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CASE REPORT

Orbital extramedullary plasmacytoma masquerading as lacrimal gland tumor: A case report

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Abstract

A 66-year-old man presented with worsening left upper eyelid swelling and redness for a week as well as new-onset diplopia and weight loss. Unilateral proptosis and hypoglobus were noted, and extraocular movements were limited. Imaging showed an extraconal mass with globe displacement and bone destruction. The mass was first suspected lacrinal gland tumor. However, when biopsy results returned normal, a subperiosteal origin was determined following a radiology consult. An urgent biopsy was performed, with notable intraoperative bleeding from the lesion and eroded infiltrated bone. Pathology results were consistent with extramedullary plasmacytoma, and hematological workup was diagnostic of multiple myeloma. The patient was started on systemic chemotherapy. The disease course was complicated by a pathologic humeral fracture, which was treated after patient stabilization. After two cycles of systemic chemotherapy, there was a significant improvement in pain, proptosis, and extraocular movements.

Introduction

Multiple myeloma (MM) is an insidious hematological malignancy characterized by monoclonal proliferation of plasma cells in the bone marrow. Extramedullary plasmacytoma is a rare manifestation of this systemic disease. Most extramedullary lesions occur in the upper respiratory tract. Orbital involvement, although uncommon, poses great risks to both vision and life.[1]

There is no consensus regarding the best treatment approach to orbital plasmacytoma, and randomized studies are nearly impossible to conduct due to case paucity. Most specialists agree that primary orbital plasmacytoma may be treated with radiotherapy, while secondary plasmacytoma requires systemic chemotherapy.[2]

Case Report

A 66-year-old retired policeman with diabetes mellitus type 2, an old shrapnel injury, and no pertinent ophthalmic history presented in the ophthalmology emergency room (ER). He complained of worsening left upper eyelid swelling and redness for a week. The patient also complained of binocular diplopia, both horizontal and vertical, worsening on the left gaze and upgaze. He denied fever, visual loss, or pain on extraocular motions. The patient has noted a left-sided headache for 3 months with no recent change in quality or severity. He denied tremor, heat intolerance, or mood changes but admitted to weight loss of 11 kg for the past 6 months, which he attributed to diabetes treatment and tight glucose control. A week previously he was prescribed cefalexin by his family physician, which provided only partial relief, and was consequently referred to urgent care.

On physical examination, the patient had normal vital signs. Visual acuity was20/20 OU. On external examination, left-sided proptosis and hypoglobus were noted; no orbital mass could be palpated. Extraocular movements in the left eye were restricted in upgaze and abduction [Figure 1]. Eyelid swelling and redness were noted, as well as mild conjunctival injection. Cobalt blue filter showed superficial punctate keratopathy. The rest of the examination was normal.

Computed tomography scan showed an superotempotal extraconal mass in the left orbit with inferomedial globe displacement. Bone destruction was noted, encroaching the left maxillary and frontal sinuses and possibly the cranium [Figure 2].

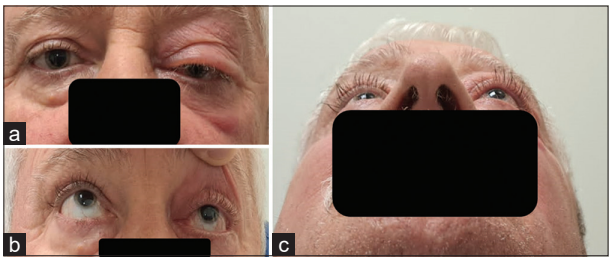

Figure 1: External photograph of the patient on presentation, showing upper eyelid edema (a), upgaze restriction (b), and left-sided proptosis (c)

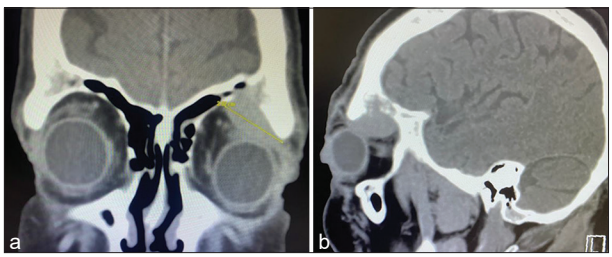

Figure 2: Computed tomography, coronal (a), and sagittal (b) views, showing left-sided upper orbital mass with notable globe displacement and bone destruction

Lacrimnal gland tumor with secondary orbital cellulitis was suspected. An urgent biopsy of the lacrimal gland was scheduled and the patient was discharged with oral antibiotics.

Within few days, the patient reported significant clinical improvement. Eyelid redness and swelling subsided. However, proptosis was still evident. Pathology results were consistent with normal lacrimal gland tissue with few foci of mild chronic inflammation and no malignant cells. A sampling error was presumed, and the patient was sent for repeat tomography. The superotemporal orbital mass was still present, with minimal change from the previous scan. Upon further inspection and radiology consult, the mass was determined to be of subperiosteal origin, and a displaced yet normal-appearing lacrimal gland was identified in the lower temporal orbit [Figure 3].

An urgent orbital biopsy from the subperiosteal mass was performed. Intraoperatively, there was evident bleeding from the lesion and eroded infiltrated bone [Figure 4]. Pathology results were consistent with extramedullary plasmacytoma with Lambda light chain restriction. The patient was admitted to hematology, has undergone systemic workup, and was diagnosed with MM. He was started on systemic chemotherapy with daratumumab, bortezomib, cyclophosphamide, and dexamethasone (DARA-VCD).

Shortly afterward, the patient presented in orthopedic ER complaining of sharp pain in the tight arm after opening a pickle jar. Humeral fracture was diagnosed and surgical reduction was offered. However, orthopedic surgeons were advised to wait until stabilization of the systemic hematological disorder before proceeding to surgery. Positron emission tomography–computed tomography showed a focus of increased uptake in the

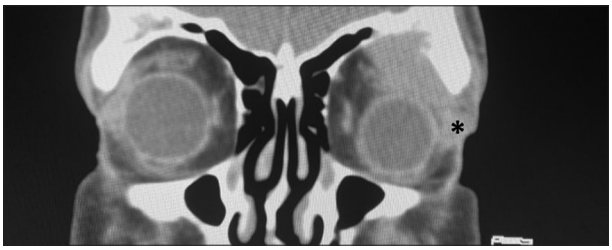

Figure 3: Repeat computed tomography, coronal view, showing the left-sided upper orbital mass and an inferiorly displaced, normal-appearing lacrimal gland (asterisk)

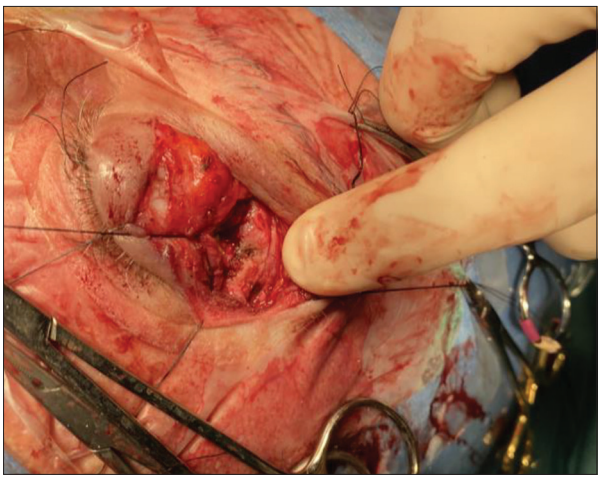

Figure 4: Intraoperative photography of the left orbital lesion, showing subperiosteal infiltration

humerus overlying the fracture line, confirming the diagnosis of a pathological fracture [Figure 5]. After stabilization, the fracture was successfully operated.

After two cycles of systemic chemotherapy and supportive treatment with granulocyte colony-stimulating factor, the patient reported relief from left-sided headaches. There was significant improvement in proptosis and extraocular movements were full.

Discussion

Orbital plasmacytoma accounts for <1% of total orbital tumors.[3] Unilateral proptosis is the most common presenting symptom. The primary manifestation of MM in this form is especially rare, yet several cases of orbital plasmacytoma have been recently described. Permaisuri reported two patients in their forties presenting with a rapid onset of unilateral proptosis with a palpable mass in the superior orbit. Ocular motility was restricted, and CT showed significant bone destruction. Systemic workup showed MM in one patient, whereas the other died before systemic workup could be completed. Both patients died <2 months after diagnosis, underscoring a very poor prognosis.[3]

Matos reported a 71-year-old man who presented in the ER with several weeks of unilateral progressive proptosis. He also

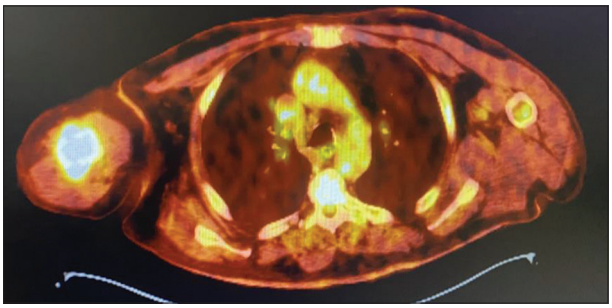

Figure 5: Positron emission tomography–computed tomography, showing a focus of increased uptake in the humerus overlying the pathological fracture line

complained of headache, epistaxis, and visual loss. CT showed bone destruction and sinusitis with suspected periorbital abscess. The patient was admitted to the intensive care unit and started on systemic antimicrobial and antifungal therapy. Systemic workup was consistent with severe MM. Therapy was proposed but he was deceased a few days after admission.[4]

Silverman reported a 60-year-old man who presented to the ER with intermittent diplopia and unilateral pain above the eye for 2 weeks. The patient also reported ipsilateral progressive ptosis for 1 week. Mydriasis and poor reactivity to light were notable on examination, as well as ptosis and mild exotropia. Extraocular movements showed limitation on supraduction and infraduction. A clinical diagnosis of cavernous sinus syndrome was made. CT demonstrated a clival mass with cavernous sinus and sphenoid sinus extension and associated bone erosion. Systemic workup was diagnostic of MM with high-risk cytogenetics and multiple lytic lesions, including several vertebral bodies. The patient was started on pulse steroids and planned to start systemic therapy, which was delayed due to COVID-19 infection. Bortezomib was started but discontinued due to hypercalcemia and altered mental status. Imaging showed worsening lesions of the thoracic spine with cord compression. The decision was then made to stop chemotherapy and pursue palliative care measures. The patient was deceased shortly thereafter, 5 months after diagnosis.[5]

Mani reported a 50-year-old woman who presented with 1 month of unilateral proptosis. Restriction of all extraocular movements was noted on examination, as well as elevated intraocular pressure. CT showed a superolateral orbital mass with lytic lesions in the calvarium. Systemic workup was diagnostic for MM. The patient was started on systemic chemotherapy with bortezomib, dexamethasone, and lenalidomide (VRD). Significant improvement in proptosis and extraocular movements was noted on follow-up.[6]

Conclusion

The clinical presentation of orbital plasmacytoma is highly versatile and clinically challenging. The most common presentation is subacute progressive unilateral proptosis in a middle-aged man. Variable amounts of pain, diplopia, and visual loss might be present. Additional signs, including palpable orbital masses, epistaxis, and cranial nerve palsies, may also be found. Orbital plasmacytoma may be the first manifestation of systemic disease. A high index of suspicion and a multidisciplinary approach is needed to diagnose and manage this potentially devastating, although rare, condition.

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