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## Rosai-Dorfman disease of the ocular surface

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**Purpose:** Extra nodal Rosai-Dorfman disease is a rare benign condition recently reported to sometimes show features of IgG4-related disease. Ophthalmic manifestations are seen in 11% of cases including orbit (most common), eyelid, nasolacrimal system, conjunctiva and uvea. The purpose of this study was to describe the corneal-limbal manifestation of the entity and to investigate whether numerous IgG4-positive plasma cells are associated with the disease at this site.

**Methods:** We report two cases of extra nodal RDD presented as isolated limbal masses in young patients. Histopathological, immunohistochemical and pyro sequencing analysis of the lesions were performed in the surgical pathology and molecular laboratories at the Johns Hopkins Hospital using standard techniques. This is an interventional retrospective small case series.

**Results:** We report two cases in young patients presented as pink, elevated, triangular limbal lesions associated with neovascularization. No systemic involvement was present in both cases. Patients underwent excisional biopsy of the lesions for histopathological examination as well as immunohistochemistry, molecular and genetic studies. On microscopic examination both cases were characterized by an atypical histiocytic infiltrate present in the substantia propria with chronic inflammation including histiocytes with engulfed lymphocytes (emperipolesis). Immunohistochemistry was performed and the atypical histiocytes were found to be CD68 positive, S-100 positive and IgG was also positive. CD1a and IgG4 were negative. Although there is association described between extra nodal RDD and IgG4 plasma cell expression, we do not found such in our two limbal lesion cases. Point mutations (V600E) in the BRAF oncogene were absent.

**Conclusions:** Rosai-Dorfman disease should be considered in the differential diagnosis of limbal mass lesions. Involvement at this site was not associated with BRAF mutation or IgG4 abnormalities in the cases examined.

## Biography

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