Masquerading solitary plasmacytoma; an eyelid lump in disguise

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Abstract:
Solitary plasmacytoma is a rare orbital lesion, most commonly appearing in patients with multiple myeloma. We report a case of a 75-year-old woman who presented with a left upper eyelid lesion, initially misdiagnosed and treated as a chalazion. Histopathological testing revealed plasmacytoma originating from the frontal sinus. This case demonstrates a rare presentation for this malignancy.

Keywords:
Chalazion, multiple myeloma, orbital tumor, plasmacytoma, radiation

Introduction
Plasma cell malignancy can be primary or secondary.[1] Although rare, primary orbital plasmacytomas can originate from bone or from soft tissue. Secondary plasmacytomas are manifestations of multiple myeloma, which are more common,[2] and have a tendency to be more aggressive. Plasmacytomas are a neoplasia of abnormal plasma cells producing monoclonal immunoglobulin chains. When solitary, they are called solitary plasmacytoma of the bone. In the orbit, these tumors most commonly originate from the bone.[3] They can also, more rarely, be found in soft tissues, called extramedullary plasmacytomas; these cases carry a more favorable prognosis. We present a case of a woman with a left upper lid mass treated as a chalazion and after biopsy was determined to be a solitary extramedullary plasmacytoma.

Case Report
A 75-year-old woman presented with a left upper lid mass for a 2-week duration. She was seen in a local clinic, diagnosed as a chalazion, and was treated with neomycin/polymyxin/dexamethasone ointment and hot compresses. She was sent to our department after exacerbation of the symptoms and growth of the mass. On review of symptoms, the patient had no other significant medical history, was not immunocompromised and took no regular medications. On examination, her visual acuity was 20/20 OU; the physical examination was significant for large immobile nonfluctuating supra-tarsal mass with red and erythema surrounding the mass [Figure 1]. There were no orbital signs observed on examination; there was full ocular motility in both eyes, no proptosis and color vision was intact. In addition, the dilated funduscopic examination was normal. Computed tomography (CT) scan of the orbits was performed, revealing a large mass extending from the frontal sinus through the soft tissue of the eyelid [Figure 2a and b]. There was extensive bony erosion with no inter-cranial extension. Initial laboratory investigations were performed, including a complete blood count, chemistry, and coagulation studies, all within the normal limits. A biopsy was performed with a combined approach; both ears, nose, and throat (ENT) team, and oculoplastic teams were present [Figure 3]. The ENT team
approached endoscopically using a navigation system to properly locate the lesion within the area of the frontal sinus. The oculoplastic team approached through a lid crease incision, and the material was sent to biopsy. Biopsy revealed an inflammatory infiltrate with many plasma cells and granulation tissue with a lymphocytic inflammatory infiltrate with plasma cells and neutrophils. There was an abnormal ratio of kappa to lambda and a high level of CD: 138 expression. Polymerase chain reaction was also performed and a monoclonal B-cell proliferation was seen. The histopathological diagnosis was plasmacytoma. The hematology department was consulted, and a systemic workup was performed. On further review of systems, there was no weight loss, fevers, night sweats, or malaise. Further blood work was performed, including; liver function tests and immunological studies, which included immunoglobulin levels, which were normal, and immunofixation studies, which revealed polyclonal expression. The free light chains were isolated and the ratio of kappa: Lambda was two (free $\kappa = 25.8$, free $\lambda = 13$). A positron emission tomography CT scan was also performed. There was evidence of residual uptake in the frontal sinus, with no other loci of systemic uptake. A diagnosis of solitary extramedullary plasmacytoma was made with no systemic evidence of multiple myeloma. The patient was treated with radiation directed to the orbit, with a total dosage of 50 gray, divided into 25 fractions of 2 gray. At 2 weeks postradiation, she had evidence of superficial punctate keratitis. At 4 weeks postradiation treatment, there was improvement in the conjunctival hyperemia, edema, and corneal signs. During her last ENT follow-up, 1 year since diagnosis, a fiber optic examination was performed and the sinus was open and clean. At the last hematology follow-up of 22 months post diagnosis, the blood work was stable. Last PET CT scan revealed no evidence of systemic disease.

**Discussion**

A presentation of plasmacytoma in the orbit is rare. Often the plasmacytoma is subdivided into two categories depending on the origin; plasmacytoma arising from the bone, or plasmacytoma arising from the soft tissue.$^3$ Plasmacytoma of the orbit is rare, and in most presentations of orbital plasmacytoma, multiple myeloma is present.$^4$ A recent review described plasmacytoma in terms of the orbital anatomical location of origin.$^2$ In this review, all orbital plasmacytomas were found exclusively in patients with multiple myeloma, which was diagnosed before or immediately after orbital plasmacytoma. Our case is solitary, extramedullary, without evidence of systemic disease almost 2 years after the first presentation. According to the aforementioned review, our patient belongs to the group in whom the mass clearly originated from the sinuses and only secondarily involved the orbit.$^2$ They found that these plasmacytomas were usually very large and involved
the inferior or medial orbit. Our patient had a lesion involving the superior orbit, originating from the frontal sinus. Since most plasmacytomas are not truly solitary; often the orbital component is the presenting sign for systemic disease.\[5\]

**Conclusion**

Orbital Manifestations of solitary plasmacytoma are rare. It is imperative to continue close follow-up in cases of solitary plasmacytoma with negative systemic workup because patients can develop multiple myeloma even years after original presentation of plasmacytoma.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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