Blepharochalasis at early age: A case report

Irene Loscos*; Anna Hermosa; José Carlos ABT; Daniela Rego; Noemi Roselló; Sabina Luna

Ophthalmology Department, Hospital de Sant Pau i la Santa Creu, Barcelona, Spain.

Abstract

Background: Blepharochalasis is an infrequent palpebral sickness. Few cases have been reported. Patients suffer from recurrent episodes of painless and nonpruritic eyelid edema that begins in childhood and ends up producing atrophy and periocular skin depigmentation, tissue relaxation and ptosis in adolescence or adult age.

Case presentation: Eleven-year-old male was admitted to Ophthalmopediatric unit of our hospital with a history of recurrent outbreaks of eyelid edema with secondary palpebral hyperlaxity of years of evolution, after multiple ineffective previous treatments, including antihistamine therapy. Eyelid examination showed signs of severe dermatochalasis with thin, atrophic skin and visualization of subcutaneous telangiectatic vessels. The patient was diagnosed with blepharochalasis and topical and oral antihistamines were stopped. At last physical examination, he had a receding secondary eyelid fold with secondary ptosis due to dermatochalasis worsening. Currently the patient is waiting the indication of surgery depending on the evolution.

Conclusion: Although there is no specific treatment, Pediatricians should considerable pharochalasis in the differential diagnosis of infants with recurrent eyelid edema and refer them to an ophthalmologist. Without the correct management and diagnosis, eyelid tissue can become deformed in the medium to long term. In addition to the importance of ruling out associated systemic pathology, many of these children are subjected to multiple ineffective treatments.

Keywords: Blepharochalasis; Non-pruritic eyelid edema; Dermatochalasis; Ptosis; Amblyopia.

Introduction

Blepharochalasis is an infrequent palpebral sickness. Until 2009 only 67 cases had been reported. It is characterized by recurrent episodes of painless and nonpruritic eyelid edema that begins in childhood and ends up producing atrophy and periocular skin depigmentation, tissue relaxation and ptosis in adult age [1-3]. Other manifestations include lower eyelid retraction, pseudo-epicanthal folds, and fat prolapse of orbital and lacrimal gland [2]. According to recent studies blepharochalasis is more frequent in women and usually begins in the adolescence, at an average age of 10 years old [2,3]. This condition can be bilateral or unilateral, the latter being less frequent and of later onset [3]. Only one case has been reported of a patient with both, unilateral and bilateral outbreaks [1]. Episodes last between hours and days, becoming less frequent as the years go by. Although the majority of patients enter at quiescent phase in adult age, there has been one case reported with persistent episodes at the age of 40. In the cases described, the condition is limited to the ocular level. Only one case has been associated with renal, cardiac, and skeletal anomalies [4].
The diagnosis of blepharochalasis is clinical. Image tests are unnecessary, as they do not demonstrate anomalous results, and palpebral biopsy does not demonstrate specific results, frequently showing elastic fibers decrease as the episodes progress [1,2]. Regarding pharmacological treatment, there is poor response to oral or local antihistamines and cortisone [5]. Once ptosis and dermatochalasis appear the only effective treatment is surgery. A quiescent period of 6 months to 2 years is recommended before any intervention is performed [1,4,6,7].

We present a case of progressive blepharochalasis that started episodes at the age of 5 years, long before the average age of presentation, and with both unilateral and bilateral outbreaks.

**Case presentation**

Eleven-year-old male with no systemic alterations is referred to our service for recurrent outbreaks of eyelid edema with secondary palpebral hyperlaxity of years of evolution. His mother referred self-limited episodes of unilateral and bilateral eyelid edema since the age of 5 years without prodromal signs. The patient had been treated with multiple antibiotics and topical and oral antihistamines with no results. Outbreaks had only decreased in frequency and severity over the years. Both upper eyelids became erythematous, edematous and touch sensitive (Figure 1a and 1b). There was no coexisting facial or lingual condition and no visual changes occurred during these episodes. The mother denied any traumatic or family history of interest. No arachnodactyly, marfanoid phenotype, scoliosis or double lip were observed during the dermatological evaluation. There was no thyroid goiter and no lax skin in other locations.

Uncorrected visual acuity was 1.00 in both eyes. Extrinsic ocular motility and binocular vision were preserved. Palpebral examination showed severe dermatochalasis with thin atrophic skin and visualization of subcutaneous telangiectatic vessels (Figure 2). The rest of the ophthalmological examination was within the normal range. At the present time the patient is 12 years old. The last episode of eyelid edema was in February 2021, seven years after the condition onset and with a quiescent period of 1.5 years now. In the last physical examination, he had a receding secondary eyelid fold and dermatochalasis. Twelve years old.

**Discussion**

Blepharochalasis is an infrequent self-limited illness, with onset in late childhood usually at the age of 10. Without the correct diagnosis and treatment, this condition can deform the palpebral tissue in the medium to long term.

Although blepharochalasis etiology is unknown, stress, fever and upper respiratory tract infections have been proposed as possible triggers [1,5]. Several authors believe that Ig A antibodies play a fundamental autoimmune role in the destruction of elastic fibers [8,9]. Moreover, recent studies highlight the role of matrix metalloproteinases and their relationship to collagen and elastin degradation, postulating that Doxycycline could be an effective treatment by inhibiting (MPM) metalloproteinases [5,9-12].

Two cases treated with a descending regimen of Doxycycline 50 mg for 8 and 10 months have been reported, reducing in both cases the frequency of exacerbations [12].

In most patients, as the average age of presentation is 10 years-old, Doxycycline could be the first option. But not in early debuts, such as our case, as Doxycycline is contraindicated in children under 8 years of age, because it affects correct osseous and dental development.

There are no well-defined protocols to treat inflammatory attacks. The first-choice treatment in severe outbreaks during early childhood is topical and oral prednisone. However, only a few reports based on case series suggest only some improvement. Considering the potential side effects of steroids, this treatment is not accepted as general rule for treating blepharochalasis [5,10]. Although oral treatment with antihistamines and antibiotics has not proven to be effective, it is not unusual that these treatments are prescribed, as in the case of our patient, due to an incorrect diagnosis. In mild outbreaks or quiescent periods, treatment should be conservative. Even if the patient presents ptosis or dermatochalasis secondary to the illness, in most of the reported cases, surgery is not considered...
Conception of the work AND/OR Design of the work

The main differential diagnosis in a child with recurrent eyelid edema is recurrent angioedema. Angioedema occurs secondary to an allergic etiology and is characterized, in addition to eyelid edema, by swelling of the extremities, abdominal pain, urticaria and pruritus [13]. Unlike blepharochalasis, these patients respond well to oral antihistamines and oral corticosteroids. The differential diagnosis should also include hereditary angioedema where localized edema without pruritus can also appear on all four extremities, pharynx, glottis or at the periocular level. It is differentiated from blepharochalasis by examining the levels of inhibitor of the first component of human complement (CI-INH), which are decreased in hereditary angioedema [14]. Another much less likely diagnosis due to its low incidence is Ascher Syndrome, where in addition to the ocular symptoms we would find phenotypic alterations, double upper lip and nontoxic goiter [15] (Table 1).

Few cases of outbreaks at such a young age as our patient have been reported. When this happens, we could expect an early cease of the outbreaks [1,2,7,16]. In both retrospective studies by Koursh et al. and Zhou J et al. [2,3], patients are clinically classified as unilateral or bilateral, with no patient having both uni and bilateral episodes. The case reported by Dworak et al. [1]. Calls our attention as also being a very early debut whose condition began with both uni and bilateral out breaks.

**Conclusion**

In the case of pediatric patients with history of bilateral, unilateral or mixta, non-pruritic, recurring palpebral edema with poor response to oral and topical antihistamines we must suspect blepharochalasis. During exacerbations, conservative treatment in combination with cool-pack bathing for symptomatic control is the choice. Only some cases could be tributaries to oral cortisone. For the adolescent period, we could consider Doxycycline as an alternative to Prednisone and as a maintenance therapy. Treating outbreaks conveniently and limiting their duration to a minimum will prevent palpebral tissue changes in adulthood.

Although exacerbations can be managed on an outpatient level, it is important to refer to an Ophthalmologist when dermatochalasis or ptosis appear due to the risk of amblyopia. Pediatricians should be aware of the differential diagnosis of non-pruritic palpebral edema in childhood in order to avoid unnecessary treatments and inappropriate use of resources.

Declarations of interest: none. This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Author’s contributions**

Loscos, I

GROUP 1: Conception of the work AND/OR Design of the work AND/OR Acquisition of data AND/OR Analysis of data AND/OR Interpretation of data.

GROUP 2: Drafting the work AND/OR Revising the work critically for important intellectual content.

GROUP 3: Final approval of the version to be published.

GROUP 4: Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Luna, S

GROUP 1: Conception of the work AND/OR Design of the work.

GROUP 2: Revising the work critically for important intellectual content.

GROUP 3: Final approval of the version to be published.

GROUP 4: Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Roselló, N

GROUP 1: Conception of the work AND/OR Design of the work.

GROUP 2: Revising the work critically for important intellectual content.

GROUP 3: Final approval of the version to be published.

GROUP 4: Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.
### Table 1: Differential diagnosis of palpebral edema in childhood.

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Symptomatology</th>
<th>Biopsy</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Blepharochalasis</td>
<td>Localized, recurrent, painless, non-pruritic palpebral edema</td>
<td>Decrease of elastic fibers as the episode progresses</td>
<td>Prednisone, Doxycycline, Oral antibiotics, Oral antihistaminic, Oral antibiotics, Surgical</td>
</tr>
<tr>
<td>Stye</td>
<td>Localized area of redness, tenderness and swelling near the lid margin.</td>
<td>Increase in perivascular mononuclear infiltrate</td>
<td>Oral antibiotics, Topic antibiotic and cortisone, Oral antibiotics, Prednisone, Doxycycline</td>
</tr>
<tr>
<td>Acute angioedema</td>
<td>Eyelid edema, tenderness, erythema and induration. Can present systemic symptoms</td>
<td>Decreased levels of C1-INH</td>
<td>Surgical, Oral antihistaminic, Oral antihistaminic, Oral antihistaminic, Surgical</td>
</tr>
</tbody>
</table>

### References


