Rosai-Dorfman disease: Isolated epibulbar mass in patient with vernal keratoconjunctivitis

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Abstract:

A 29-year-old male known case of vernal keratoconjunctivitis (VKC) presented with 5-month history of a rapidly growing mass on the temporal aspect of the right cornea. The 9 × 9 mm epibulbar lesion was excised and histologically showed features of extranodal Rosai Dorfman disease (RDD) with emperipolesis of eosinophils. The lesion did not recur following excision. The association of RDD with VKC has not been previously reported; however, the causal relationship remains unclear.

Keywords:

Limbal mass, rosai dorfman disease, vernal keratoconjunctivitis

Introduction

Rosai-Dorfman disease (RDD) which is known also as sinus histiocytosis with massive lymphadenopathy (SHML),[1] is a rare histiocytic disorder in which non-Langerhans sinus histiocyte over-produced without clear cause and accumulate in the lymph nodes or less commonly in other areas of the body which may lead to organ damage. RDD is an entity that uncommonly involves the eye. We report a case of vernal keratoconjunctivitis with RDD presenting as an isolated epibulbar mass without lymphadenopathy in a healthy young patient.

Case Report

A 29-year-old male patient known case of keratoconus presented with a rapidly growing mass of the temporal aspect of the right cornea for five months which was associated with redness and itching. Systemic history was significant for positive sickle cell disease and eczema. His family history included diabetes mellitus in the mother and social history suggested he was a smoker. On examination his uncorrected visual acuity in the right eye was 20/20. His uncorrected visual acuity in the left eye was 20/40 and 20/20 with +0.75–3.35 × 130. Intraocular pressure was 16 and 14 in the right and the left eye respectively. Slit-lamp examination of the right eye showed diffuse papillary hypertrophy on the superior tarsus with mucoid nodules around the limbus and Horner-Trantas dots. A 9X9 mm solid round non-mobile non-tender pinkish mass with whitish dots on its smooth surface was noted. The mass extended from the temporal limbus and invaded one third of the cornea [Figure 1a]. The surrounding bulbar conjunctiva was injected with enlarged and tortuous blood vessels. The central cornea was clear and the remaining ocular examination was unremarkable. Examination of the left eye showed diffuse papillary hypertrophy mainly on the superior tarsus.

Corneal topography with Pentacam showed changes consistent with keratoconus in both eyes. Ultrasound biomicroscopy of the right eye lesion showed a large mass temporally invading the cornea but no scleral invasion. B-scan ultrasonography was unremarkable. The lesion was excised and submitted for histopathology.

Post-operatively, the patient was treated with Prednisolone Acetate 1% drops four times daily and Ofloxacin drops four times daily. In addition, the vernal keratoconjunctivitis was

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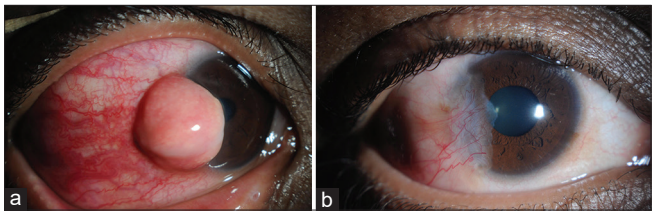

Figure 1: (a) Photograph showing large solid non-mobile epibulbar mass with injected surrounding conjunctiva. (b) Photograph showing scarring and pannus at the site of excision four months postoperatively

treated with Epinastine hydrochloride 0.05% drops twice daily and lubrication in both eyes and cyclosporine 1% drops four times daily in the right eye. On subsequent post-operative visits, he was symptom-free and maintain his pre-operative good vision. On clinical examination there was a reduction in the papillary reaction in both eyes with a scar and pannus at the site of excision [Figure 1b]. Although pannus reached near the center of the cornea at the site of excision but its central edge remains stable and the patient vision does not drop below 20/20. Such pannus formation is most probably due to damage to the limbal stem cells at that area because of the lesion itself or as the complication ofthe surgery. In case of vision drop the patient may benefit from removal of this pannus and autologouslimbal stem cell transplantation. One year later, there was no recurrence in the right eye but his keratoconus progressed more in the left eye for which he underwent corneal cross-linking.

By light microscopy the conjunctival nodule showed extensive infiltration by a mixture of eosinophils, lymphocytes and large foamy histiocytes. The infiltrate extended from the subepithelial region deep into the substantia propria. Histiocytes showed emperipolesis of eosinophils, polymorphonuclear leukocytes and lymphocytes. The histiocytes stained positively with CD68 and S100 and were negative for CD1A. Scattered CD 3 and CD 20 immunolabeling was also noted [Figure 2]. These pathologic findings were consistent with RDD of the limbus and the eosinophilic conjunctivitis was consistent with the history of VKC.

Discussion

Extranodal involvement by RDD has been described in 43% of cases, and most frequently involves the skin, soft tissue, upper respiratory tract, bone and orbit either with lymphadenopathy or as an isolated disease.[2] Our patient has sickle cell disease which was reported in association with RDD.[3] Extranodal Rosai-Dorfman disease (RDD) rarely involves the eye (8–11%).[2,4] Ocular structures involved include the conjunctiva, cornea, uveal tissue, sclera, lacrimal gland, ocular adnexa and the orbit.[2,4-6] Epibulbar lesions are rare and have been described in 16 cases.[6-9] The nodules appear to be solitary, rarely bilateral, pinkish in color and occurred in patients ranging in age from 13 months to 74 years. Systemic involvement with conjunctival masses is rare.[9]

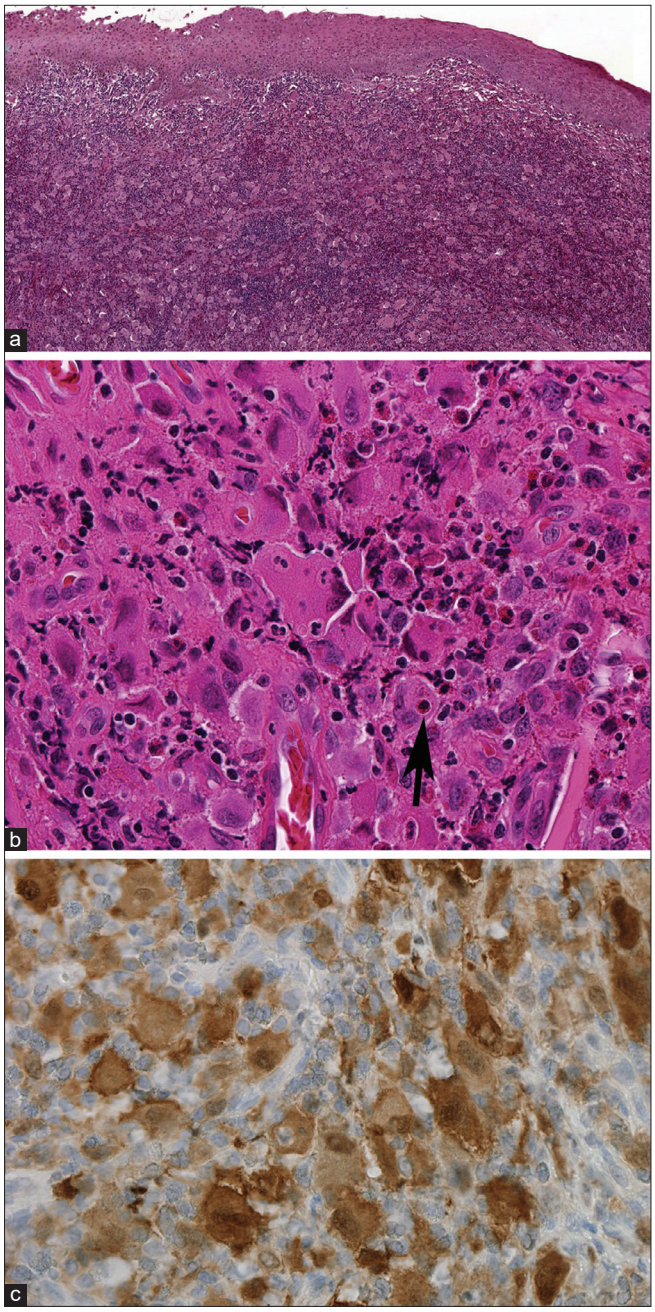

Figure 2: (a): Photomicrograph of limbal mass at low magnification. Note the extensive inflammatory infiltrate in the substantia propria and large histiocytic with pale staining cytoplasm (hematoxylin and eosin; original magnification ×4). (b): an aggregate of histiocytes at high magnification. Note the emperipolesis of eosinophils (arrow), lymphocytes and polymorphonuclear leukocytes within the histiocytes (hematoxylin and eosin; original magnification ×20). (c): note positive S-100 staining of the histiocytes (DAB chromogen; original magnification ×40)

The unusual feature of our case was the association of the limbal mass with the clinical history of VKC which has not been previously reported. Other associations like joint disease, glomerulonephritis, neoplastic and hematological disease have been described.[2] Also, the histopathological features of RDD with a background of extensive eosinophilic

conjunctivitis was noteworthy. Of interest was the observation of "eosinophil emperipolesis". Typically, lymphocytes and polymorphonuclear leukocytes are seen engulfed by the large atypical histiocytes. The relationship between VKC and RDD in this case is unclear. Whether VKC incited the occurrence of RDD is to be determined.

The clinical differential diagnosis of RDD at the limbus includes limbal dermoid, pyogenic granuloma, lymphoma, reactive lymphoid hyperplasia and juvenile xanthogranuloma. The histologic differential diagnosis includes Langerhans cell histiocytosis in which CD1a stain would have been positive and juvenile xanthogranuloma where one would have expected to see Touton type giant cells and S-100 stain would have been negative. Angiolymphoid hyperplasia may demonstrate eosinophilia, however the presence of histiocytes and emperipolesis as well as other pathologic features make the diagnosis unlikely.[10] Epstein-Barr virus[2] and human herpes virus 6[11] has been detected in RDD tissues but its presence was not tested in our case. As previously reported in other cases[2,5-9] the course of extranodal RDD is fairly benign and does not typically recur after excision.

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Conflicts of interest

There are no conflicts of interest.

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