Nonsyndromic Maxillary Double Lip: A Case Report and Review of Literature

ABSTRACT

Double lip is an uncommon facial anomaly which may be either congenital or acquired. Most often it is nonsyndromic, however it is also known to present as syndrome associated with other systemic disorders. Besides treating the orofacial defect, it is essential to recognize and diagnose the possible systemic disorders with this anomaly. This manuscript reports on management of a case of nonsyndromic double lip with a relevant review of literature.


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INTRODUCTION

Double lip is a rare deformity which can be either acquired or congenital. Acquired double lip may affect upper or lower lip, which is usually secondary to trauma. Congenital double lip involves the upper lip more frequently than the lower lip. It is known to occur as an isolated entity or as part of a syndrome. The syndrome most frequently associated with double lip is Ascher’s syndrome. It is a triad comprising of double upper lip, blepharochalasis and nontoxic thyroid goitre.1,2 The case presented here was eventually diagnosed of having a nonsyndromic congenital double upper lip without any other features of the Ascher’s syndrome. Double lip is a benign condition and the most common reason for patients seeking treatment is due to the facial disfigurement. This manuscript presents a case of congenital double upper lip and relevant review of literature. An online literature search showed reports of 29 such cases from various parts of the world, of which 8 were associated with Ascher’s syndrome and one report claimed an association with a possible new syndrome. The focus of this manuscript is on the diagnosis and the surgical technique in treating double lip.

CASE PRESENTATION

A 15-year-old male patient presented with complaint of deformed upper lip. The main motive of the patient for seeking treatment was social embarrassment caused by his deformity that accentuates when he smiles. There was no history of trauma/surgical procedure to the upper lip. On interviewing his parents, the boy was known to have increased thickening of his upper lips at birth. They assumed it to fade with the boy’s growth and had never consulted a doctor before, until now. It was a congenital deformity and there was no relevant family history. Systemic examination showed no significant clinical findings.

The upper lip deformity consisted of an excess fold of mucosal tissue giving an appearance of characteristic ‘double lip’. At rest, the deformity was limited to mild eversion of the upper lip (Fig. 1A). On smiling, it was very apparent causing significant unesthetic appearance (Fig. 1B). The excess fold of tissue during smile measured about 45 mm in length and 6 mm in width covering nearly half the labial surfaces of the maxillary incisors. The excess tissue had a notch in the midline near the frenulum attachment.

On thorough clinical examination, the patient did not appear to have any other components of Ascher’s syndrome. He had no thyroid enlargement and his thyroid function tests (TFT) were found to be within normal limits. There were no extraoral/intraoral abnormalities. A clinical diagnosis of congenital maxillary double lip was made. It was planned to excise the excess mucosal tissue under local anesthesia. The patient was explained about the surgical procedure needed to correct his deformity. Consent for the surgery was obtained after possible complications, such as swelling, paresthesia of bilateral infraorbital nerve causing temporary/permanent numbness of his upper lips, wound infection and dehiscence were explained to the patient. (This is not

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a clinical research, hence ethical committee approval is not required).

**Surgical Procedure**

Bilateral infraorbital nerve blocks were administered. 3 mL of 1:3,000,000 adrenalinized saline was infiltrated into the upper lip around the excessive mucosal tissue. An elliptical incision was given around the tissue to be excised to the depth of orbicularis oris muscle (Fig. 2A). After excision the wound was sutured with 3-0 black silk (Fig. 2B). The healing was uneventful and the patient was satisfied with the esthetic result (Fig. 3). The excised tissue was sent for biopsy. Histopathology report showed normal mucosal tissue with numerous minor salivary with no pathology.

**DISCUSSION**

The diagnosis of double lip is purely clinical. Histopathology may be done for academic value. A double lip in most of the cases have only cosmetic or social implications. An interesting feature of a double lip is its association with other syndromic abnormalities, which the patient may not be aware. It is essential for the treating clinician to have knowledge of various other abnormalities associated with double lip. Congenital double lip can occur in isolation or part of a syndrome. Paramar and Muranjan reported on a possible new syndrome comprising of double upper and lower lip, unilateral ptosis, hypertelorism, blepharochalasis, broad nose and bilateral 3rd finger clinodactyly. Costa-Hanemann et al described double upper lip associated with hemangiomas. There are reports of double upper lip associated with bifid uvula, chelitis glandularis and cleft palate. The etiology of double lip is unknown and its occurrence is sporadic. A recent report by Palma and Taub has suggested a male predilection of 7:1. The upper lip develops by fusion of maxillary, medial and lateral nasal processes. The vermilion border of the upper lip demarcates the cutaneous and the mucosal portion of the lip. The lip vermilion consists of a dry portion (Pars glabrosa) and a wet portion (Pars Villosa). Pars glabrosa is smooth and similar to skin. Pars villosa is similar to mucosa. Double lip is thought to arise in 2nd or 3rd trimester of gestation due to persistence of an exaggerated sulcus between the pars glabrosa and pars villosa. The treatment for double lip is surgical excision of the mucosal and submucosal tissue. Various incision techniques have been reported which include transverse elliptical incision, double ellipse incision combined with ‘Z’ plasty and ‘W’ plasty. The transverse elliptical incision is common and a simple surgical technique, which was used to treat the patient reported in this manuscript. However, care should be taken not to remove excessive of the vermilion. This could result in insufficient exposure of upper lip vermilion during smile.

The association of double lip with other abnormalities is well established in Ascher’s syndrome. It is a triad comprising of double upper lip, blepharochalasis and non-toxic thyroid enlargement. It was first described by an ophthalmologist, Ascher K. W. in 1920. Blepharochalasis was first described by Beer in 1817 and the term was given by Fusch in 1896. It is characterized by recurrent episode of painless edema of upper eye-lids. After repeated episodes, the upper eye-lids becomes wrinkled, thinned and redundant resulting in ptosis. In severe cases, there is narrowing of the palpebral fissure causing...
visual disturbances. Benedict was first to describe two stages of blepharochalasis. An early stage characterized by eyelid edema that does not pit on pressure and a late stage characterized by atrophy of the lid tissue resulting in ptosis. Treatment of blepharochalasis is surgical. Redundant skin should be excised, prolapsed fat should be removed and eye-lid crease should be recreated. The patient presented was examined by an ophthalmologist and had no edema of the upper eye-lids. Thyroid enlargement are late manifestation of Ascher's syndrome. This patient was asymptomatic and the TFT showed euthyroid values. Long-term follow-up is essential to assess on development of thyroid dysfunction.

CONCLUSION

Maxillary double lip can occasionally be associated with syndromes. It is imperative for the treating dentist to overlook beyond the facial deformity in management of such patients.

REFERENCES