

CASE REPORT

Sino-orbital Plasmacytoma: A Rare Entity

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ABSTRACT

Tumors of plasma cell origin are termed as plasmacytomas. They can be primary or secondary. Primary tumors are invasive locally and do not metastatize. Involvement of the eye and orbit is very rare, and only few cases have been reported around the globe. We outline the case of a 48-year-old male, who presented with primary plasmacytoma of the maxilla and orbit, and how it was managed using the combined approach technique.

Keywords: Multiple myeloma, Plasmacytoma, Sino-orbital neoplasms.

How to cite this article: Subash A, Gupta AK, Singh A, Bansal S. Sino-orbital Plasmacytoma: A Rare Entity. Clin Rhinol An Int J 2017;10(3):163-165.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Tumors of plasma cell origin are termed as plasmacytomas. They can be primary or secondary. Primary tumors are invasive locally and do not metastatize, unlike secondary plasmacytomas that are manifestations of multiple myeloma and are invariably metastatic. Involvement of the eye and orbit is very rare and only few cases have been reported around the globe.¹ We present the case of a 48-year-old male who presented with primary plasmacytoma of the maxilla and orbit.

CASE REPORT

A 48-year-old male solidier presented with progressive decrease in mouth opening and bleeding from the nose for the past 1 year. Patient was initially investigated at other hospitals and was treated symptomatically. A diagnostic nasal endoscopy revealed a friable mass filling the right nasal cavity. At presentation, patient also

had blurring of vision in the right eye and diplopia. A contrast-enhanced computed tomography (CT) revealed heterogeneous soft tissue filling the right nasal cavity, right paranasal sinuses, and extending to the infratemporal fossa and right orbit (Figs 1 and 2). A biopsy from the tumor came out to be plasmacytoma. This diagnostic surprise warranted further workup to confirm if it was a primary or secondary plasmacytoma. A skeletal profile was ordered (Fig. 3); a bone marrow study was performed and was unremarkable. After confirming a diagnosis of primary plasmacytoma, a combined approach of subtotal maxillectomy with endoscopic excision of orbital tumor was decided on. This approach helped better visualization of the disease, allowing complete removal with conservation of orbital contents and thus preserving



Fig. 1: Contrast-enhanced CT of paranasal sinus (coronal)

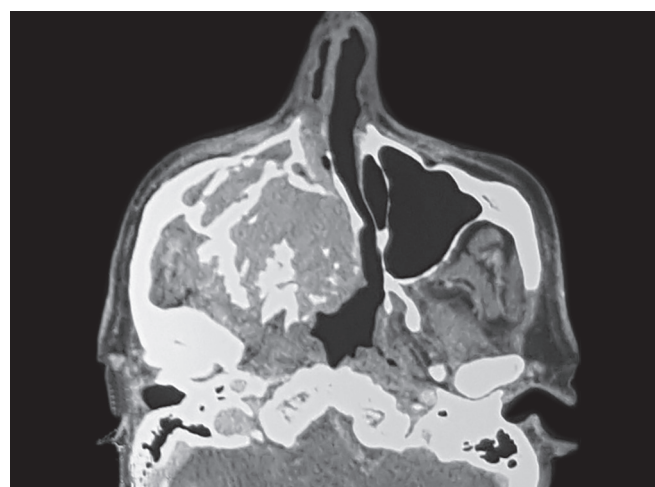
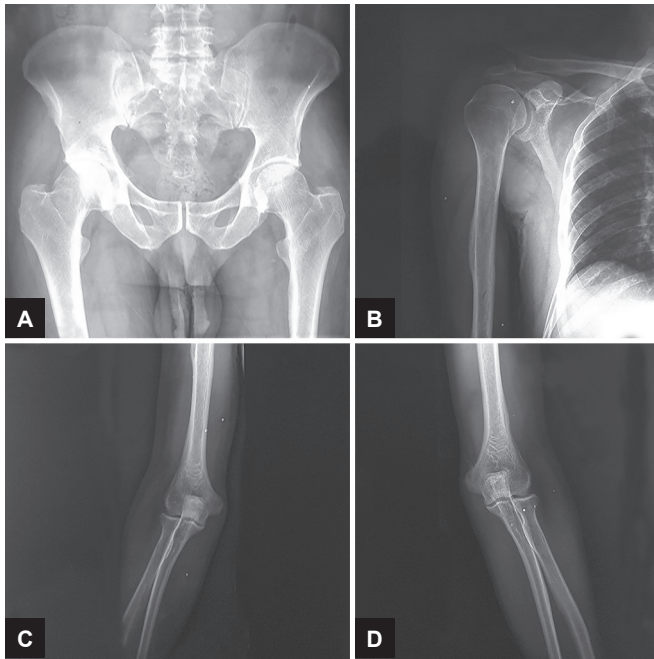


Fig. 2: Contrast-enhanced CT of paranasal sinus (axial)

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Figs 3A to D: Skeletal profile

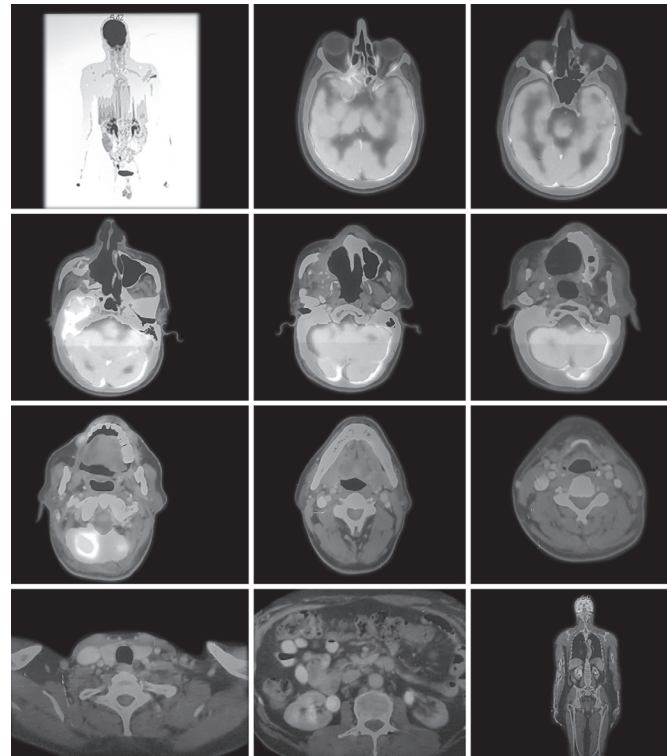


Fig. 4: Whole body PET-CT

the vision and cosmesis. Despite tumor-free resection margins, patient was given 46 gray (Gy) in 23 fractions of radical radiotherapy and four cycles of chemotherapy over 4 weeks. A whole body positron emission tomography (PET) CT was done to rule out distant metastasis (Fig. 4). Patient has been symptom-free and is on regular follow-up with nasal endoscopy for more than 2 years now (Figs 5 and 6).

DISCUSSION

B-lymphocytes can transform to immunoglobulin producing cells and are termed plasma cells. Tumors of plasma cell origins are termed as plasmacytomas. The criteria for the diagnosis of primary plasmacytoma include a negative nodal status, a skeletal survey, a bone marrow

study, and appropriated whole body imaging.¹ Treatment of plasmacytoma includes external beam radiotherapy or local excision.²⁻⁴ The mean survival is over 8 years.^{3,5}

Orbital involvement in multiple myeloma is uncommon. Bonavolontà et al,⁶ who studied orbital tumors extensively, have reported that orbital plasmacytoma has a prevalence of 3% and accounts for <1% of total orbital lesions. In our case, the primary site of origin was maxilla and the tumor had extended to orbit, which is extremely rare.

Using a combined approach, organ preservation can be better performed. In our case, use of endoscopes provided better visualization and helped us in complete extirpation of tumor with preservation of orbital contents

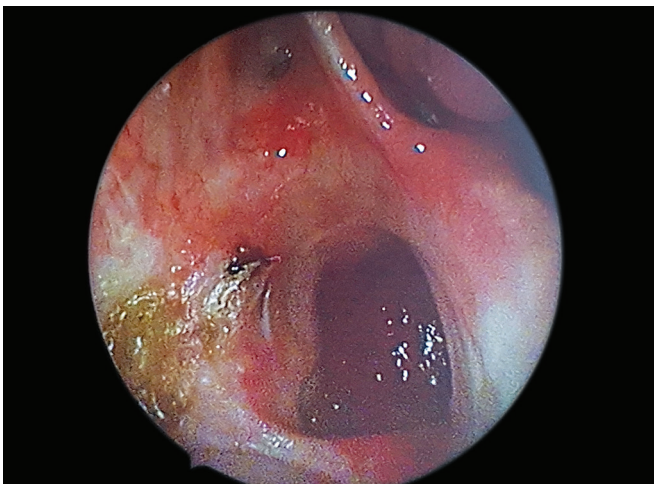


Fig. 5: Postoperative endoscopy: well-healed cavity (2 years)

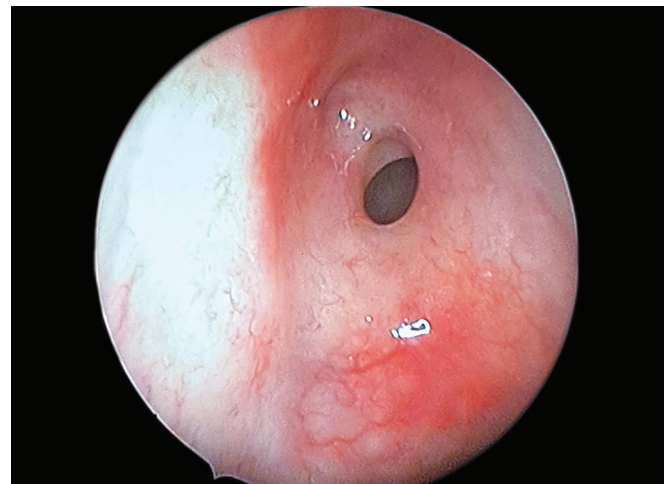


Fig. 6: Postoperative endoscopy: sphenoid ostium (2 years)



Fig. 7: Patient after 2 years of surgery

and orbital sling. This prevented postoperative diplopia and maintained the esthetics of face (Fig. 7).

CONCLUSION

Sino-orbital plasmacytoma is a rare tumor. Appropriate treatment includes surgical extirpation followed by

concurrent chemoradiotherapy. If treated appropriately, the disease has a good prognosis.

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