Skull Base Plasmacytoma in a Patient with Light Chain Myeloma

Zeki Ustuner, M.D.,¹ Mert Basaran, M.D.,¹ Talat Kiris, M.D.,²
Bilge Bilgic, M.D.,³ Serra Sencer, M.D.,⁴ Burak Sakar, M.D.,¹
Yavuz Dizdar, M.D.,¹ Sevil Bavbek, M.D.,¹ and Haluk Onat, M.D.¹

ABSTRACT

Skull base involvement of plasmacytoma is reported in a patient with light chain myeloma. A 39-year-old man was admitted after experiencing paresthesia on the left side of the face and left arm, intermittent diplopia, and hoarseness for 2 years. Cranial magnetic resonance imaging revealed a large midline mass extending from the middle and posterior skull base into the upper two cervical vertebrae. An extramedullary plasmacytoma associated with light chain multiple myeloma was diagnosed after biopsy of the mass and laboratory investigations. The imaging findings and clinical features associated with this rare site of extramedullary plasmacytoma involvement are reported.

KEYWORDS: Skull base, multiple myeloma, light chain, plasmacytoma

Extramedullary plasmacytomas occurring as solitary (primary) tumors or secondary manifestations of multiple myeloma most often involve the upper airways and associated paranasal sinuses. These tumors represent less than 1% of all head and neck tumors.¹ Extramedullary plasmacytomas involving the skull base can produce a variety of signs and symptoms, some of which may be vague. Consequently, presentation and appropriate diagnostic investigations may be delayed.² Cranial nerve palsy may

be a significant complication of plasma-cell neoplasms involving this anatomic site.³ Treatable skull base lesions, including plasmacytomas and multiple myelomas, must be considered in the differential diagnosis of patients who demonstrate a progressive course or multiple cranial neuropathies,⁴ even those younger than 40 years old.⁵ We present a patient with light chain multiple myeloma who became symptomatic with signs and symptoms associated with plasmacytoma of the skull base.

Skull Base, volume 13, number 3, 2003. Address for correspondence and reprint requests: Zeki Ustuner, M.D., Istanbul Universitesi, Onkoloji Enstitüsü, Capa, 34390, Istanbul, Turkey. E-mail: zekiustuner@superonline.com. ¹Institute of Oncology, and Istanbul Medical Faculty, Departments of ²Neurosurgery, ³Pathology, and ⁴Radiology, Istanbul University, Istanbul, Turkey. Copyright © 2003 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2003,13,03,167,172, ftx,en;sbs00346x.

ILLUSTRATIVE CASE

A 39-year-old man was admitted with a 2-year history of progressive paresthesia of the left side of the face and left arm, intermittent diplopia, dysphagia, and hoarseness. His physical examination was unremarkable. His neurological examination revealed ninth and tenth cranial nerve palsy on the right. Routine laboratory evaluation was normal except for mild anemia (hemoglobin, 12.8 gr/dL) and an elevated erythrocyte sedimentation rate (31 mm/hr). Serum calcium, alkaline phosphatase levels, and renal function tests were normal.

Cranial magnetic resonance imaging (MRI) disclosed a large midline mass extending from the ethmoid and sphenoid sinuses into the clivus and petroclival junctions. The tumor also invaded the atlantoaxial joint and odontoid process (Fig. 1). A presumptive diagnosis of hematological malignancy, chordoma, or invasive adenoma was made.

Transnasal trans-sphenoidal biopsy of the tumor was performed to obtain a definitive diagno-

sis. Intraoperative imprint smear and paraffin sections of the biopsy specimen showed extensive infiltration of plasma cells (Fig. 2), which stained positive for CD 38 and kappa light chain, immunohistochemically. The lambda light chain stain was negative. Bone marrow biopsy showed almost diffuse neoplastic plasma cell infiltration. Serum immunoglobulin (Ig) levels were as follows: IgG, 453 mg/dL; IgA, 35.5 mg/dL; and IgM, 45.5 mg/dL. Beta-2 microglobulin level was 2.6 mg/L. Urine immunoelectrophoresis showed increased (8 gr/day) light chain excretion. Light chain multiple myeloma stage II A according to the Durie-Salmon staging system was diagnosed.

The patient received radiation (52 Gy) for the skull base mass followed by systemic vincristine, adriamycin, and dexamethasone (VAD) combination chemotherapy. Cranial MRI performed after the patient completed four cycles of chemotherapy showed a partial response with some regression of the tumor (Fig. 3). A follow-up examination showed no residual neurological deficit and the patient was

Figure 1 (A) T1-weighted sagittal gadolinium-enhanced MRI shows homogeneously enhancing soft-tissue mass in the ethmoid and sphenoid sinuses and clivus and extending into upper two cervical vertebrae. (B) T2-weighted coronal MRI shows that the solid tumor (intermediate signal) involves the sphenoid sinus and sellar region.

Figure 2 Neoplastic plasma cell infiltration (hematoxylin and eosin 500 $\times$).

symptom free. The patient is expected to receive high-dose melphalan and an autologous stem cell transplant.

DISCUSSION

Multiple myeloma has been reported in only 2% of patients under 40 years of age.⁶ Our patient was in this rare age range for disease presentation. Skull base lesions are usually asymptomatic, but larger tumors can cause symptoms. Multiple myeloma rarely involves the skull base. Cranial nerves II, III, IV, and VI are most often involved, but large lesions can affect other cranial nerves.^{3-5,7} Plasma cell neoplasms should be considered in the differential diagnosis of skull base tumors associated with cranial nerve palsy. Our patient had ninth and tenth cranial nerve palsy due to the extensive involvement of the skull base by the lesion. His intermittent diplopia was probably related to increased intracranial pressure.⁸ He did not exhibit the common initial findings of myeloma such as bone pain, anemia, renal insufficiency, and hypercalcemia.

Extramedullary plasmacytoma can be a harbinger of multiple myeloma⁶ in 20 to 30% of the patients¹ and may develop several years after disease onset.^{8,9} The risk for a plasmacytoma of the skull base to progress to multiple myeloma over the course of long-term follow-up is high.¹⁰ The longstanding

Figure 3 After radiotherapy, a T2-weighted sagittal MRI shows regression of the tumor and some treatment-related tumor necrosis.

symptoms and relatively insidious course of the disease in our patient implies that the extramedullary plasmacytoma had progressed to myeloma.

Plasmacytomas are composed of a proliferation of abnormal plasma cells that exhibit monoclonal intracellular immunoglobulins with immunological staining techniques. Our patient was diagnosed after cells expressing monoclonal kappa light chain were detected in the specimen obtained from the skull base mass biopsy as well as in the urine. Atypical plasma cells were also present in the bone marrow biopsy.

Plasma cell neoplasms are highly radiosensitive. Local irradiation is the primary mode of treatment for extramedullary plasmacytomas, occasionally followed by surgical resection of the residual tumor.⁷ When multiple myeloma is diagnosed, the treatment of choice is systemic combination chemotherapy followed by high-dose melphalan and autologous stem cell transplant after palliative local treatment.¹⁰

As presented in this report, the clinical and radiological features of the skull base plasmacytoma, a rare location in an unexpectedly young patient, may aid our understanding of the disease.

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Commentary

Ustuner and colleagues report a 39-year-old male who presented with left face and arm paresthesia, intermittent diplopia, dysphagia, and hoarseness, which had progressed for 2 years. After a trans-sphenoidal biopsy was performed, he was diagnosed with a plasma cell tumor involving the clivus and nasopharynx. Finally, multiple myeloma was diagnosed after a bone marrow biopsy. The patient was treated with radiotherapy for the skull base disease and with chemotherapy for the systemic component of the pathology. After he underwent radiotherapy, MRI showed a dramatic response at the skull base and the patient's neurological deficits resolved. Unfortunately, his long-term oncological outcome was not provided.

Plasma cell neoplasms are divided into multiple myelomas, solitary plasmocytomas of bone, and extramedullary plasmocytomas. Whereas multiple myelomas represent systemic disease without the potential for cure, solitary plasmocytomas of bone and extramedullary plasmocytomas represent local forms of plasma cell neoplasms. Their prognosis depends on their likelihood of progressing to multiple myeloma. The potential for malignant systemic

progression is higher for solitary plasmocytomas of bone than for extramedullary plasmocytomas. Local plasma cell neoplasms can involve skull base structures. They may occur at the clivus or petrous apex where they represent a solitary plasmocytoma of bone, or they may originate within the submucosa of the sinu-nasal and nasopharyngeal tracts where they represent an extramedullary plasmocytoma. In the present case, the lesion involving the clivus and nasopharynx was part of disseminated disease, and multiple myeloma was diagnosed.

Before considering major skull base resection, the authors appropriately obtained a tissue bi-

opsy. As this case documents, excellent clinical and radiological results can be obtained with radiation of plasma cell tumors of the skull base. These tumors are known to be radiosensitive. Therefore, surgical resection should be reserved for salvaging unresponsive local plasma cell tumors involving the skull base. Chemotherapy is the treatment of choice for systemic disease. This case again highlights the importance of obtaining a biopsy of skull base tumors to determine the appropriate treatment.

Iman Feiz-Erfan, M.D.¹

Randall W. Porter, M.D.¹