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Blepharochalasis at early age: A case report

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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 pharochalasisin 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 bilateraleyelid 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 lipwere observed during the dermatological evaluation. There was no thyroid goiter and no lax skin in other locations.

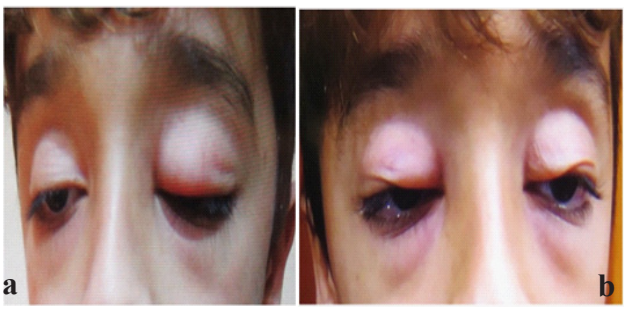

Figure 1: (a): Boy with unilateral outbreak. Nine years old. **(b):** Boy with bilateral outbreak. Nine years old.

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 worsening due to the last out breaks (Figure 3). We are waiting to indicate surgery according to evolution during this quiescent period.

Discussion

lepharochalasis 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.

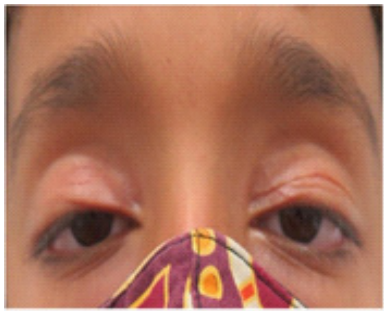

Figure 2: Boy on first visit showing dermatochalasia. Eleven years old.

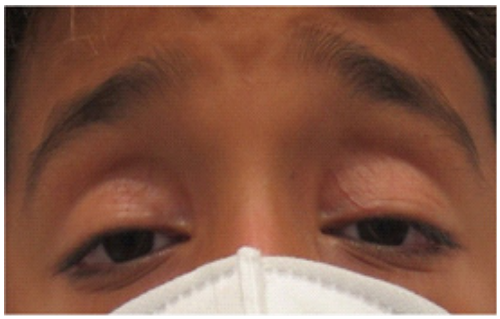

Figure 3: Boy on his last visit with signs of receding palpebral secondary folds and dermatochalasia. Twelve years old.

Although blepharochalasis etiology is unknown, stress, fever and upper respiratory tract infections have been proposed as possible triggers [1,5]. Several authors believe that lg 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

until 6 to 12 months after the last outbreak [1,4,6,7]. *Zhou J et al* [3] conducted a retrospective study with 93 cases, analysing the risk of surgical reintervention according to the quiescent period length before undergoing surgery. They concluded that after 2 years of stability the risk of reintervention was lower.

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. Itis 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.

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Author's contributions

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

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

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Table 1: Differential diagnosis of palpebral edema in childhood.

	Etiology	Symptomatology	Biopsy	Treatment
Blepharochalasis	Hereditary?? Triggers? IgA?	Localized, recurrent, painless, non-pruritic palpebral edema	Decrease of elastic fibers as the episode progress	Prednisone Doxycycline
Stye	Staphylococcal infection of Zeiss and Moll	Localized area of redness, tenderness and swelling near the lid margin. Localized pain	---	Topic antibiotic and cortisone
Preseptal cellulitis	Infection (Common bacteria: *Staphylococcus epidermis, Staphylococcus aureus, Streptococcus pyogenes*)	Eyelidedema, tenderness, erythema or induration. Can present systemic symptoms. Pain	--- Leukocytosis in analytics	Oral antibiotics
Hereditary angioedema	Hereditary	Localized non-pruritic edema in different parts of the body Pain	Decreased levels of CI-INH	Oral antihistaminic
Acute angioedema	Allergic	Localized (bilateral or unilateral) facial swelling and pruritic edema	Perivascular mononuclear infiltrate	Oral antihistaminic
Ascher syndrome	Autosomal dominant	Classic triad: Non-toxic goiter Double lips Dermatochalasia	Lacrimal gland hypertrophy	Surgical

References

1. Dworak DP, Patel SA, Thompson LS. An Unusual Case of Blepharochalasis. *J Ophthalmic Vis Res.* 2017; 12: 342-344.
2. Koursh DM, Modjtahedi SP, Selva D, Leibovitch I. The blepharochalasis syndrome. *SurvOphthalmol.* 2009; 54: 235-244.
3. Zhou J, Ding J, Li D. Blepharochalasis: clinical and epidemiological characteristics, surgical strategy and prognosis-- a retrospective cohort study with 93 cases. *BMC Ophthalmol*. 2021; 21: 313.
4. Ghose S, Kalra BR, Dayal Y. Blepharochalasis with multiple system involvement. *Br J Ophthalmol.* 1984; 68: 529-532.
5. Browning RJ, Sanchez AT, Mullins S, Sheehan DJ, Davis LS, et al. Blepharochalasis: Something to cryabout. *J Cutan Pathol.* 2017; 44: 279-282
6. Nagano H, Domoto T, Azuma R, Kiyosawa T. Step wise surgery with variable adjustments for severe blepharochalasis with multiple chemical sensitivity: A case report. *Case Reports Plast Surg Hand Surg*. 2020; 7: 20-22.
7. Wang G, Li C, Gao T. Blepharochalasis: A rare condition misdiagnosed as recurrent angioedema. *Arch Dermatol*. 2009; 145: 498-499.
8. Motegi S, Uchiyama A, Yamada K, Ogino S, Takeuchi Y, et al. Blepharochalasis: Possibly associated with matrix metalloproteinases. *J Dermatol*. 2014; 41: 536-538.
9. Paul M, Geller L, Nowak-Węgrzyn A. Blepharochalasis: A rare cause of eye swelling. *Ann Allergy Asthma Immunol.* 2017; 119: 402-407.
10. Ortiz-Perez S, Patel BC. Blepharochalasis Syndrome. 2022 Aug 1. In: StatPearls [Internet]. TreasureIsland (FL): Stat Pearls Publish-

ing; 2022 Jan–. PMID: 32809455.

11. Xin Y, Xu XL, Li Y, Ding JW, Li DM, et al. *Zhonghua Yan Ke ZaZhi*. 2020; 56: 205-210.

12. Karaconji T, Skippen B, Di Girolamo N, Taylor SF, Francis IC, Coroneo MT, et al. Doxycycline for treatment of blepharochalasis via inhibition of matrixmetalloproteinases. *Ophthalmic Plast Reconstr Surg*. 2012; 28: e76-e78.

13. Tachdjian R, Johnston DT. Angioedema: Differential diagnosis and acute management. *Post grad Med*. 2021; 133: 765-770.

14. Mansour E, Veronez CL, Craig T, Grumach AS. Hereditary angioedema in children and adolescents. *Allergol Immunopathol (Madr)*. 2022; 50: 1-6.

15. Zhao ZL, Wang SM, Shao CY, Fu Y. Aschersyndrome: A rare case of blepharochalasis combined with double lip and Hashimoto's thyroiditis. Int *J Ophthalmol*. 2019; 12: 1044-1046.

16. Brar BK, Puri N. Blepharochalasis-A rare entity. *Dermatol Online J.* 2008; 14: 8.