Case Report

Macrocheilia - A Rare Clinical Entity!!

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ABSTRACT

Double lip or "macrocheilia," is a rare anomaly which consists of a fold of excess or redundant hypertrophic tissue on the mucosal side of the lip. The upper lip is affected more commonly than the lower lip. The congenital double lip is believed to be present at birth and becomes more prominent after eruption of teeth and results in compromised esthetics and interference with speech and mastication. Simple surgical excision produces good esthetic and functional results. Here we report a case of non-syndromic congenital maxillary double lip.

Key words: Double lip, macrochelia, Congenitol anomoly.

Introduction:

A double lip is a rare anomaly characterized by a horizontal fold of redundant mucosal tissue that is situated proximal to the vermilion border. It may be either congenital or acquired and has no gender or race predilection.[1] It occurs most often in the upper lip, although both upper and lower lips are occasionally involved. The deformity may be present at birth and become more prominent as the patient grows.[1,2] Double lip may interfere with speech, mastication and esthetics.[3]

Double lip is caused by excessive areolar tissue and non-inflammatory labial mucosa gland hyperplasia of the pars villosa.[4] During smiling, the lip is retracted and the mucosa is positioned over the maxillary teeth, resulting in a “Cupid's bow” appearance. Double lip may require surgical correction for esthetic reason.[5]Here we present a case of maxillary double lip.

Case Report

A 44-year-old man presented to the department of oral medicine and radiology complaining of a lip defect. The lip defect, he stated, was present since childhood. He denied any complaints other than cosmetic ones. Oral examination revealed thickened folds of redundant mucosal tissue on the inner surface of the upper lip with central constriction giving a Cupid's bow appearance (Figure1). The mucosal tissue was smooth, with no palpable masses or surface changes. The patient denied any other significant medical history. A provisional diagnosis of double lip was given.

Bilateral infraorbital blocks and local anesthesia were administered. Elliptical excisions were made over each side of redundant tissue, combined with a central Z-plasty to release the constricting band and the defects were closed in layers (Figure2). No postoperative problems occurred, and the cosmetic result was good(Figure3).Histologic examination of the excised material revealed sections of soft tissue covered by stratified squamous epithelium with parakeratosis. Numerous minor salivary glands, with moderate lymphocytic infiltration, were present in the underlying connective tissue. A few muscle fibers were also present in the specimen.

Discussion:

Double lip is an uncommon congenital or acquired anomaly that can have important consequences for the patient.[2] It consists of a fold of excess or redundant hypertrophic tissue on the mucosal side of the lip. The double lip occurs most often bilaterally on the upper lip, but may be unilateral; and can affect both the lips.[6] The condition, also referred to as macrocheilitis or hamartoma, has no predilection in terms of race or sex.[5,7]

The congenital form of double lip is thought to arise during the second or third month of gestation from a persistence of the sulcus between the Pars Glabrosa and the Pars Villosa of the lip. During fetal development, the upper lip mucosa consists of two transverse zones, an outer zone, which is smooth and similar to skin, the Pars Glabrosa and the inner zone, which is villous and similar to the oral mucosa, the Pars Villosa.[8] The furrow dividing the double lip represents the exaggerated boundary line between the two zones. In the double lip the buccal villous part becomes hypertrophic. In some patients, the central constriction is apparently due to the attachment of the upper frenulum. Although present at birth, the congenital condition may become apparent only after eruption of the teeth.[9]

The acquired form of double lip may be secondary to trauma and oral habit, and may

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syndrome which consists of the triad of blepharochalasis, nontoxic thyroid enlargement and double lip.[6,10] The first case of double lip and blepharochalasis was reported in 1909, and the association of these findings with thyroid enlargement was noted by Ascher.[4,6] It is not clear whether thyroid enlargement is a consistent or necessary feature of the syndrome. The lip becomes enlarged in a manner suggestive of angioneurotic edema and, over time, this swelling partially resolves.

The acquired form of double lip may be secondary to trauma and oral habit, and may develop in association with Ascher's syndrome which consists of the triad of blepharochalasis, nontoxic thyroid enlargement and double lip.[6,10] The first case of double lip and blepharochalasis was reported in 1909, and the association of these findings with thyroid enlargement was noted by Ascher.[4,6] It is not clear whether thyroid enlargement is a consistent or necessary feature of the syndrome. The lip becomes enlarged in a manner suggestive of angioneurotic edema and, over time, this swelling partially resolves. The association of congenital double lip with other abnormalities as bifid uvula and cleft palate has been described.[11]

Another uncommon acquired condition is Cheilitis Glandularis, an inflammatory hyperplasia with varying degrees of inflammation of the lower labial salivary glands.[10] The etiology of cheilitis glandularis is unknown, although familial inheritance and congenital predisposition, bacterial infection and irritation from sun, chemicals and tobacco have been observed as causes.

The differential diagnosis of cheilitis glandularis and congenital double lip is important, because cheilitis glandularis has been associated with an increased risk of the development of squamous cell carcinoma.[10] The differential diagnosis should also include vascular tumors, lymphangioma, angioedema, cheilitis granulomatosis, Meischer syndrome, mucocele, salivary gland tumours, inflammatory fibrous hyperplasia,
sarcoidosis, and plasma cell cheilitis. Such lesions are frequently associated with a uniformly enlarged lip without a midline constriction dividing the lip.[11]

This congenital or acquired abnormality caninterfere with chewing, speaking and esthetics. Recognition of the double lip and appropriate surgical treatment can reduce these potential problems.[3] The surgery involves excision of excess mucosa and submucosa without involving the underlying
muscular layer. Treatment can be done either under general or local anesthesia.[12,13] In case of central constriction of the lip, Eski M and other indicated use of double elliptical incisions combined with central vertical Z plasty.[14] However, according to Santos and Altamirano W plasty can also achieve similar results. Daniels stated that because of the progressive nature of disorder or suspected Ascher's syndrome the patient should be followed up because blepharochalasis and non toxic enlargement of thyroid gland can develop later.[15] Whereas Palma and Taub reported a case of double upper lip with recurrence.[16]interfere with chewing, speaking and esthetics. Recognition of the double lip and appropriate surgical treatment can reduce these potential problems.[3]

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Conclusion:

Double lip is of special interest in dentistry because the general practitioner is often the first professional to detect and establish the diagnosis of this uncommon condition. The treatment of congenital double lip is indicated when the excess tissue interferes with mastication or speech or leads to such habits as sucking or biting the redundant tissue or is of esthetic concern to the patient.

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Figure 1: Frontal View Of Patient With Maxillary Double Lip With Cupid's Bow Appearance

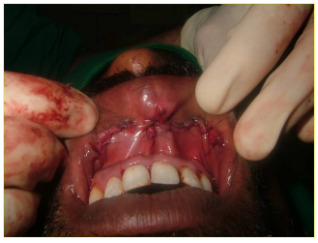

Figure 2: Photograph Showing Sutures Placed On The Inner Surface Of The Upper Lip

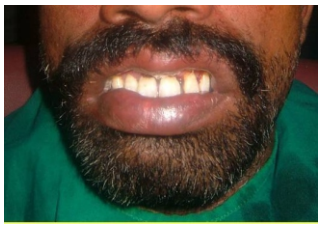

Figure 3: Postoperative Photograph After 3 Months

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