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Bilateral lacrimal sac swellings in the elderly: Not always a mucocele

A 62-year-male, hypertensive, had a 6-month history of painless, progressive bilateral (OU) lacrimal sac region swellings without epiphora. Examination revealed pea-sized swelling over the lacrimal sac regions OU (left eye > right eye) [Fig. 1a]. On syringing and irrigation, lacrimal system was freely patent. A detailed evaluation revealed inferior dystopia of the right eye (OD), and pinkish-red, lobulated, subconjunctival mass with vascularization was noted in the right superior and left inferior fornices [Fig. 1a]. The rest of ophthalmic examination was normal. MRI orbits revealed similar soft-tissue lesions in both orbits, lacrimal sacs, and nasolacrimal ducts (NLD) [Fig. 1b]. Lacrimal sac fine needle aspiration cytology (FNAC) [Fig. 1c], and orbitotomy-incision biopsy [Fig. 1d] suggested a low-grade non-Hodgkin's mucosa-associated lymphoid tissue (MALT) lymphoma (predominant CD20+).

What is Your Next Step?

A. Orbitotomy and excision biopsy + dacryocystorhinostomy
B. Orbitotomy and excision biopsy + dacryocystectomy
C. Systemic workup + orbitotomy and mass excision + dacryocystorhinostomy
D. Systemic workup followed by chemotherapy ± radiotherapy.

Correct Answer: D

Findings

The systemic workup revealed splenomegaly, enlarged para-aortic lymph nodes, and bone marrow infiltration with lymphomatous cells. Systemic chemotherapy was advised by the hemato-oncology department, including cyclophosphamide, doxorubicin, vincristine, and prednisolone. After the 6th cycle, the patient showed complete resolution of both orbital and lacrimal sac region masses [Fig. 1e]. Radiotherapy was deferred due to complete resolution post-chemotherapy. The patient was alive and disease-free locally, and systemically at 2 years follow-up recently.

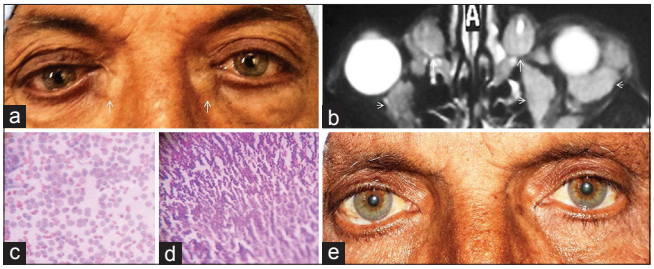

Figure 1: (a) Bilateral lacrimal sac region swellings (white arrows), right inferior dystopia, and left inferior orbital sulcus fullness. (b) MRI orbits (axial T2-weighted) shows bilateral lacrimal sac wall thickening (white arrows) and iso-intense orbital masses (arrowheads). (c) Cytology (10×) shows small round blue cells with coarse chromatin and scanty cytoplasm. (d) Sheets of round blue cells suggestive of lymphomatous malignancy (10×, H and E). (e) After chemotherapy, bilateral lacrimal sac regions and the right eye appears normal

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Diagnosis

Bilateral lacrimal sac and orbital non-Hodgkin's MALT lymphoma.

Discussion

The MALT lymphomas are common among all lacrimal sac lymphomas (LSL).[1] The diffuse large B-cell lymphoma behaves more aggressively, hence, have a poorer prognosis.[1,2] Among Japanese, 50% of LSL is of diffuse B-cell type while in western literature, the MALT lymphoma is reported to be more common (33%).[2] The majority of LSL patients do not have epiphora due to the patent lumen of NLD as the lymphoma cells infiltrate the lacrimal drainage system associated lymphoid tissue (LDALT).[1-3] The lack of prominent symptoms may lead to delayed presentation and diagnosis of LSL. MRI provides adequate information for orbital-adnexal lymphomas.[1-3] Chemotherapy provides a successful outcome in terms of both local and systemic disease control.

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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