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Case Report

Ascher syndrome: Review of literature and case report

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ABSTRACT

A 13 year old girl presented with aesthetic deformity of upper lip since birth. She also presented with eyelid swelling on and off for 11 months. She was diagnosed to be a rare case of Ascher syndrome. Ascher syndrome commonly presents with double lip and blepharochalasis, sometimes associated with goitre. The deformity of her double upper lip was corrected by appropriate surgery. Because her blepharochalasis is in active stage now, she is under periodic follow up for appropriate intervention. This article describes the management of the patient and brief overview of the syndrome. Ascher syndrome is often missed or misdiagnosed commonly.

KEY WORDS

Ascher syndrome; blepharochalasis; double lip

INTRODUCTION

Ascher syndrome was first described in 1920 by an ophthalmologist. [1] This syndrome presents as blepharochalasis, double lip and nontoxic thyroid enlargement. The thyroid enlargement is not present in all cases of this syndrome. The syndrome is often undiagnosed because of its rarity.

CASE REPORT

A 13-year-old girl was presented with aesthetic deformity of the upper lip [Figure 1]. Patient's mother said that the deformity was present since birth. She was going to school and often teased by her friends. Hence she was depressed. There was no history of lip trauma. She did not have lip-sucking habit. She also gave history of recurrent painless swelling of both upper eyelids, each episode lasting several days, for past 11 months. She did not have family history of similar compliants. On examination, her upper lip was bulky

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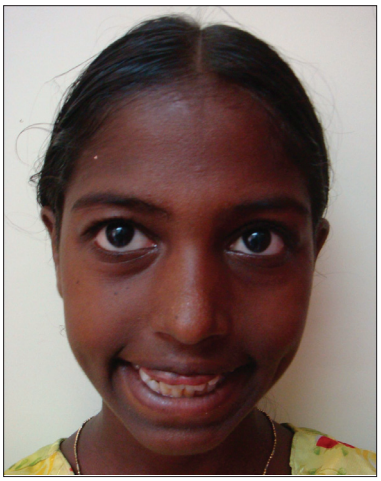

Figure 1: Ascher syndrome: Blepharochalasis and double upper lip

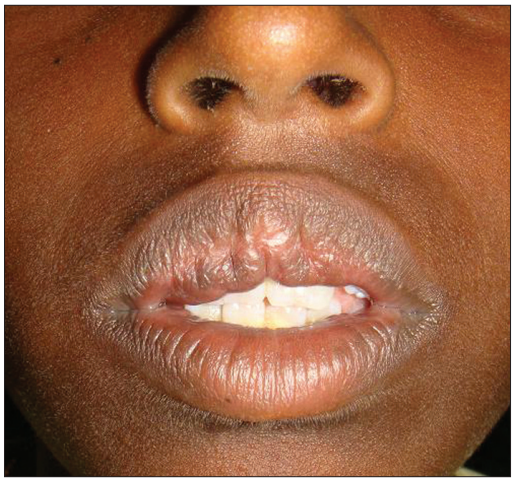

Figure 2: Preop repose: Bulky upper lip

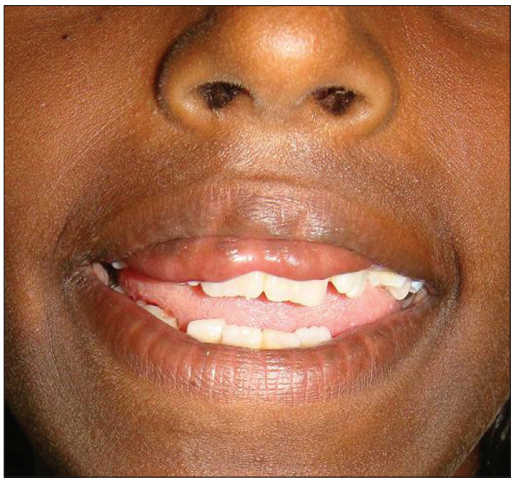

Figure 3: Preop smiling: Double upper lip

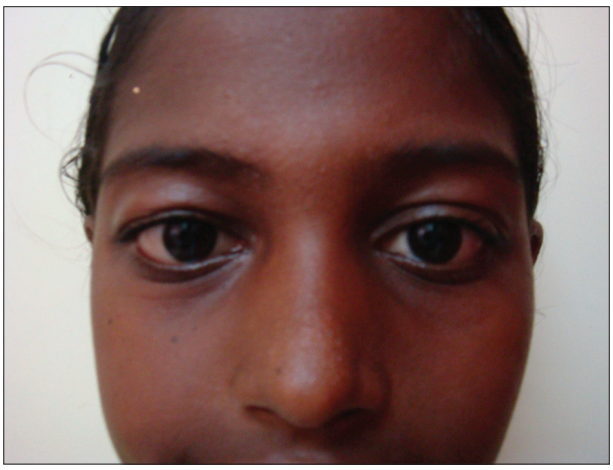

Figure 4: Blepharochalasis: Prominent on right side and excess skin more laterally

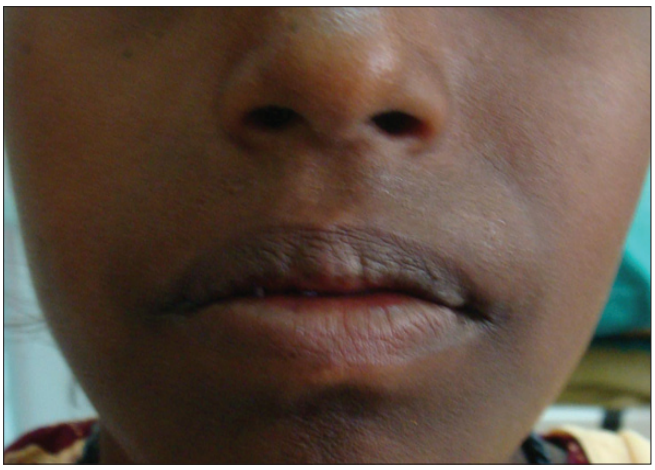

Figure 5: Postop repose: Bulk reduced

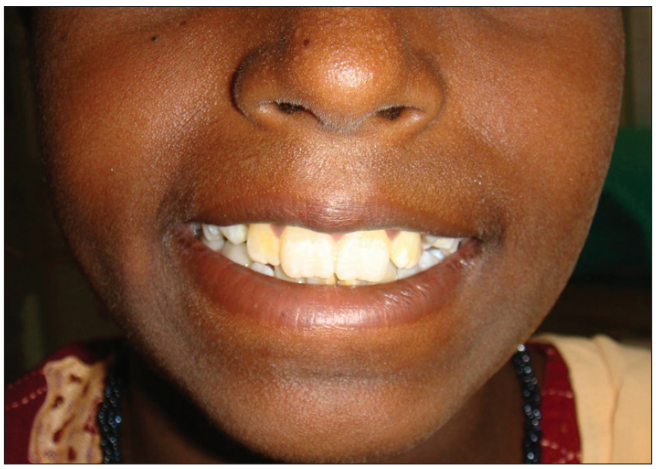

Figure 6: Postop smiling: Lip symmetry maintained

in repose [Figure 2]. When she smiled, a horizontal sulcus appeared in the upper lip making it appear as if she had two upper lips [Figure 3]. The buccal part of double upper lip measures 4.3 cm by 1.6 cm. It was soft in consistency and non-compressible. The lower lip was normal. Skin over both upper eyelids was thin, boggy and flaccid. Blepharochalasis was present more on right side. The excess skin was consistently more prominent laterally than medially [Figure 4]. Her lower eyelid and visual acuity were normal. She did not have goiter. She also did not have any systemic ailment. There was no laxity of skin elsewhere on the body. Patient was planned for surgical lip correction. Preoperatively horizontal elliptical marking was made encircling the buccal part of double upper lip. Small extension of elliptical marking continued up to commissure on either side. Under anaesthesia, marked buccal part of double upper lip tissue was excised. Only mucosa and submucosa was removed without damaging underlying muscle. The maxillary labial frenum was left intact. The wound closed with 4-0 absorbable interrupted sutures.

Her lip wound healed without complications and lip symmetry maintained [Figures 5 and 6]. She recovered from depression.

She has been under follow-up for the past 7 months. There is no development of goiter. Surgery for blepharochalasis deferred for her as the disease is in active phase.

DISCUSSION

Ascher syndrome presents as combination of blepharochalasis, double lip and non-toxic thyroid enlargement.[1] Ascher syndrome, like other syndromes, rarely has all components together at presentation. Enlargement of the thyroid is present in only 10-50% cases of Ascher syndrome.[2] Aetiology of the syndrome is unknown. Most cases are sporadic, though rarely family history of Ascher has been reported. The syndrome is often undiagnosed by general practitioner because of its rarity.

Double lip usually affects the upper lip and produce unpleasant appearance on smiling. It can rarely involve lower lip or both lips. Double lip occurs due to lip maldevelopment. The lip normally develops during the second or third month of gestation from the pars glabrosa (outer cutaneous zone) and the pars villosa (inner mucosal zone) with disappearance of horizontal sulcus between them. Persistence of horizontal sulcus with hypertrophy of pars villosa leads to double upper lip. The treatment for double lip is surgical excision. Surgical excision of double lip indicated when it interferes with chewing, speaking or for aesthetic reasons.[3-6]

Blepharochalasis is present in more than 80% cases of this syndrome. It starts at puberty and usually both upper eyelids are involved. Pathologically it is a form of localized angioedema with decrease in dermal elastin. Blepharachalosis is characterized by three stages.[7] First stage or oedema stage presents with intermittent painless swelling of lids. Second stage or atonic ptosis stage presents with ptosis due to dehiscence of levator aponeurosis or lax thin skin falling as redundant folds over lid margin. Third stage or ptosis adiposa presents with medial fat pad atrophy, orbital fat prolapse and lacrimal gland prolapse. Surgery should be deferred for at least 1 year from the previous attack of eyelid oedema.[8] Indications for surgery are visual acuity disturbance or ocular complications.

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