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Case Report

Orbital MALT Lymphoma: A Case Report

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Abstract:
A case of orbital MALT (mucous associated lymphoid tissue) lymphoma is reported for its rarity. It presented as a large tumor obscuring the whole eye with loss of vision, without any signs of dissemination and remained free of recurrence or metastasis 12 months after undergoing simple surgical excision.
Key Words: Orbital MALT Lymphoma

Case Report

A 52 years old man presented with a slow growing mass arising from the right orbit, (Fig. 1,2) encroaching upon the eyeball over the last 4 months resulting in loss of vision. He also had blood stained discharge from the right eye.

Figure 1: Mass arising from the right orbit

The patient had undergone a combined surgery (for cataract and secondary open angle glaucoma) in the same eye 8 years ago, as per his previous records.

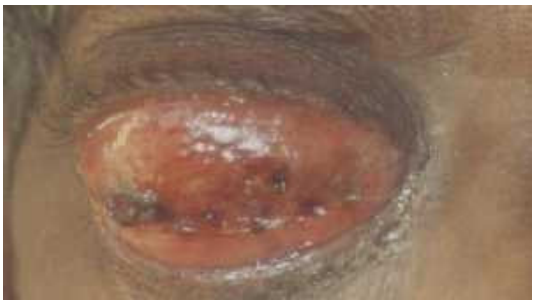

Figure 2: Mass arising from the right orbit

Examination revealed a right orbital mass filling the interpalpebral area, firm with irregular surface, non-tender, non-pulsatile, with pigments present on the surface of the mass. Right eyeball was not visible. The other eye was normal on examination. There were no enlarged lymph nodes and no significant findings on systemic examination. Routine blood investigations were within normal limits. B scan showed clear orbital cavity with hyper-echoic shadow surrounding it. An excisional biopsy was performed.

On gross appearance the mass appeared white and fleshy. Cut section was white, homogenous, surrounding the whole of the anterior part of the eye. Tumour was extending into the sub-conjunctival space and anterior chamber.(Figs.3,4,5)

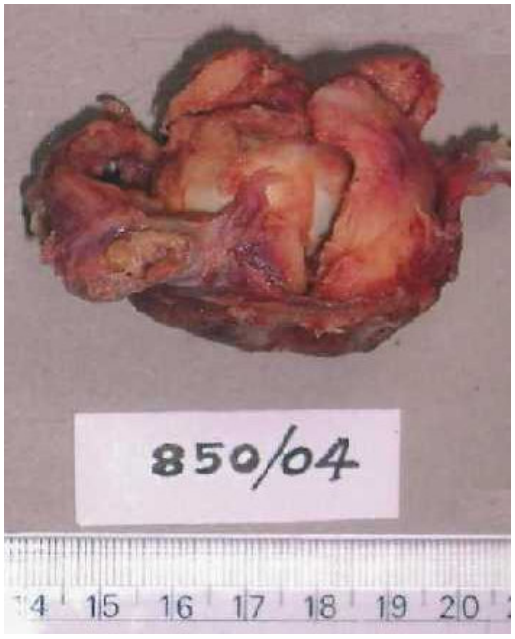

Fig. 3: Eyeball surrounded by the tumor

Histopathological examination revealed diffusely arranged, densely cellular lymphoid cells in a sparsely cellular stroma. Tumour cells were medium sized with irregular nuclei, clumped chromatin, inconspicuous nucleoli and abundant pale cytoplasm. Admixed with these were small lymphoid cells with oval nuclei and scanty cytoplasm. A few immunoblasts and plasmacytoid cells were also seen.

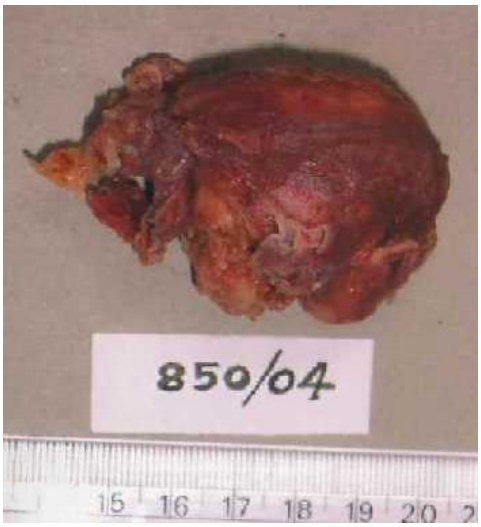

Figure 4: Posterior view of the tumor

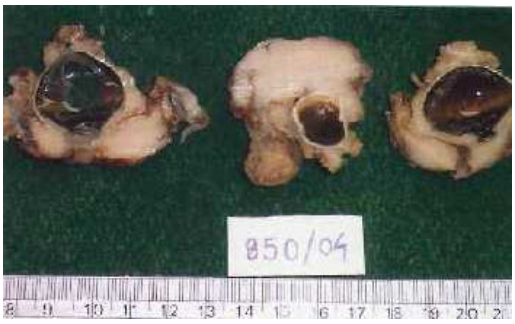

Figure 5: Enucleated eyeball with cut-section of the tumor

Figure 6: Histopathology of orbital MALT Lymphoma

On the basis of these findings, a diagnosis of MALT Lymphoma was made.

Patient is under follow-up and has remained free of recurrence or systemic dissemination after 6 months of surgery.

Discussion:

MALT lymphoma is part of a group of low-grade B cell lymphomas presenting in the gut, lungs, thyroid, salivary gland and conjunctiva. It may arise in lacrimal gland affected by Sjogren's syndrome.(1) Clinical manifestations of MALT Lymphoma vary from mild photophobia to mild redness and irritation and orange pink or pale masses arising from either the upper or lower fornices. Typical appearance of MALT Lymphoma shows centrocyte like cells infiltrating around lymphoid follicles and intraepithelial B cells.(2)

MALT Lymphoma differs from other forms of B cell extra-nodal non-Hodgkins lymphoma in that it is not as aggressive in behaviour and remains localized to mucosal surfaces rather than disseminating systemically. This appears to account for the better prognosis observed with MALT Lymphoma compared to that with non-MALT Lymphoma arising at the same site. Petrella et al (3) had also described a case of unilateral conjunctival lymphoma (MALT type) which showed no signs of dissemination at presentation and remained free of recurrence or metastasis 12 months after undergoing simple surgical excision.

There has been some speculation on why orbital lymphomas tend to be confined to the orbit for long periods before metastasis. One possibility is that they are initially reactive lesions and only subsequently become truly neoplastic. The absence of lymphatic drainage channels might also impede dissemination.(4)

The present case had a history of ocular surgeries 8 years prior to the development of the MALT Lymphoma. We ask whether there could be any relationship between the two?

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