Transfacial excision of brainstem plasmacytoma

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Three distinct varieties of plasma cell tumors have been described: multiple myeloma (involvement of multiple osseous sites, bone marrow infiltration, and monoclonal gammopathy), plasmacytoma of bone (involving the spine or skull usually), and extramedullary plasmacytoma (EMP). EMPs, comprising only 3% of plasma cell neoplasms, are rare tumors with an annual incidence reported to be in the range of 3:100,000.[1] Initially described by Schhridde in 1905, EMPs appear to represent an autonomous lymphoproliferative disease process whose natural history is distinct from that of solitary plasmacytomas of bone marrow. EMP may occur in any organ containing reticuloendothelial cells, with approximately 90% reported to occur in the head and neck region, most commonly in the upper respiratory tract (nose and paranasal sinuses).[2] These tumors most commonly affect males (male/female ratio, 3:1) in the sixth and seventh decades. Although solitary plasmacytomas almost always eventually progress to multiple myeloma, this occurs infrequently with EMPs, which tend to be locally destructive.

Dural EMP is exceedingly rare, with only 16 cases reported to date.[3] To our knowledge, no cases of isolated dural EMP of the brainstem have been reported to date. In this brief article, we outline the presentation, treatment, and outcome of a patient with isolated dural brainstem EMP who presented to our institution.

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CASE REPORT

A 57-year-old black woman presented with a 2-week history of progressive frontal headaches and left-sided abducens palsy. Subsequent investigations included a computed tomography scan of the sinuses, which revealed mild sphenoid sinusitis. Magnetic resonance imaging of the head revealed the presence of an extradural mass centered on the brainstem with extension through the clivus (Fig 1). Results of a full systemic workup including computed tomography of the neck, chest, abdomen, and pelvis were within normal limits.

The patient was brought to the operating room for both diagnosis and treatment of a presumed skull base tumor with evidence of brainstem involvement. Bilateral nasofacial incisions along the aesthetic subunits of the nose were used to gain access to the midfacial skeleton. Miniplates were preadapted along the planned osteotomy sites in the usual fashion. Subsequently, a high Le Fort I osteotomy with a paramedian palatal split allowed a bimaxillary swing approach to be accomplished (Fig 2). The clivus was noted to be partially eroded by the tumor. Drilling this structure with a medium-sized bur under microscopic guidance provided excellent exposure to the bulk of the lesion, which was peeled off of the dura anterior to the brainstem. The lesion was completely extradural and was completely resected. The preadapted hardware was then replaced. The patient's postoperative course was quite uneventful, and her abducens palsy resolved.

Histologic examination demonstrated an almost pure population of plasma cells with the formation of cytoplasmic Russell bodies. Occasional lymphocytes and neutrophils were also present. Immunohistochemical staining was grossly positive for light chains. Subsequent serum electrophoresis, skeletal survey, lumbar puncture, and bone marrow biopsy were within normal limits.

The patient received postoperative adjuvant external beam radiotherapy to a dose of 50 Gy to the primary site. The patient remains disease free at 3 years postoperatively.

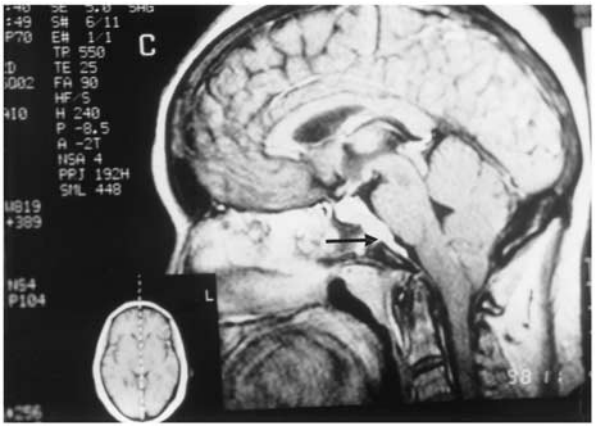

Fig 1. Sagittal magnetic resonance imaging demonstrating extradural enhancing lesion of the brainstem *(arrow)*.

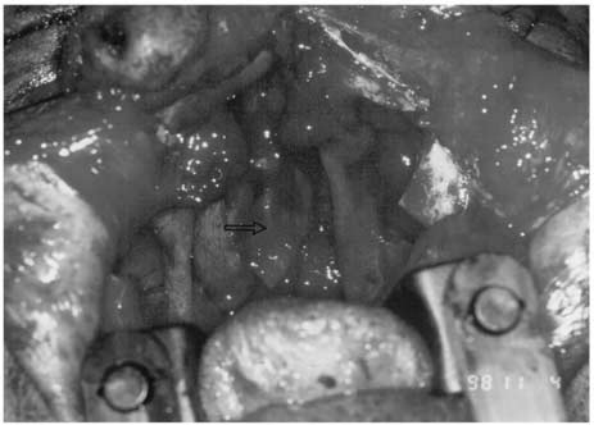

Fig 2. Exposure of the brainstem lesion *(arrow)* achieved after a bimaxillary swing approach to the skull base.

DISCUSSION

EMP is a rare disease whose natural history, if left untreated, is not clear. Surgery and external beam radiotherapy are the accepted treatment modalities for this disease process; however, due to the small number of cases reported to date, a definite survival advantage with either modality has not been demonstrated.[4] Improvement in local control has been demonstrated with complete surgical removal as well as with irradiation. A dose of at least 40 Gy appears to be associated with fewer local recurrences in the treatment of EMP.[5] Local control is likely important to achieve for 2 reasons: to decrease the local destruction caused by tumor progression, especially in the neurologically sensitive skull base area, and to decrease the likelihood of progression to multiple myeloma. Although rare progression is noted, prognosis for this small cohort of patients has been more favorable than de novo multiple myeloma.[5] There are no markers available to date that are able to predict which cases will progress to multiple myeloma and which will remain localized EMP.

In summary, complete eradication of EMP either with surgery or radiation is potentially curative. However, before initiating treatment, a full systemic workup to rule out the presence of multiple myeloma is important, because the treatment in such a circumstance is chemotherapy. Close follow-up of this patient population is important to detect early transformation to multiple myeloma or to detect the presence of locally recurrent disease.

Transfacial access to brainstem plasmacytoma facilitates safe removal.

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