based on the dura. Glioma could also be considered. In a number of reports, a preoperative diagnosis of falx meningioma or glioma was made on the basis of the presence of an M-protein peak on CSF protein electrophoresis.

In 1989, Weidenheim, et al., reviewing the spectrum of plasmacytic central nervous system lesions, introduced the terms “plasma cell granuloma” and “atypical monoclonal plasma-cell hyperplasia” as precursors of “solitary plasmacytoma.” Plasma cell granulomas are inflammatory pseudotumors with a proliferation of inflammatory cells including a mixed population of plasma cells, lymphocytes, and histiocytes admixed with granulation tissue and fibrosis. They are polyclonal in their expression of immunoglobulin light chains. Atypical monoclonal plasma cell hyperplasia contains monoclonal plasma cells and is an ill-defined entity that probably represents a type of solitary plasmacytoma. Meningiomas with a conspicuous plasma cell and lymphocyte component also must be considered. Amyloid association with intracranial plasmacytomas has frequently been noted.

Solitary intracranial plasmacytoma has a favorable prognosis (Table 1). Durable, sustained remission may occur after subtotal or radical resection combined with radiation therapy. Corticosteroid medications may have therapeutic effects. Plasma cell granuloma may be successfully treated by surgery without adjunctive therapy and may not progress to multiple myeloma during the follow-up period. Atypical monoclonal plasma cell hyperplasia may also respond very effectively to radiation therapy alone. Multiple myeloma may develop during
long-term follow-up periods in patients with intracranial solitary plasmacytomas, but it is unlikely to do so unless myelomatous changes occur in the early postoperative period.

Conclusions

Plasmacytoma should always be included in the differential diagnosis of dural-based intracranial lesions. The presence of abnormal serum or CSF immunoglobulins in the preoperative electrophoresis studies will be a strong indicator for diagnosis. Radical neurosurgical intervention is not indicated in the case of diffuse dural plasmacytoma. Stereotactic biopsy of the neoplasm followed by radio- and chemotherapy would be the treatment of choice. Complete cure of the disease may be achieved by applying these management modalities.

References


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