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Conjunctival plasmacytoma

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Abstract

BACKGROUND: Plasmacytomas are plasma cell tumors that may be a primary or secondary tumor focus, the latter of which are associated with multiple myeloma. We present a rare case of a solitary extramedullary plasmacytoma involving the conjunctiva.

CASE REPORT: A 33-year-old white man presented with the initial complaint of redness in both eyes, more in his right than left eye. A vascularized conjunctival lesion was noted in his right eye. The patient underwent excisional biopsy, which found a conjunctival plasmacytoma.

CONCLUSION: Although plasmacytomas of the eye and orbit are rare, it is important to be familiar with these tumors that may be associated with multiple myeloma. Definitive diagnosis is made by biopsy and histopathologic examination of the tissue. Plasmacytomas may be treated with external beam radiation, local excision, or radiotherapy after surgical excision. Long-term follow-up, including periodic systemic evaluation, is required to establish that orbital involvement is not an early manifestation of multiple myeloma.

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Plasmacytomas were first described by Dalrymple and Bence Jones in 1846.[1] Plasmacytomas are collections of plasma cells that have become modified to produce large amounts of immunoglobulin and are classified as a type of B-cell non-Hodgkin lymphoma.[2] Although the etiology of plasmacytoma is still unknown, a link to a hepatitis C viral infection has been suggested[3]; high hepatitis C virus sero-prevalence has been detected in patients affected by B-cell non-Hodgkin's lymphoma,[4] and active hepatitis C virus, including replication, has been identified in the bone marrow of patients with multiple myeloma.[5]

Plasmacytomas may be a primary or secondary tumor focus, the distinction of which is based on the presence of systemic disease. The most malignant plasma cell neoplasm is multiple myeloma, which may manifest as a secondary plasmacytoma and is aggressive, metastatic, and represents 1% of all cancers[6] and 10% of the hematologic malignancies.[7] Plasmacytomas typically affect patients between the ages of 50 to 80 years.[3] Systemic complications, such as localized bone symptoms, alterations in blood calcium levels, and kidney damage, may be present not only in multiple myeloma but also in patients with a solitary plasmacytoma.[3] Specifically, patients may report bone pain caused by bone lysis and/or bone fracture from altered osteoclast/osteoblast activity leading to calcium mobilization from affected bone; elevated calcium levels lead to tubular damage and subsequent kidney damage.[3]

Primary plasmacytomas may be divided into medullary, referring to bone lesions, or extramedullary, referring to soft tissue lesions.[6] Known as *solitary extramedullary plasmacytomas* (SEMP), primary lesions represent 3% of plasma cell neoplasms,[8] are locally invasive, and do not metastasize often. Extramedullary plasmacytomas occur between the fourth and seventh decades of life, with the majority of cases involving the upper respiratory tract[9]

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but may rarely manifest in and around the eye. Extramedullary plasmacytomas may precede multiple myeloma.[8]

In a series reviewing conjunctival tumors, 128 lymphoid tumors of the conjunctiva were identified, representing the fifth most common conjunctival tumor, and accounting for 8% of all conjunctival tumors.[10] Plasmacytomas accounted for 1% of the lymphoid tumors; other lymphoid tumors included were malignant lymphoma (77%) and benign reactive lymphoid hyperplasia (22%).[10] Lymphoid tumors of the conjunctiva were most commonly found in the fornix (37%), followed by the extralimbal bulbar conjunctiva (33%), and lastly the limbus (8%); lymphoid tumors were most likely located diffusely (34%), followed by superiorly (24%), inferiorly (20%), nasally (17%), and temporally (5%).[10] The mean age of detection for malignant lymphoid, along with epithelial, lipomatous, secondary, and leukemic tumors, was 60 years, and most patients in the survey were classified as white (89%); 37% of lymphoid tumors presented bilaterally in this series.[10] In a series focusing on conjunctival lymphoid tumors at the same institution, systemic lymphoma was detected in 47% of the 117 patients with bilateral disease and 17% with unilateral disease.[11] We report on a case of a conjunctival solitary extramedullary plasmacytoma.

Case report

A 33-year-old white man presented with a chief concern of constant redness in both eyes, more in his right than left eye. The patient's ocular and medical histories were negative, and he denied any taking medications or having allergies. His family ocular history was positive for glaucoma in his paternal grandfather. External evaluation found the pupils were equally round and reactive to light without any afferent pupillary defect, extraocular motilities were unrestricted in all gazes, and confrontation visual fields were full to finger counting in both eyes.

His uncorrected visual acuities were 20/20− in each eye, with a manifest refraction of +0.50 in both eyes yielding a best-corrected visual acuity of 20/20 in each eye. Intraocular pressures (IOPs) were 20 mmHg in each eye predilation and 23 mmHg in each eye postdilation with Goldmann applanation tonometry (GAT). Anterior-segment evaluation with slit lamp examination found clogged meibomian glands and inflamed nasal pterygia (1.8 mm across the limbus in his right eye and 2.4 mm in his left eye). A mildly elevated, mobile, faint pink, vascularized, 4.5-mm by 4.5-mm lobular temporal bulbar conjunctival lesion in his right eye was also noted. His dilated fundus examination findings were unremarkable in both eyes with a cup-to-disc ratio of 0.40 with healthy rims in both eyes.

The patient had inflamed pterygia and meibomitis diagnosed in both eyes and a suspicious bulbar conjunctival lesion diagnosed in his right eye. He was started on ketorolac tromethamine 0.5% ophthalmic solution 4 times a day in both eyes for 7 days, lid hygiene was initiated, and he was asked to return to clinic in 3 months for monitoring. Based on his postdilated IOP spike, he was also considered a glaucoma suspect, and an IOP check and baseline threshold visual field examination was planned at his follow-up visit.

The patient failed to return at the recommended follow-up time and instead returned to the clinic a year and a half later reporting continuing redness in both eyes for the prior 2.5 years; he requested eye drops to decrease redness. He reported that the ketorolac tromethamine 0.5% ophthalmic solution previously prescribed did not help, and occasional use of naphazoline hydrochloride 0.025% and pheniramine maleate 0.3% ophthalmic solution helped only temporarily. He also commented that the vascular lesion in his right eye had been stable for the prior 1 to 2 years. Slit lamp biomicroscopy found a mild follicular reaction in the inferior palpebral conjunctiva of both eyes, stable nasal pterygia in both eyes, and the suspicious bulbar conjunctival lesion in his right eye. IOPs were 16 mmHg in both eyes with GAT. He deferred dilation. Olopatadine hydrochloride 0.1% ophthalmic solution and artificial tears (polyvinyl alcohol 1.4% ophthalmic solution) were started twice a day and 4 times a day, respectively, in both eyes for follicular conjunctivitis, warm compresses and lid hygiene were re-advised for meibomitis, and a consult to ophthalmology (within 2 weeks) was arranged for further evaluation of the conjunctival lesion in his right eye. An IOP check, baseline visual field determination, baseline stereo disc photos, and dilation were also deferred to the follow-up examination.

Two weeks later at our cornea/anterior segment clinic, the findings were similar to those of prior examinations, and IOPs were 20 mmHg in both eyes with GAT. The dilated fundus examination was unremarkable, and a trial of prednisolone acetate 1% ophthalmic suspension every 2 hours in the right eye with a slow taper was initiated to rule out an inflammatory process in the conjunctival lesion. Follow-up was scheduled in 3 weeks for consideration of excisional biopsy, but the patient failed to return as instructed and was lost to follow-up.

Three years later, the patient returned for evaluation of the conjunctival lesion. He reported growth of the conjunctival lesion since his prior examinations with increased redness and foreign body sensation in both eyes. He denied using any medications, including eye drops, and his presenting visual acuities remained 20/20 in each eye. Anterior segment evaluation with a slit lamp found a papillary reaction in both eyes and nasal pterygia extending now 2.0 mm across the limbus in his right eye and 2.7 mm in his left eye. The elevated conjunctival lesion previously noted in the right eye was now noted to be 4.6 mm by 5.8 mm, and 2 larger "feeder" vessels in the temporal bulbar conjunctival lesion in his right eye were also noted. Baseline anterior segment (*see* Figure 1) and optic nerve head photos were taken in both eyes with patient consent. An excisional biopsy of the conjunctival lesion in the right eye was planned with subsequent excisional biopsy of the pterygia also considered.

An excisional biopsy of the conjunctival lesion was performed, and histopathologic examination found a