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An analysis of conjunctival and periocular venous malformations: clinicopathologic and immunohistochemical features with a comparison of racemose and cirsoid lesions

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ABSTRACT

Vascular tumors (in contrast to dilations or ectasias) of the conjunctiva and other adnexal tissues are rare, with no previous convincing example of a congenital, purely venous conjunctival malformation having been described. A 33-year-old man with a previously well-tolerated racemose conjunctival lesion present from birth developed bothersome symptoms when it underwent multifocal thrombosis with papillary endothelial cell hyperplasia as part of the process of thrombotic organization. Conservative subtotal excision with placement of an amniotic graft led to an acceptable cosmetic appearance, abatement of symptoms, and retention of full ocular function. Histopathologically, the lesion was composed of patulous vascular channels with thin walls displaying a negligible and irregular muscularis, diffuse supportive mural fibrosis, and the absence of an elastic lamina. Immunohistochemically the endothelial cells were CD31- and CD34-positive (vascular origin) but D2-40-negative (lymphatic origin). An associated neovascular capillary bed was not detected. Venous (racemose or grape-like) malformations should be distinguished from: arteriovenous (cirsoid or twisted) malformations in which the vessels possess thicker and more uniform muscular walls, some of which are endowed with an elastica; varices (hemorrhoidal dilations typically of a pre-existent vein); and venous angiomas (noncongenital lesions acquired in middle life) composed of regularly structured muscular channels devoid of an elastic lamina. Other conditions not to be confused with congenital venous malformations include hemorrhagic lymphangiectasia (of Leber), hemorrhagic lymphangiomas, and complex lymphaticovenous malformations.

Vascular tumors of the conjunctiva are less frequent than those of the other periocular tissues, accounting for only 4% of nonmelanocytic tumors of this tissue.[40] Examples include pyogenic granuloma,[10,14,26,33,46] lymphangioma,[35] lymphangiectasia,[3,4,12,20,34,38] capillary hemangioma,[44] Kaposi sarcoma (with and without immunodeficiency),[9,16,23,25,36,50] varix,[41] putative cavernous hemangioma,[44] glomus hemangioma,[42] hemangiopericytoma,[14] and so-called acquired

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sessile hemangioma.[43] A previous description of conjunctival racemose/cirsoid hemangiomas is questionable.[44] In the absence of any published comprehensive comparative review, many earlier reports of periocular vascular entities have not had rigorous or clear-cut diagnostic criteria. We report a dramatic example of a conjunctival venous malformation (racemose or grape-like) with clinicopathologic and immunohistochemical findings. This case illustrates the benefits that accrue diagnostically when clinical features are correlated with histopathologic findings. We also clarify some of the salient clinical and nosological differences among conjunctival and other ocular adnexal vascular anomalies such as varix, venous angioma, complex lymphaticovenous malformation, and racemose or cirsoid malformations (often confusingly referred to as hemangiomas).

1. Case report

A 33-year-old healthy white male presented with a conjunctival vascular lesion in his right eye present since birth (Fig. 1A–1D). The lesion did not appear to enlarge with exercise or Valsalva maneuver or during viral infections. For over 20 years it had been evaluated periodically, was considered benign, and remained asymptomatic until 8 months prior to presentation. He then complained of tearing, burning, photophobia, throbbing pain, and blurred vision following an apparent increase in size and darkening hue. He was using Refresh Liquigel (Allergan, Irvine, CA) as needed for comfort. Otherwise, he had no significant past medical or ocular history.

Best-corrected visual acuity was 20/20, and intraocular pressure was 15 mm Hg in both eyes. Slit-lamp examination of the right eye revealed a large mobile, lobulated, non-pulsatile, red-blue vascular lesion involving a large part of the inferotemporal epibulbar quadrant of the globe and adjacent fornix (Fig. 1A) that extended up to 2 mm away from the corneal limbus (Fig. 1B). There was also lateral and superior forniceal extension (Fig. 1C). The thickest portion was elevated 4 mm. Associated large, tortuous vessels were present in the inferior fornix (Fig. 1D). There was no obvious enlargement or change in configuration of the mass during a Valsalva maneuver. The cornea was clear without a sectoral arcus, and the iris and the lens were normal. There was no proptosis, globe displacement, or motility disturbance. Magnetic resonance imaging (MRI) and angiography disclosed an enhancing, crescentic lesion measuring 24 × 20 × 5 mm that displaced the right lacrimal gland superiorly (Fig. 1E, 1F) indicative of primary involvement of the anterior orbit. There were

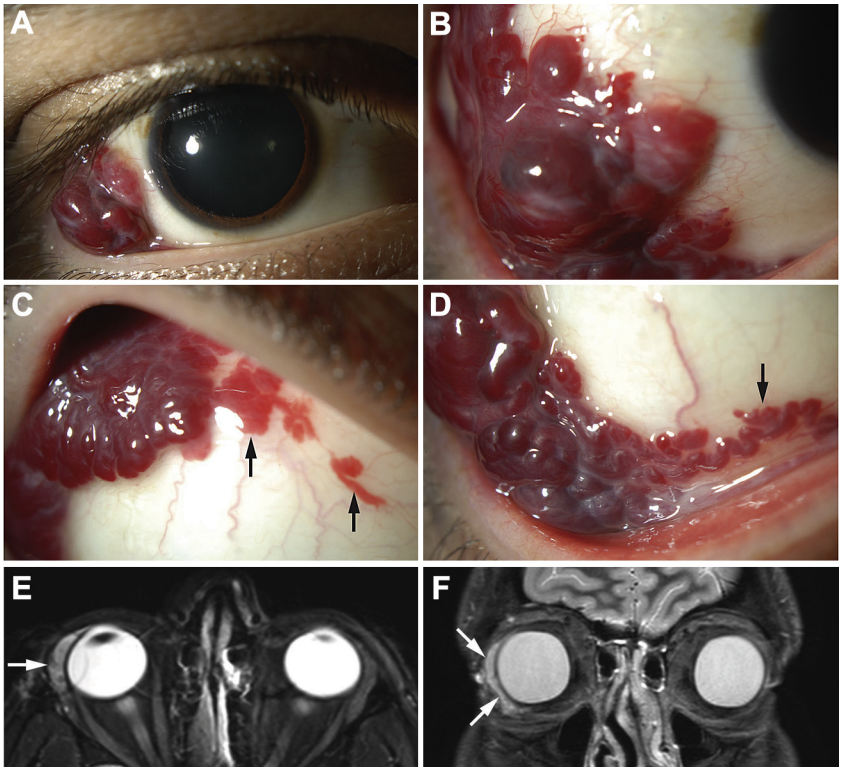

Fig. 1 – Conjunctival venous malformation. *A*: A reddish-blue multilobular, movable mass in the inferotemporal quadrant of the right eye of a 33-year-old man had been present since birth. It had been asymptomatic until a recent abrupt enlargement. *B*: Close-up view of the lesion with irregular epibulbar margin. *C*: The mass extended to the superolateral fornix and was associated with small flat subconjunctival hemorrhage (*arrows*). *D*: Enlarged, probably venous channels (*arrow*) in the inferior fornix blend with the mass. *E*: Axial magnetic resonance image reveals an anterior orbital component of the lesion in the vicinity of the lacrimal gland (*arrow*). *F*: Coronal MRI demonstrates that the anterior orbital component involves the tissues lateral to the globe from the lacrimal region (*top arrow*) to the inferior orbit (*bottom arrow*).