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Case Report

Conjunctival lymphangiectasia in the setting of cavernous sinus thrombosis

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Abstract

We present a unique case of cavernous sinus thrombosis as a likely etiology of conjunctival lymphangiectasia, with resolution of symptoms with anticoagulation. A 58-year-old male presented with 6-months of chemosis and conjunctival lymphangiectasia. Magnetic resonance imaging revealed a right cavernous sinus and superior ophthalmic vein filling defect, consistent with thrombosis. The patient received anticoagulation, with subsequent resolution of the conjunctival lymphangiectasia. This case highlights the importance of imaging in patients who present with conjunctival lymphangiectasia to rule out possible life-threatening pathology.

Keywords: Conjunctival chemosis, Cavernous sinus thrombosis, Lymphangiectasia

Introduction

Conjunctival lymphangiectasia is a relatively uncommon ocular surface disorder, presenting as dilation of the lymphatic vessels of the conjunctiva [1]. This can present as diffuse dilation with chemosis or focal dilation of vessels, seen as single or multiple cysts. It is primarily acquired due to disruption or obstruction of the lymphatic and/or venous system in the setting of prior surgery, trauma or inflammation, which leads to retrograde flow through the lymphatic system [1]. It can also be associated with other systemic syndromes such as Turner syndrome or Klippel-Trenaunay-Weber syndrome [2]. Diagnosis of lymphangiectasia is most commonly confirmed with biopsy. Histologically it presents with dilated lymphatic channels lined by a flattened endothelium [1]. The surrounding lamina propria is often edematous secondary to vessel leakage. D2-40 chemical staining can confirm the presence of lymphatic endothelium. Diagnosis can also be made based on anterior segment OCT (AS-OCT). Work up must rule out any head and neck masses or lymphadenopathy. It is generally self limited but may require intervention such as lubrication, topical anti-inflammatory medications, surgical removal, or cryotherapy, in the setting of ocular surface irritation. We present a patient with unilateral conjunctival lymphangiectasia of a unique etiology.

Case Report

A 58-year-old hypertensive male initially presented with about 6 months of chemosis in the right eye that was unresponsive to topical steroids or antibiotics. He denied vision changes, headaches, or other associated symptoms and reported otherwise feeling well. On initial presentation, visual acuity was 20/25 OD and 20/20 OS. His pupils, intraocular pressure, and extraocular movements were all normal. Of note, the patient had no cranial neuropathies or proptosis. Slit lamp examination of the right eye revealed diffuse conjunctival lymphangiectasia with focally dilated blood vessels and inferior chemosis (Figure 1). Slit lamp exam of the left eye was normal. Dilated fundus examination of both eyes was normal, including no disc edema. His exam was felt to be consistent with conjunctival lymphangiectasia. A biopsy was obtained and pathology confirmed the diagnosis of conjunctival lymphangiectasia (Figure 2). Magnetic resonance imaging (MRI) and magnetic resonance venography (MRV) were ordered to rule out compressive pathology of the lymphatic system.

MRI and MRV of the brain and orbits with and without contrast showed right cavernous sinus and superior ophthalmic vein filling defects, consistent with cavernous sinus thrombosis. The patient subsequently underwent a hypercoagulable work-up, which was negative.

The patient was started on anticoagulation. Within 6 weeks of initiating anticoagulation, the patient noted resolution of his conjunctival lymphangiectasia (Figure 3). He did not develop any other symptoms during this time period.

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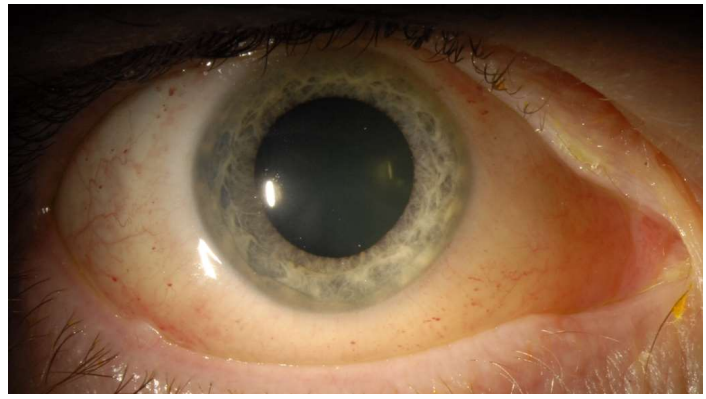

Figure 1: Slit lamp photo of the right eye showing conjunctival lymphangiectasis and chemosis.

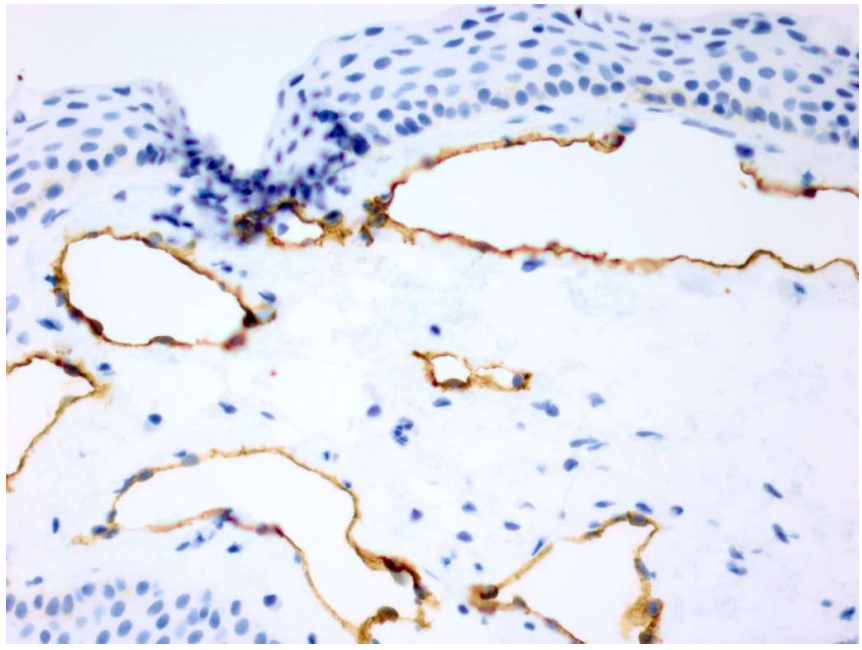

Figure 2: Pathology slide stained with D2-40, indicating lymphatic endothelium.

Figure 3: Anterior segment photo of the right eye showing resolution of conjunctival lymphangiectasis and chemosis.

Discussion

Conjunctival lymphangiectasia is a relatively rare entity associated with the presence of dilated lymphatic vessels within the conjunctiva. While it is known that outflow obstruction causes lymphangiectasia, we present cavernous sinus thrombosis as a likely, previously unreported, etiology of this outflow obstruction to highlight the need for a thorough workup in every patient that presents with this diagnosis.

Outflow obstruction leading to lymphangiectasis is generally thought to be acquired secondary to lymphatic scarring or mechanical outflow obstruction due to prior surgery, trauma, or inflammation [1]. Pertinent etiologies include any known conditions resulting in hypoproteinemia, local venous hypertension, and increased vascular permeability, as these can all cause chemosis and vascular and lymphatic dilatation. Patients often present with focal dilatation of the lymphatic vessels or with diffuse chemosis, as in our patient. While this is a clinical diagnosis, pathology and/or imaging (i.e. anterior segment OCT or B-scan) can help confirm the diagnosis and rule out other possibilities [1-4].

Work up generally involves a thorough systemic review of symptoms and ruling out any head and neck masses or lymphadenopathy that could cause outflow obstruction. While the incidence of compressive or obstructive pathology as the cause of conjunctival lymphangiectasia is unknown, missed or delayed systemic diagnosis can lead to significant systemic complications, making it important that an ophthalmologist recognize this entity and appropriately obtain proper work up. In our case, head and neck imaging was concerning for a cavernous sinus thrombosis in a patient that was otherwise completely asymptomatic.

Conjunctival lymphangiectasia generally does not require intervention unless the patient experiences persistent ocular surface irritation. Oftentimes, as with our patient, reversing the outflow obstruction is enough to resolve the lymphangiectasia. If it persists, topical anti-inflammatory medications or lubrication are the first line treatments [1]. If these treatments fail, reported treatment options include surgical removal, cryotherapy, or subconjunctival injection of anti-vascular endothelial growth factors (anti-VEGF) [1,5-7].

Venous dilation is a known symptom of cavernous sinus thrombosis but there have been no case reports documenting an association between biopsy-proven conjunctival lymphangiectasia and cavernous sinus thrombosis. It is interesting to note that our patient did not have any other manifestations of cavernous sinus thrombosis or superior ophthalmic vein thrombosis. However, given the patient improved with anticoagulation, we suspect that venous backflow was the cause of his conjunctival lymphangiectasia. We recommend intracranial imaging for patients in which the cause of lymphangiectasia is unknown or when there is a high suspicion for an underlying intracranial etiology in order to rule out any life threating causes.

Statement of Ethics

The subject has given informed consent to publish their case. Information revealing the subject's identity has been avoided.

Disclosure Statement

The authors have no conflicts of interest to declare.

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