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Case Report

Diffuse large B-cell lymphoma of the orbit: A tertiary eye care center experience in Saudi Arabia ☆

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Abstract

Primary Diffuse Large B-cell Lymphoma (DLBCL) represents the 2nd most common lymphoma occurring in the orbit, after Mucosal Associated Lymphoid Tissue (MALT) lymphoma. A total of 5 cases of ocular adnexal DLBCL were diagnosed over 25 years of experience at our tertiary eye care center. Two cases involved the lacrimal sac and one case involved the lacrimal gland. In this paper we are presenting the remaining 2 non-lacrimal cases of DLBCL. The first case is a 32 year old male who was referred with a slowly growing, painless mass involving the left medial canthal area as a case of dacryocystitis. The mass was found to be extending into the orbit inferiorly with upward displacement of the left globe. The second case is a 65 year-old lady who presented with unilateral proptosis as a result of a right orbital mass extending to the orbital apex. Histopathologic examination and immunohistochemical analysis of the incisional biopsy in both cases confirmed the diagnosis of Diffuse Large B-cell Lymphoma (DLBCL).

Keywords: B-cell lymphoma, Large cell, Orbit

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Introduction

Lymphoproliferative disease of the orbit includes a wide range of disorders including lymphoid hyperplasia and indolent lymphomas. Primary Diffuse Large B-cell Lymphoma (DLBCL) represents the 2nd most common lymphoma occurring in the orbit, after Mucosal Associated Lymphoid Tissue (MALT) lymphoma. The diagnosis requires careful clinical, radiological, histopathological and immunohistochemical examination.[1]

Five cases of ocular adnexal DLBCL cases were diagnosed in our institution over a 25-year experience. Two cases involved the lacrimal sac, one case involved the lacrimal gland and the remaining last 2 cases showed different non-lacrimal orbital involvement. We are reporting these 2 cases of DLBCL with an overall clinical, histopathologic and radiologic data. Both cases presented initially with ophthalmic manifestations and no history of systemic lymphoma. The mass in the first case involved the inferior orbit with no proptosis but visible extension to left lacrimal sac area and the case was referred as dacryocystitis. The second case presented with a typical right orbital mass resulting in unilateral proptosis. The systemic work up in the first case was completely negative, thus this lesion was considered primary. Work up of the second case revealed mediastinal lymphadenopathy indicating a possible secondary orbital involvement.

Case 1

A 32 year old male was referred by a general ophthalmologist as a non-urgent case of left chronic dacryocystitis. The patient gave a history of tearing and a slowly growing, painless swelling just near the left medial canthal area for several months. There was no history of trauma, blood transfusion or

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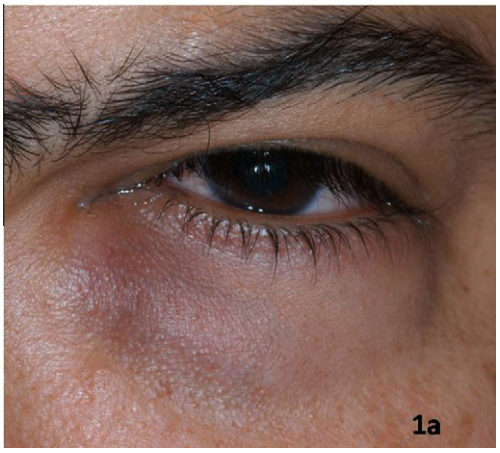

Figure 1a. The clinical appearance of the left medial canthal area swelling in case 1.

extramarital exposure. No history of fever, weight loss or other systemic symptoms. Past medical and drug history were not significant. Family history was also unremarkable.

On examination, the best corrected visual acuity was 20/20 in both the eyes. The intraocular pressure (IOP) was 16 mmHg in each eye. Examination of the right eye was within normal limit. External examination of the left eye revealed fullness of left lower lid involving the medial canthal area. A palpable tender firm mass was felt extending from the medial canthus to the anterior part of the orbit just inferior to the left globe (Fig. 1a). The globe was displaced upward with no evidence of proptosis. Regurgitation from upper and lower puncta was negative. Ocular motility was full. The remaining ocular examination was within normal limits. His systemic examination revealed no abnormality.

A Computerized Tomography (CT) scan of the left orbit showed a mass with intermediate attenuation measuring 2.9 × 2.8 cm in the left inner canthus region in close proximity to the lacrimal sac (Fig. 1b). Magnetic Resonance Imaging (MRI) of the left orbit showed a soft tissue mass of intermediate signal intensity on T1 and T2 weighted images in the area of the left lacrimal sac with intra-orbital, extra-conal

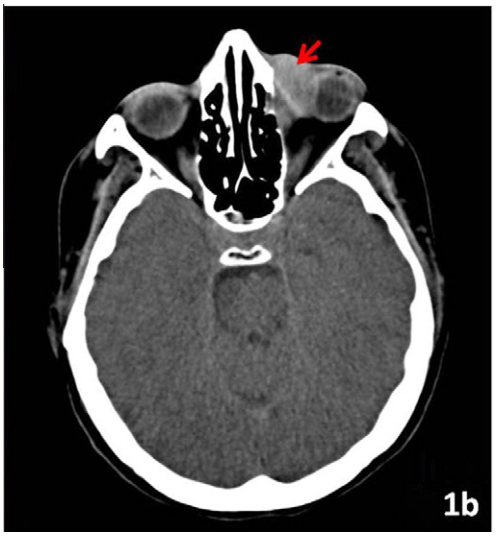

Figure 1b. Computerized tomography (Axial) of the left orbit with contrast enhancing mass medially (red arrow).

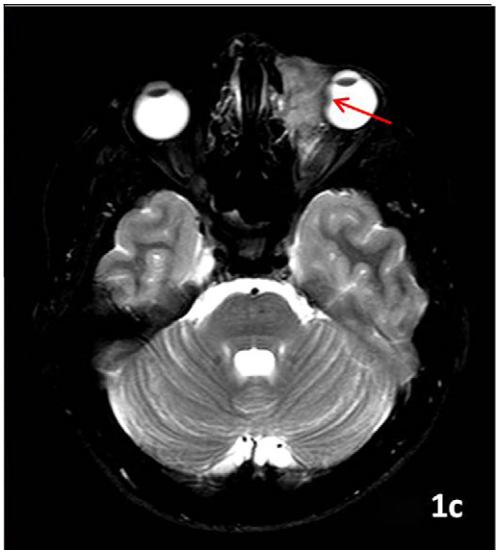

Figure 1c. Axial T2 weighed magnetic resonance image of the left orbital lesion. Note the globe indentation by the mass (red arrow).

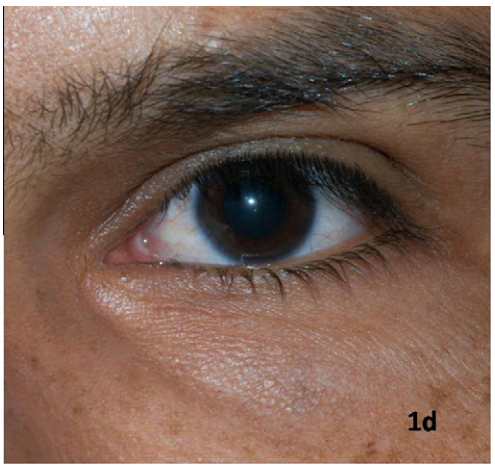

Figure 1d. The clinical appearance of the same patient showing resolved swelling following treatment.

component, all measuring 3 × 1.8 cm (Fig. 1c). The globe was pushed laterally with abnormal contour of its wall medially. The mass showed mild homogenous contrast enhancement except for a non-enhancing center.

The patient underwent debulking of the lesion through a lower eyelid incision. The mass was found intra-operatively to be separate from the lacrimal sac. The Histopathologic examination revealed diffuse proliferation of sheets of large lymphocytes with frequent mitotic figures. The cells showed evidence of pleomorphism with vesicular nuclei and prominent nucleoli (Fig. 2a). The immunohistochemical staining showed that the cells were CD20, Bcl2 and Ki-67 positive (Figs. 2b,c,d) while negative to CD10. The differential histopathologic diagnosis based on the morphology included Diffuse Large B-Cell Lymphoma (DLBCL) and Burkitt lymphoma, however the immunohistochemical staining (specifically the strong positivity to CD 20 and the negative CD10) supported the diagnosis of DLBCL. The specimen was also submitted for FISH analysis for c-myc translocation, which was negative, thus ruling out the diagnosis of Burkitt lymphoma.