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Ascher Syndrome: A Rare Syndrome

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Abstract
Ascher syndrome (Laffer-Ascher syndrome) is a rare disorder of unknown etiology, with a prevalence <1 case/million populations worldwide. The classical clinical triad is blepharochalasis, double lip and non-toxic thyroid enlargement. The syndrome can occur without nontoxic enlargement of the thyroid gland and is then considered as the forme fruste or incomplete form.

Introduction

Ascher syndrome (Laffer-Ascher syndrome) was first described in 1920 by an ophthalmologist K W Ascher.[1] It is a very rare disorder with prevalence <1 case/million population, with <100 cases reported in the literature worldwide, of which very few are reported from India.[2] It usually presents as a triad of blepharochalasis, double lip and non-toxic thyroid enlargement.[3,4] The syndrome can occur without nontoxic enlargement of the thyroid gland and is then considered as the forme fruste or incomplete form. In 10% of cases, non-toxic idiopathic thyroid enlargement also occurs completing the typical triad.[5]

Case report

A 23-year-old female presented with a history of recurrent swelling of both upper eyelids since last 7 years. The swelling was abrupt in onset and gradually subsided within 3-4 days. About 3-4 months later, she noticed drooping of her upper eyelids. Followed by this, since last 5-6 years she noticed progressive increase in the size of her upper lip. There was no response to treatment with antihistamines. There was no relevant personal history of any major medical or surgical illness in the past nor was there any familial history of similar illness.

Clinical examination showed lax skin with ptosis of both the upper eyelids. The upper lip was swollen and enlarged with a shallow transverse sulcus between the mucosa and the skin of the lip giving it an appearance of double lip. When the patient smiled, a deformity with a fold of excess tissue on the mucosal aspect of the upper lip was remarkably prominent. (Fig. 1) There was no clinical evidence of thyroid enlargement. Her

thyroid gland was normal in size and consistency on ultrasonic examination and thyroid function tests were normal. Ophthalmological examination was normal too.

The patient was referred to plastic surgery, but the patient refused for any surgical intervention.

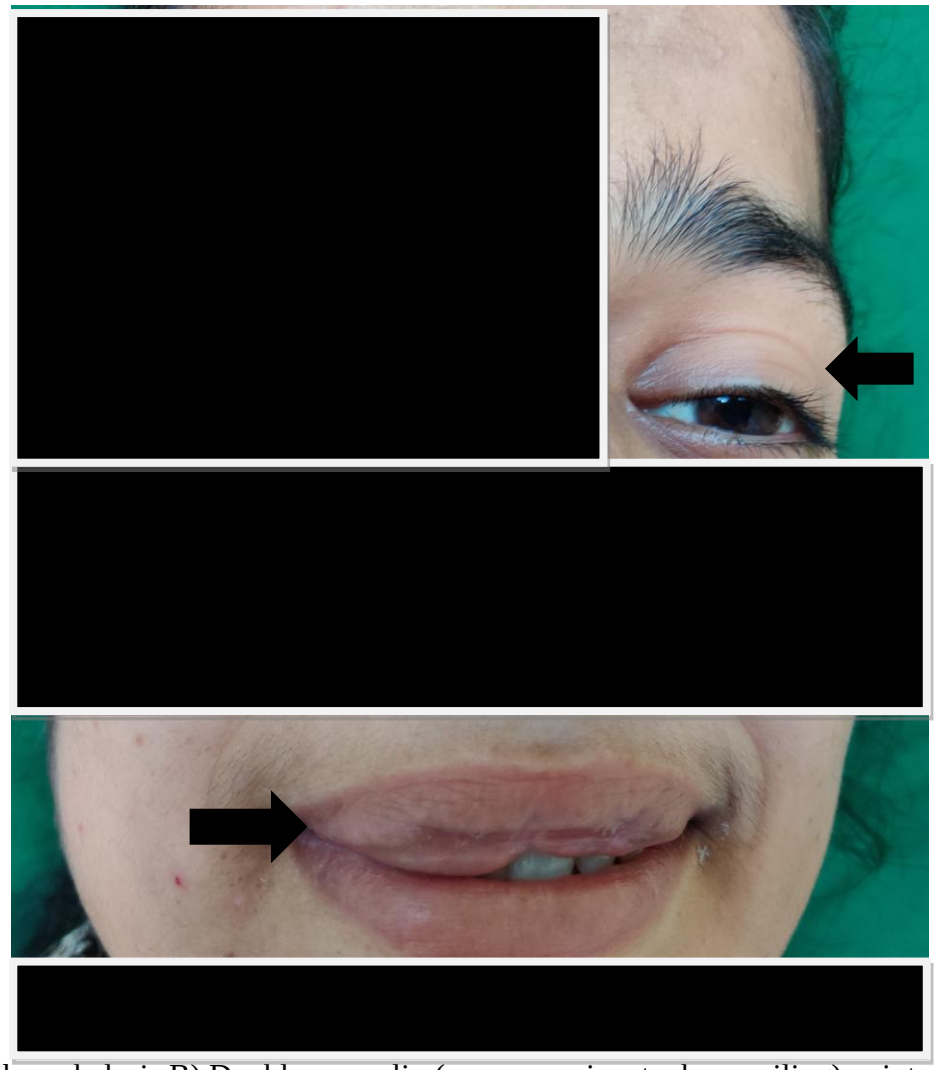

Figure 1: A) blepharochalasis B) Double upper lip (more prominent when smiling) pointed by black arrows.

Discussion

Ascher syndrome is a condition of unknown etiology, most cases are sporadic, but familial cases suggestive of autosomal dominant inheritance have been reported[3] It has no racial, gender or geographical predilection. Ascher syndrome presents within the first 20 years of life, usually seen at puberty. It is a combination of blepharochalasis with progressive enlargement of the upper lip due to hypertrophy and inflammation of the labial salivary glands. There are multiple attacks of painless edema of upper eyelids which may cause weakness of the orbital septum, ptosis and these episodes often result in eyelid skin redundancy. Diagnosis of Ascher syndrome is purely clinical. Thyroid enlargement usually presents several years after the initial eyelid and lip edema and is seen in 10%–50% of patients. However, is not essential for the diagnosis. The differential diagnosis includes hereditary angioedema, orofacial granulomatosis, Melkersson - Rosenthal syndrome, cutis laxa and sarcoidosis.[6]

No specific pharmacological treatment exists for the syndrome. Cosmetic surgery is the treatment of choice when the condition interferes with

vision, speech, and chewing. Surgical treatment to excise the excess tissue of eyelids or lips can be carried out.

Conclusion

The scientific literature describes less than 100 cases of Ascher's syndrome. Early diagnosis of Ascher syndrome prevents treatment delays. An adequate aesthetic correction is possible enough to bring the patient back to normal and avoid an impact on the psychological state of the patient, with a minimum risk of recurrence. The importance of recognizing the triad must alert a dermatologist to propose a diagnosis of the aforementioned syndrome so as to avoid unnecessary investigations and inappropriate surgery.

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Conflicts of interest

There are no conflicts of interest.

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