Do you know this syndrome? Ascher´s syndrome: clinical findings of little known triad*

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CASE REPORT

A 58-year-old female patient, with a background of hypertension and rheumatoid arthritis, complained of long standing increase of volume of the upper lip. On physical examination, we could observe normochromic nodular lip projections, with a smooth surface, partially covering the maxillary central incisors and fully covering the maxillary lateral incisors (Figure 1). On palpation, both projections were soft, resembling physiological labial mucosa. Bilateral, symmetrical and painless edema of the upper eyelids was also observed; that was said to be chronic and recurrent by the patient (Figure 2). Facing the clinical the clinical findings, the diagnosis of Ascher syndrome was suggested. Thyroid, liver and renal function, as well as relevant autoimmune screening, and thyroid ultrasound with doppler were all normal.

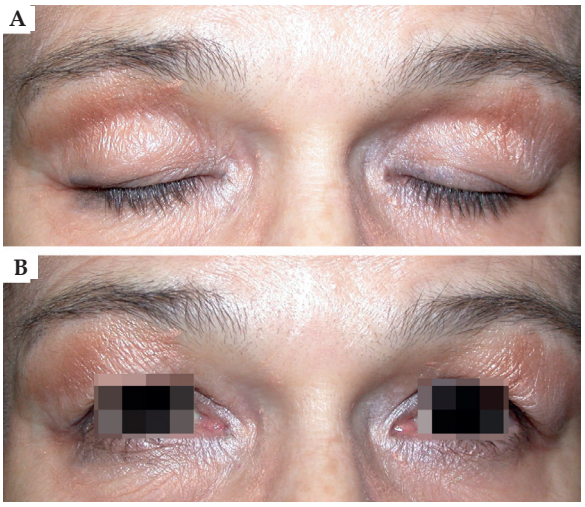

Figure 2: Blepharochalasis. **A.** Physical examination of the patient with her eyes closed, showing upper eyelid edema, with marked projection on the lateral corner of the eye. **B.** Patient with her eyes open: it is still possible to observe the lax projection of the upper eyelids, symmetrically

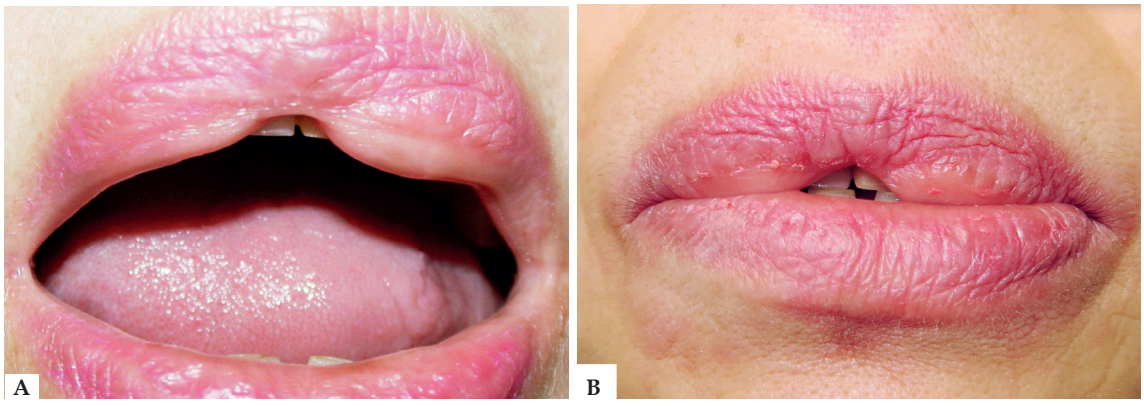

Figure 1: Double lip. **A.** Clinical examination showing two well defined, normochromic projections, with a smooth surface, centrally localized in the upper labial mucosa, better seen when the patient keeps her mouth open. **B.** It is still possible to observe the labial projections with the mouth closed, overlying the anterior teeth

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DISCUSSION

Ascher's syndrome is characterized by the triad: double lip deformity, blepharochalasis and non-toxic goiter.[1] Described in 1920, it is considered to be a rare, benign condition with unknown etiology, affecting all races and both sexes equally.[2]

It is usually manifested before 20 years of age and is uncommon during childhood.[3] Hormonal dysfunctions and autosomal dominant inheritance have been implied as possible causes.[4]

The double lip deformity represents hyperplasia of the labial mucosa and can present as a congenital or acquired type. In both types, the most common presentation affects the upper lip bilaterally, and occasionally both the upper and lower lip can be affected.[5] Recurrent traumas and bad habits such as sucking the lips are associated to the acquired type. The congenital type happens due to the persistence of the sulcus between the internal (pars vilosa) and external (pars glabra) portions of the lip, that would otherwise disappear on the third month of gestation.[6] The deformity, barely seen at rest, becomes evident when the patient talks or smiles because of the lip retraction that positions the excess mucosa in front of the anterior teeth, giving an aspect similar to the cupid's bow.[7]

Blepharochalasis is present in approximately 80% of the cases and represents atrophy and progressive skin laxity with subsequent orbital fat herniation. It predominantly involves the upper eyelids, that become redundant, discolored, anetoderma-like, giving a wrinkled appearance. It is clinically manifested as recurrent painless edema of the eyelids, with no erythema, progressing to fat ptosis and eventually prolapse, the lacrimal glands included.[2,5]

Although it was described as a part of the triad, non-toxic goiter is only present in 10-50% of cases, and it is not essential for the diagnosis.[5] In the presented case, only labial and eyelid increase was observed, reinforcing the infrequency of the thyroid involvement.

Differential diagnoses should include Melkersson-Rosenthal syndrome, hereditary angioedema, acquired cutis laxa, among others.[8]

Surgery is the treatment of choice for both the labial and eyelid involvement, whenever the deformity interferes with the appearance or function.[8-10] In our case, we proposed a surgical approach, but the patient opted to continue with clinical follow up only. ❑

Abstract: Ascher's syndrome consists of double lip, blepharochalasis and sometimes non-toxic thyroid enlargement. It is a rare, benign, rarely reported, and sometimes misdiagnosed condition that most often affects patients under 20 years old. The etiology remains unknown, although factors such as trauma, hormonal dysfunction and heredity have been suggested. Treatment based on surgical intervention is indicated to improve aesthetics or function. The presented case shows a patient diagnosed with Ascher's syndrome based on clinical findings such as double lip and blepharochalasis.
Keywords: Goiter; Lip; Eyelids

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