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CASE REPORT
Cosmetic

Incidental Diagnosis of Four Lid Orbital Lymphoma during a Blepharoplasty

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Summary: Chronic lymphocytic leukemia is characterized by the accumulation of mature CD5-positive B-cells in the lymphoid organs.[1] Extranodal involvement occurs in up to 10% of cases and can arise in various tissues, including the orbit. Less than 400 cases of orbital lymphoma are diagnosed per year in the United States, typically manifesting as a form of B-cell non-Hodgkin lymphoma, with extranodal marginal zone B-cell lymphoma being the most common subtype. Orbital lymphoma typically presents with proptosis and a palpable mass; however, patients may also have a relatively benign examination. Here, we present a 76-year-old man with symmetric dermatochalasis and marked fat prolapse of all four lids, who was incidentally diagnosed with secondary orbital lymphoma in all four eyelids during a cosmetic four lid blepharoplasty. His history was significant for RAI Stage 0 chronic lymphocytic leukemia diagnosed 15 years before consultation. Orbital lymphoma presenting as orbital fat prolapse has only been reported a few times in the literature. To our knowledge, this is the first case of secondary orbital lymphoma in all four eyelids found incidentally during an aesthetic four lid blepharoplasty.
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Chronic lymphocytic leukemia (CLL) is characterized by the clonal proliferation of predominately CD5-positive B-cells.[1] Extranodal involvement of CLL is rare, with rates up to 10%, and is most commonly found within the cutaneous, renal, and central nervous systems, but can occur in various tissues like the orbit.[1]

Leukemic infiltration of orbital tissues comprises less than 1% of all orbital tumors and is typically a manifestation of acute rather than chronic forms of leukemia.[2] Approximately, 43% of orbital lymphoma arises from the orbit, 21% from the conjunctiva, 26% in the lacrimal gland, and 9% in the lid.[3] The incidence of orbital lymphoma is approximately 1.14 cases per million people annually in the United States.[4] It is most common in the seventh decade of life, with a male-to-female ratio of 1:2.5.[3] Orbital lymphoma typically manifests as a form of B-cell non-Hodgkin lymphoma, with extranodal marginal zone B-cell lymphoma being the most common subtype.[4]

Orbital lymphoma typically presents with proptosis and a palpable mass.[5] Bilateral presentation is rare, ranging from 7% to 24%. Interestingly, there are no lymphatic connections between the orbits, which means that there are likely systemic predisposing conditions that render both orbits at risk.[3]

The diagnostic criteria of primary orbital lymphoma include a biopsy consistent with orbital lymphoma, without evidence of lymphoma in other organs or prior history of lymphoma.[5] Secondary orbital lymphoma is diagnosed with biopsy in patients with history of lymphoma originating from other organs.[5] Approximately 73% of orbital lymphomas are primary tumors; 27% are secondary to metastatic spread.[6] The differential diagnosis includes malignant or inflammatory pseudo-tumors of the orbit and thyroid-associated orbital disease.[3]

Secondary orbital lymphoma is rare. To our knowledge, this is the first case of secondary orbital lymphoma in all four eyelids found incidentally during an aesthetic four lid blepharoplasty. The patient did not demonstrate any typical signs or symptoms of orbital lymphoma. Orbital lymphoma presenting as orbital fat prolapse has been reported in the literature only a few times.[7]

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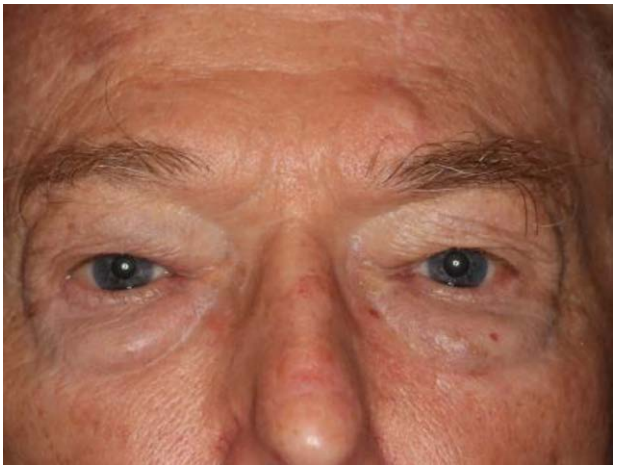

Fig. 1. Preoperative photographs showing symmetric dermatochalasis and marked fat prolapse of all four eyelids.

CASE

A 76-year-old man presented to an oculoplastic surgeon with symmetrical dermatochalasis of all four eyelids for approximately 1.5 years (Fig. 1). He was interested in improving the aesthetic appearance of his lids. His history was significant for CLL, diagnosed 15 years prior (RAI stage 0). His blood counts were stable; therefore, he was followed up without treatment. He had bilateral brow ptosis, dermatochalasis, marked fat prolapse of all four lids, and mechanical ptosis of bilateral upper lids. TSH, free T4, and thyroid antibodies were ordered to rule out a thyroid-related ophthalmopathy contributing to his fat prolapse, which were all within normal limits. The patient was then consented for an elective four lid blepharoplasty.

The procedure began with an upper eyelid blepharoplasty. There was anterior orbital fullness consistent with prolapsing orbital fat pads. When an incision was made through the orbital septum, an orbital mass associated with the orbital fat and lacrimal gland was identified (Fig. 2). The mass had an irregular, firm consistency and gray discoloration. The anterior orbit was dissected anterior to the levator, and the mass was biopsied and sent to pathology. The suspicion for malignancy was significant, especially in the setting of the patient's history of CLL, so it was felt appropriate to complete the bilateral upper lid blepharoplasty, anterior orbitotomy and biopsy, but defer the lower lid blepharoplasty, as the lower lid fat prolapse was now suspicious for malignancy.

The pathology was consistent with CLL/small lymphocytic lymphoma. A postoperative PET scan was negative for metastasis. The patient was referred to oncology and placed on rituximab and prophylactic acyclovir. Clinical follow-up at 8 months revealed no signs of orbital lymphoma, with persistent lower eyelid dermatochalasis and fat prolapse (Fig. 3). The patient is considering lower lid blepharoplasty in 1 year if he has no signs of recurrence.

DISCUSSION

Signs of orbital lymphoma include periorbital edema; erythema; epiphora; diplopia; visual acuity changes; pain; asymmetric dermatochalasis; limited eye motility; or most commonly, a palpable mass (80%) and proptosis (52%).[5,8] Although these findings are nonspecific, they warrant further workup and referral to ophthalmology. In our case, the patient had no concerning signs or symptoms; each medial fat pad felt of normal consistency, and the fat pad herniation was symmetric in all four lids. The intent of the blepharoplasty was for purely aesthetic reasons, to improve the appearance of the patient's tired eyes.

Few cases of incidental orbital lymphoma have been reported in the literature. This is the first case we know of secondary orbital lymphoma (CLL/small lymphocytic lymphoma) incidentally presenting in all four eyelids during a cosmetic blepharoplasty. The only patient factor

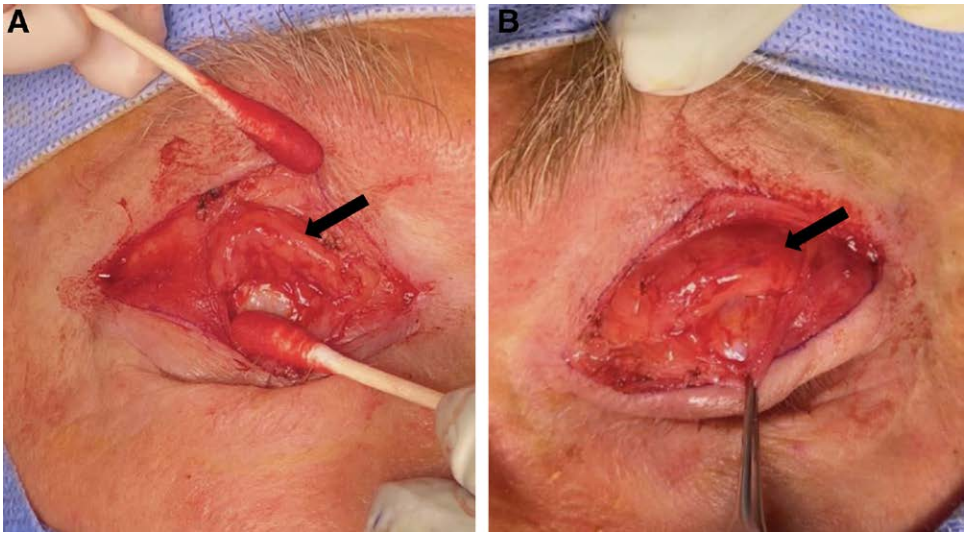

Fig. 2. Intraoperative appearance of the orbital masses found deep to the septum. A, Right upper eyelid. B, Left upper eyelid. A forceps is being used to retract the septum.

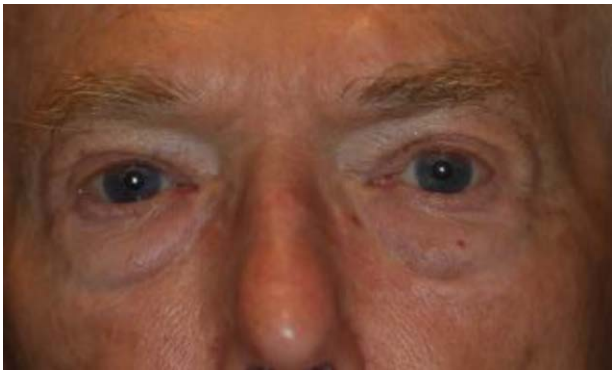

Fig. 3. Postoperative photograph at 8 months following treatment with Rituximab and resolution of lymphoma on PET scan. The patient has persistent lower eyelid dermatochalasis and fat prolapse.

provoking suspicion for malignancy was his history of CLL diagnosed 15 years prior. It is thus important to be able to distinguish normal from diseased tissue. Intraoperative appearance of orbital fat ranges widely in the literature. In the upper lid, the medial fat pad is usually lighter in color, more rounded and fibrous compared with the central fat pad.[9] In orbital lymphoma, the fat most commonly has a gray discoloration with an irregular, tougher consistency, like in our patient, but may also present as a pink, firm, highly vascularized mass.[7]

Upon discovery of questionably diseased tissue, it is recommended to abort the planned operation because these tumors require an oncologic evaluation to detect the degree of systematic involvement and required treatment. Additionally, aborting the procedure avoids potential oncologic seeding. In our case, the bilateral upper lid blepharoplasty was started simultaneously by two surgeons, so it felt appropriate to biopsy both sides.

CONCLUSIONS

In the United States, over 300,000 blepharoplasties are performed annually.[10] Although encountering incidental tumors is uncommon, it is critical not to overlook the possibility of orbital lymphoma disguising itself as herniated fat pads, even in all four eyelids. Obtaining a thorough history and physical is paramount. Although a palpable mass and proptosis are most commonly found in orbital lymphoma, patients may have a relatively benign examination. If intraoperative appearance is questionable, we recommend obtaining a biopsy and aborting the planned procedure, especially in patients with history of lymphoma and in procedures where the orbital fat has a firm consistency and gray discoloration.

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DISCLOSURES

The authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

The patient provided written consent for the use of their image.

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