Case Report

# An Unusual Case of Blepharochalasis

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## Abstract

**Purpose:** To describe a rare case of blepharochalasis that progressed from unilateral to bilateral involvement at five years after disease onset.
**Case Report:** A previously healthy five-year-old white boy presented to our hospital for a screening visual examination. He was found to have a 2-mm right eyelid ptosis with crepe-like skin and subcutaneous telangiectatic vessels. His mother noted that since the age of three, the child has been having two to seven day-long episodes of right upper eyelid swelling and edema with tenderness. The episodes eventually progressed to involving the left eyelid as well. Oral steroid taper was found to effectively resolve these exacerbations, and a diagnosis of blepharochalasis was made.
**Conclusion:** Blepharochalasis should be in the differential diagnosis for young children presenting with unilateral or bilateral ptosis with periorbital skin abnormalities.

**Keywords:** Bilateral Ptosis; Blepharochalasis; Oculoplastics; Orbital Inflammation; Unilateral Ptosis

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## INTRODUCTION

Blepharochalasis is a rare disorder characterized by episodes of non-tender, non-pitting eyelid edema progressing to periorbital skin atrophy.[1] Other presenting symptoms include ptosis, conjunctival redness, blepharophimosis, erythema, and proptosis.[2] Both sexes are equally affected and the onset of the disease is usually during childhood and adolescence, with an average presenting age of 11 years.[2] Most episodes last anywhere from hours to days. These episodes become less frequent with age, and most patients enter a quiescent stage;[3] however, one patient has been described as having episodes for 40 years.[4]

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Diagnosis can be difficult since there is no confirmatory test as blepharochalasis is diagnosed clinically. Computed tomography (CT) scans in previous cases have shown no abnormal findings; biopsy results are nonspecific, showing mostly a decrease in elastic fibers.[2,5] Differential diagnoses include local transient edema, orbital cellulitis, recurrent angioedema, hereditary angioedema, and recurrent contact dermatitis.[2]

Treatment is also very challenging since it is often refractory to anti-histamines and corticosteroids; however, the use of short course prednisone taper has shown improvement during exacerbations.[5] Surgery was the only option for many years, but it does not cure blepharochalasis. Once remission occurs, these patients should be monitored since recurrences have been reported to occur up to six years after the previous episode.[6]

The objective of this report is to present a rare clinical case of progressive blepharochalasis in a five-year-old boy who presented initially with right eyelid ptosis.



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His disease was rare as it progressed from unilateral to bilateral eyelids after five years.

## CASE REPORT

An otherwise healthy five-year-old white boy presented for a routine kindergarten eye exam with a noticeably ptotic right eyelid. His uncorrected visual acuity was 20/40 OD and 20/200 OS. His refractive error was +5.50 + 0.75 × 105 (20/25) OD and +7.25 + 0.50 × 128 (20/80) OS. He was orthotropic with full extraocular movements and 80 seconds of arc of stereoacuity.

His mother stated that since the age of three, he has had episodic right upper eyelid swelling. These episodes occurred about every two to three months and lasted about two to seven days with spontaneous resolution. Fever and flu-like prodromal symptoms were noted before most episodes. The right eyelid became red, completely swollen, and tender to touch [Figure 1a]. There was no associated swelling of the face or tongue. No vision changes or other associated signs or symptoms were evident during these episodes. He denied any history of trauma, and his family history revealed only a maternal aunt with rheumatoid arthritis.

On physical exam, he had 2mm right ptosis with full, redundant crepe-like skin and visible subcutaneous telangiectatic vessels [Figure 1b]. He had good levator function at 12 mm. The left eyelid was unremarkable. Conjunctiva and periorbital tissue including the lacrimal glands were normal in both sides without any palpable mass. Valsalva produced a mild redness of the right eye and lid.

Initially, the child was treated for preseptal cellulitis with oral and topical antibiotics. Recurrent exacerbations continued and began involving the left eyelid as well [Figure 2a]. Magnetic resonance imaging (MRI) of the orbits revealed soft tissue swelling but no focal mass. Hematologic and thyroid lab work-ups were normal. C1-inhibitor deficiency was not tested. Short courses of prednisone with a quick taper helped with resolution of these exacerbations [Figure 2b]. Given the recurrent nature of eyelid inflammation resulting in thin crepe-like skin and ptosis, a diagnosis of blepharochalasis was made.

The patient is now 10 years old and has three to four exacerbations per year; in the seventh year after onset, he only had one episode. In his stable phase, he has ptosis on the right with a marginal reflex distance of 0. His refractive amblyopia in the left eye resolved with spectacles, likely facilitated by the ptosis of the opposite lid.

## DISCUSSION

Blepharochalasis is a very rare disorder. According to Koursh et al, only 67 cases of blepharochalasis had been described in the literature by 2009; they found that the initial onset of symptoms occurred at an average age of 11 years.[2] Our patient was only 3 years old during his first episode, which is considerably lesser than the average age. Only 3 of the 67 cases studied had an onset on or before the age of 3 years.[2] Fever and other upper respiratory infection symptoms have been noted before the onset of blepharochalasis.[5,7] These episodes usually last several days, but can continue for weeks; inflammation and edema may be severe and may present acutely.[2,5]

The disease can be unilateral or bilateral, usually affecting only the upper eyelids, but it may also affect both upper and lower eyelids. All 67 eyes in the study by Koursh et al were either categorized as unilateral or bilateral but never both.[2] Our case is very uncommon in that it was unilateral for 5 years before becoming bilateral.

In the quiescent stage, the eyelids become lax and have a thin, wrinkled-paper appearance, and the surrounding periorbital skin becomes atrophic and discolored. Our patient likely had prior episodes of these exacerbations

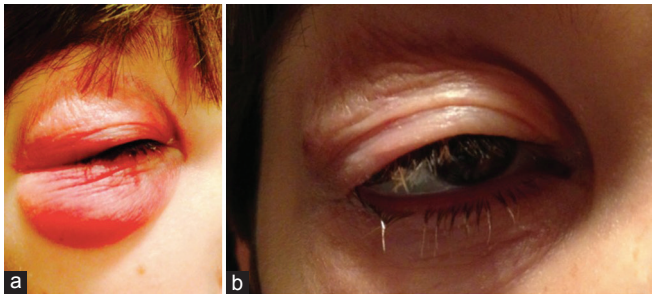


**Figure 1.** (a) Right eyelid swelling and redness during an exacerbation at 5 years of age. (b) Right ptosis with crepe-like skin and subcutaneous telangiectatic vessels at 5 years of age.

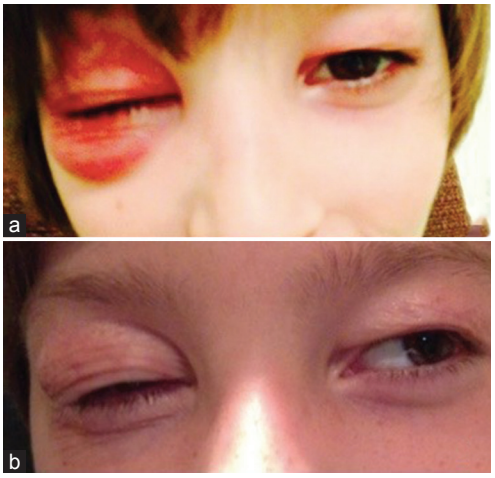


**Figure 2.** (a) Exacerbation involving the right eyelids more than the left eyelids at 10 years of age. (b) Near the resolution of the same episode in Figure 2a at 10 years of age.

by the time he presented. Ptosis is common, as found in our patient, and is present in over 75% of cases.[2] The levator aponeurosis insertion is stretched, but the function of the levator palpebrae superioris remains preserved.

The etiology of the disease remains unknown. Hormonal and allergic etiologies have not been supported; possible triggers (stress, fever, upper respiratory tract infections, etc.) have occurred in a few patients.[2] Some studies have shown that IgA antibodies may play a role by attacking elastic fibers.[8] Recent research points to a more specific immunologic mechanism that may involve matrix metalloproteinases (MMP), which degrade elastin and collagen.[8,9] Karaconji et al described two blepharochalasis cases that were treated effectively with doxycycline, which inhibits MMP.[9,10]

Differential diagnoses of blepharochalasis are numerous, and an important diagnosis to consider is idiopathic orbital inflammatory syndrome (IOIS), as it also responds to steroids and may present with periorbital edema. Signs and symptoms to look for in IOIS are proptosis, diplopia, decrease in visual acuity, acute onset of pain, and chemosis.[11] Our patient presented with none of these signs. Furthermore, IOIS rarely affects children, and if it does, it is often bilateral.

Given the age of our patient, doxycycline was not a viable option since it is contraindicated in children less than 8 years of age as it can affect teeth and bone development. In addition, surgery is not curative due to postoperative edema/exacerbation, and is often complicated by overcorrection. Takahashi et al reported that in their patient, ptosis recurred two years after surgery.[12] Furthermore, it is not recommended until attacks have subsided for six months, which our patient did not achieve.[1,6] Surgical procedures involve ptosis repair, blepharoplasty, levator resection, and lateral canthoplasty.[2] Antihistamines have also been shown to be ineffective. Our patient was administered antibiotics (for possible cellulitis) with no appropriate response. Cool compresses initially, and then corticosteroids were also administered. Oral prednisone tapers were found to resolve our patient's exacerbation. Our patient has been having recurrent episodes for seven years; however, in his seventh year, he has had only one episode, which is characteristic of blepharochalasis.

Blepharochalasis should be a differential diagnosis for young children presenting with unilateral or bilateral ptosis with periorbital skin abnormalities. Rarely, unilateral involvement may progress bilaterally as subsequent exacerbations occur.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of Interest

There are no conflicts of interest

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