SOLITARY EXTRAMEDULLARY PLASMACYTOMA OF THE ANTERIOR SKULL BASE PRESENTING WITH SEIZURES: A CASE REPORT

Anish Parekh MD, Jagdish Dhingra MD FACS, Elie Rebeiz MD FACS

Department of Otolaryngology – Head and Neck Surgery, Tufts Medical Center, Tufts University School of Medicine, Boston MA

Introduction

Extramedullary plasmacytomas are rare malignant monoclonal proliferations of plasma cells that present in the upper respiratory tract mucosa. In the head and neck, these tumors often cause nasal or pharyngeal symptoms. Nasal tumors commonly cause obstruction and epistaxis. We report a unique presentation of a 48-year-old female with seizures who was found to have a nasal plasmacytoma. We also discuss clinical presentation, diagnosis, and treatment.

Case Report

A 48-year-old female presented with new-onset seizures. In the Emergency Department, a CT scan revealed soft tissue mass lesion involving the anterior nasal cavity with osseous destruction of the septum, maxillary processes, and extension into the frontal bulla (Figure 1). The mass eroded the cribiform plate and extended intracranially to the olfactory fossa. The patient was referred to Otolaryngology for further management. Office-based nasal endoscopy was normal. Biopsy was performed under general anesthesia which revealed a diffuse infiltrate of atypical neoplastic plasma cells (Figure 2). Serum, cerebrospinal fluid, and bone marrow samples did not show any abnormality. The patient was treated with radiotherapy of 4,140 cGy over one month. She is doing well clinically and underwent 6-month follow-up MRI (Figure 3).

Discussion

Clinical Presentation
The presenting signs and symptoms of extramedullary plasmacytomas are often nonspecific but related to the mass effect and location of the lesion. As many as 80% present in the upper respiratory tract and 90% are solitary. Often, they present with nasal obstruction, epistaxis, facial asymmetry, or sensory deficit. The male to female ratios is 4:1 and 75% present during the fifth to seventh decade of life.

Diagnosis
Differential diagnosis include benign tumors such as osteoma, hemangioma, papilloma, and angiofibroma. Malignant tumors found in the nose are squamous cell carcinoma, nasopharyngeal carcinoma, adenocarcinoma and melanoma. Often times, imaging is obtained to better characterize these lesions. CT and MRI are common modalities but do not distinguish plasmacytomas from other soft tissue lesions.

Obtaining adequate tissue biopsy is necessary for accurate diagnosis of plasmacytoma. They are characterized by a proliferation of atypical plasma cells of varying maturity. Nuclei are round, eccentric and have a “clock-face” nuclear chromatin pattern. Immunohistochemistry confirms monoclonal and neoplastic nature of these tumors.

Multiple myeloma must be excluded prior to arriving to the diagnosis of extramedullary plasmacytoma.

Treatment
The mainstay of treatment for extramedullary plasmacytoma of the head and neck is radiation therapy and surgical excision.

Plasmacytomas are radiosensitive and respond well to total dose of 4500 to 6000 cGy. This has shown to result in 7% locoregional control with 5-year survival of 82%.

Surgery has shown to have a similar success rate when disease is limited and amenable to resection. This includes small, pedunculated nasal lesions, and parotid, thyroid or nodal disease.

Long-term Management
Patients with plasmacytomas require annual follow-up with a head and neck surgeon and medical oncologist. Imaging should be performed to detect recurrence and serum and urine studies to detect conversion to multiple myeloma.

Conclusion

Plasmacytomas are a rare neoplasm of the head and neck but should be included in the differential diagnosis of nasal septum mass. Treatment of these lesions includes radiotherapy with or without surgical excision.

References