

CASE REPORT



Congenital maxillary double lip: A case report and review of literature

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Abstract

Congenital upper/maxillary double lip is a rare developmental anomaly. It is present since birth; however, it becomes manifest after the eruption of teeth. It is prominently visible on talking/speech, while smiling or when asked to show teeth. It may be associated with speech and mastication difficulties; however, the most common presenting cause is unseemly appearance and the possible embarrassment in public. Surgical excision of the excess offending tissue gives excellent results improving the appearance esthetically and giving a boost to the individual confidence levels.

Keywords: Ascher syndrome, congenital anomaly, double lip

Introduction

Double lip is a rare anomaly, in which the patient has a fold of hyperplastic/redundant labial tissue on either side of the midline (occasionally unilateral). Martins *et al.* suggested the involvement of upper lip more often than the lower lip. Peterson referred it to as “macrocheilia” or hamartoma and suggested that true macrocheilia as seen in congenital double lip must be differentiated from pseudomacrocheilia which involves eversion or protrusion of the lip without excessive tissue. The individual with double lip may experience problems in conversing and in mastication. Furthermore, the affected individual reveals abnormal facial appearances while smiling or when asked to show his teeth. However, usually, the patient has no complaints except cosmetic appearance and embarrassment in public specially on smiling. It has no particular gender or race predilection. However, Palma and Taub have suggested a male-to-female predilection of 7:1. The anomaly may be congenital or acquired or may be part of a syndrome. Laffler first reported a case of double lip and blepharochalasis in 1909. Later, Ascher’s syndrome characterized by episodic edema appearing such as angioedema of the eyelids (leading later to blepharochalasis), double lip, and non-toxic thyroid enlargement (goiter) was reported. The cause of the syndrome is unknown though trauma, hormonal dysfunction, and heredity have been suggested. Surgical excision is the treatment of choice. Surgical correction of double lip may be called for mainly for esthetic and cosmetic reasons and surgery may be

necessitated due to speech and/or mastication difficulties. Good results are accomplished by excision of the excessive mucosal and submucosal tissue, leaving the underlying muscles intact.

Case Report

A 20-year-old lady reported to the Department of Oral and Maxillofacial Surgery, SDM College of Dental Sciences and Hospital, Dharwad, Karnataka, India, with a chief complaint of abnormal upper lip. The patient apparently became conscious about the abnormal upper lip only when it was pointed out to her by visiting relatives, after she delivered a baby. History taking revealed that the patient was having this abnormality since birth. There was no other specific complaint regarding speech or mastication. Medical history did not reveal any comorbidities. The patient was, however, a lactating mother. Extraoral examination revealed an unusually large upper lip with lingually placed bilateral extra folds of tissue on either side of the midline in the form of a Cupid’s bow appearance, lying over the labial surfaces of maxillary anterior teeth. A thick band of fibrous tissue separated the two-fold. There was no change in the size of the upper lip at rest or during speech. The patient had lip incompetency for which she was advised to undergo orthodontic correction, but due to financial constraints, she refused the treatment.

Examination revealed intact mucosa over the upper lip with totally normal appearance. Other congenital anomalies, for

example, lip pitting were not noted in the examination about the vermilion border of the lip [Figure 1]. The patient did not report any functional abnormalities of speech or mastication. There was no history of lip biting or lip sucking habit. However, the patient gave a history of “sucking in” the extra tissue at time of stress. Palpation revealed a swelling that was soft in consistency, mobile, and fluctuant. The excessive mucosal folds were equal on either side of midline with constriction toward the labial frenum region (fibrous band). The condition did not bother the patient until it was pointed out by a relative. The patient was also unaware of surgical correction for the same.

Congenital maxillary double lip was the provisional diagnosis entertained. Surgical correction was planned and offered to the patient. Under local anesthesia (bilateral infraorbital nerve block), the hyperplastic redundant tissue with the midline fibrous band consisting of the double lip mass was outlined, and surgical excision was carried out using transverse elliptical incision. The local infiltration was avoided in the surgical area which would obliterate the mucosal folds, thus masking the margins of mucosal folds. Maxillary labial frenum was not excised to avoid alteration in the lip form. Minor salivary glands encountered in the surgical field were excised to prevent future mucocele formation. A light compression dressing was applied for 24 h after the procedure. No post-operative problems were encountered and the patient was discharged.



Figure 1: Pre-operative maxillary double lip



Figure 2: Post-operative picture after surgical correction

At discharge and on follow-up, the patient reported complete satisfaction with the cosmetic results. Six-month follow-up showed satisfactory functional and esthetic results, without evidence of recurrence and the patient was psychologically content with the results [Figure 2]. Histological examination of the excised soft tissue revealed sections 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 a rare anomaly which is characterized by a fold of hyperplastic/redundant labial tissue on either side of the midline which occasionally may be unilateral. It involves the upper lip more often than the lower lip. The anomaly becomes prominent during talking, smiling, or when asked to show teeth. Although it is a developmental anomaly, it becomes evident when the permanent teeth have erupted completely. Literature suggests that although the evidence of double lip becomes evident during speech and mastication, the presence of excess fold of redundant tissue at rest has not been highlighted in literature.^[1-4] However, in the case reported here, the patient showed redundant tissue at rest which became further prominent on smiling.

The maxillary double lip may be congenital/developmental in origin or it may be secondarily acquired as a result of various types of trauma. The congenital variety is thought to originate in the 4th–12th weeks embryonic development. It is postulated to be secondary to the persistence of horizontal sulcus separating the pars glabra and pars villosa of the developing lip. The exact cause for the formation of the congenital double lip is unknown, but it may be transmitted as an autosomal dominant disorder.^[5] The excision of the excess mucosa and submucosa sparing the underlying muscular layer as advocated by Epker and Wolford and is carried out under either local anesthesia, using infraorbital nerve block.^[6]

Gorlin *et al.*, in 1976, established that in such cases, narrowing at the central region is due to the attachment of the labial frenum and this was supported by multiple other reports. In the present report, we found that this case too had the central constriction of mucosal fold due to high labial frenum attachment.^[7]

John Daniels suggested that the acquired variety may be secondary to trauma following orthodontic treatment, trauma secondary to ill-fitting dentures, or habits such as lip biting and sucking of the lip between diastema. In this case, although the patient had a history of the lesion since birth, she also gave a history of sucking of the loose hypertrophic tissue during stress which accentuated the condition. This may be due to the expansion of loose mucosal fold as there is no attachment of orbicularis oris muscle. Conditions as those of chronic enlargement of the lip, hemangioma, lymphangioma, angioedema, cheilitis glandularis, and glandular granulomatosis need consideration in differential diagnosis of double lip. Of these, cheilitis glandularis is of the most

interest due to its malignant potential, squamous cell carcinoma reported in 18%–35% cases. These conditions, however, show a uniformly enlarged lip without a midline constriction. John Daniels also reported double lip in a triad of Ascher syndrome which also shows blepharochalasis and non-toxic thyroid enlargement.^[8] The case reported here did not show the other features of Ascher's syndrome. The patient had successfully undergone the stress of pregnancy. The thyroid function test carried out did not reveal any abnormality. She did not show any other anomalies, for example, bifid uvula, cleft palate, etc. Therefore, this case report is congenital non-syndromic and isolated case of maxillary double lip.

Lamster suggested that the treatment of choice for double lip is surgical. It is mainly indicated for esthetic reasons or mastication and/or speech problems. The surgery may be performed under local anesthesia (infraorbital nerve block with or without ring block). GA may also be used for the procedure.^[9-12] Peterson suggested other surgical methods include electrosurgical excision and triangular excision.^[13] In the case reported, the margins were identified and surgical excision was carried out so that surgical site heals with good vermilion and mucosal proportion. Multiple surgical methods have been employed for the correction of double lip in the absences of the central constriction excision through the transverse elliptical incision is the best choice (Reddy and Rao).^[14,15]

Recurrence is occasionally reported after surgery in cases of acquired double lip. Recurrence after surgery for the congenital condition has almost never been recorded. Histopathological examination must be undertaken to rule out malignancy or tuberculosis.

The microscopy would usually reveal mucous gland with hyperplasia, areolar tissue interspersed with extensive network of capillaries with plasma cell and lymphocytic infiltrations surrounding the blood vessels. The histopathology study of the excised tissue of our patient revealed normal mucosal lining of the lip, mucous glands with hyperplasia, and extensive numbers of capillaries.

Conclusion

Maxillary double lip should be thoroughly evaluated to rule out the Ascher syndrome. The causes of acquired double lip, for example, trauma due to various causes (previous orthodontic treatments,

ill-fitting dentures, and habit of sucking the extra tissues) should be considered and if present, addressed. A psychological assessment of the individual may be advisable. Counseling of the patient, before surgery, is of utmost importance for understanding the patient's expectations from the surgical intervention as also to stress on proper follow-up. The possibility of needing repeat procedure should be explained and documented.

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