

Double Lip Associated with Ascher's Syndrome and Its Surgical Correction: Case Report

Ascher's Sendromu ile İlişkili Çift Dudak ve Cerrahi Onarımı

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ABSTRACT Double lip is a rare oral anomaly that occurs most frequently in the upper lip with acquired or congenital origin. In some of the cases double lip is an indicator of the Ascher's syndrome. Sometimes, congenital form of double lip anomaly is associated with hemangiomas. It is equally prevalent in both genders. Often appears as two masses of hyperplastic tissue on either side of the midline of the lip as an excessive areolar tissue and inflammatory labial mucosa gland hyperplasia of the pars villosa. Orbicularis oris muscle is normal and muscle fibers cannot be found in the double lip. Surgical correction is necessary when the excessive tissue interferes with mastication or speech. Special attention has to be paid in the diagnosis of the pathology because it is sometimes confused with other serious anomalies and may be associated with Ascher's syndrome. This paper is a report of an upper double lip with Ascher's syndrome and its surgical management in a 57-year-old male patient.

Key Words: Surgery, oral; esthetics; diagnosis, oral; pathology, oral; lip; lip diseases

ÖZET Çift dudak, edinsel veya doğuştan orijinli olarak en sık üst dudakta meydana gelen ve nadir görülen oral bir anomalidir. Bazı vakalarda çift dudak Archer Sendromu'nun bir belirtisidir. Bazen, çift dudak anomalisinin doğuştan olan formu hemanjiyoma ile ilişkilendirilmektedir. Her iki cinsten eşit dağılım gösterir. Çift dudak çoğu kez, pars villozanın (iç zon) iltihabi labiyal mukozal bez hiperplazisi ve aşırı bir areolar doku olarak dudak orta hattının her iki tarafında hiperplastik dokunun çift taraflı şişliği şeklinde görülür. Orbikularis oris kası normaldir ve kas liflerine çift dudak içerisinde rastlanmaz. Çift dudakta doku fazlalığı konuşmayı ve çiğnemeyi engellediğinde cerrahi düzeltme gereklidir. Çift dudak Archer Sendromu ile ilişkili olması ve diğer ciddi patolojiler ile karışabilmesi sebebiyle teşhisinde oldukça dikkat edilmesi gereken bir patolojidir. Bu vaka raporunda Archer Sendromu ve çift dudağı olan 57 yaşındaki erkek hastaya ve çift dudağın cerrahi tedavisine yer verilmektedir.

Anahtar Kelimeler: Cerrahi, oral; estetik; tanı, oral; patoloji, oral; dudak; dudak hastalıkları

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Double lip is a rare clinical entity that can be diagnosed easily by careful patient history and examination as an accessory fold of redundant mucous membrane inside the vermillion border.¹⁻⁶ It is sometimes associated with Ascher's syndrome which is combination of blepharochalasis (a condition in which there is a redundancy of eyelid skin), nontoxic thyroid enlargement and upper double lip.¹⁻¹⁰ Moreover, congenital double lip can be associated with hemangiomas.⁹ This clinical entity has to be differentiated from other resembling chronic enlargements of the

lip.^{2,4-6,8} In most of the patients double lip is present as an isolated anomaly following oral habits or a trauma to the lips.^{1,2,4,5,9,10} Surgical intervention (simple excision) is the treatment of choice and in most of the patients good functional and cosmetic results can be obtained by this type of surgery.¹⁻⁶ In this study a case of congenital double lip with Ascher's syndrome and its surgical management have been described and differential diagnosis has been discussed.

CASE REPORT

57-year-old-man living in a small town was referred to the Ankara University, Faculty of Dentistry, Oral and Maxillofacial Surgery Clinic for surgical correction of bilateral extra fold of tissue in form of a cupid's bow in the upper lip by his cousin who is a research assistant in the department mentioned above. His medical history revealed that he had Ascher's syndrome as a systemic disorder.

Clinical examination revealed a second upper lip noticeable when the lips were tensed. A central constricting band which is dividing maxillary teeth, causes "cupid's bow" appearance. The two halves double lip occurrence was most likely due to the attachment of upper frenulum (Figure 1). The patient was wearing full upper denture and had been aware of the anomaly since he was 6-year-old. It was not causing any functional problem for the patient. However he had complained of unpleasant appearance during smiling recently. He occasionally "sucked in" the extra tissue during eating and speaking. No history of trauma was evident. Extra-oral examination also revealed both eyelid ptosis and blepharophimosis. And also there was a thyroid enlargement. This clinical appearance was suggestive of congenital upper double lip with Ascher's syndrome (Figure 2). Surgical excision under local anesthesia was the surgical treatment of choice. The anaesthesia was done as far as possible to prevent distortion of the mass. The excessive labial mucosa was removed by transverse elliptical incision extending to the commissures laterally and submucosa inferiorly. The incisions were extended deep into the submucosa so that a wedge-shaped



FIGURE 1: Double upper lip appearance before surgical procedure. (See color figure at <http://dishekimligi.turkiyeklinikleri.com/>)

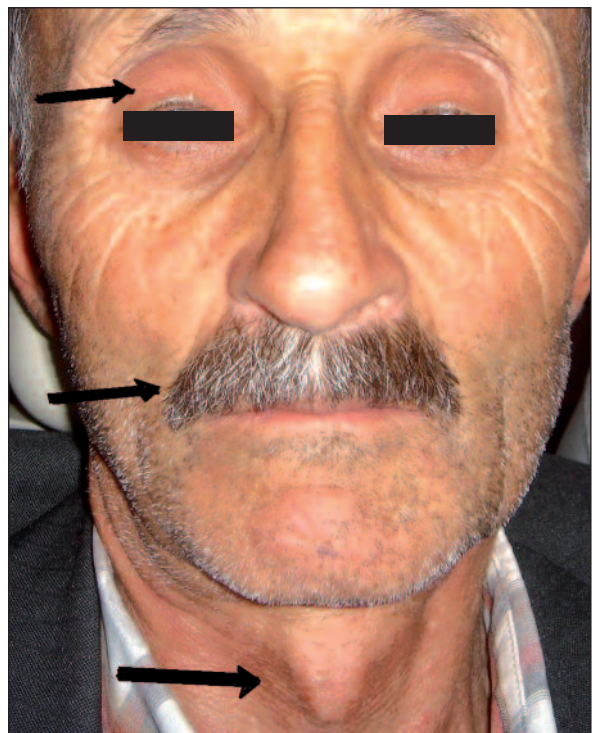


FIGURE 2: View of bilateral eyelid ptosis caused by blepharochalasis, after correction of the double lip and the thyroid enlargement. *Black arrows are showing the clinical appearance of Ascher's syndrome. ** Intermediate black arrow is showing after surgical correction of the double lip. (See color figure at <http://dishekimligi.turkiyeklinikleri.com/>)

block of tissue could be removed. Orbicularis oris muscle was left intact. The wound was closed in layers with the ordinary manner (Figure 3). A light compression dressing was applied for 2 hours after the procedure. No early postoperative complica-



FIGURE 3: View of the surgical area in just postoperative time.

(See color figure at <http://dishekimligi.turkiyeklinikleri.com/>)

tions were evident and sutures were removed in seven days. Histological examination revealed soft tissue covered by stratified squamous epithelium with parakeratosis. Numerous minor salivary glands, with moderate lymphocytic infiltration, were present in the underlying connective tissue. Long term follow-up examinations showed that cosmetic result was satisfactory and lip functions were normal. Two years following the surgery the healing was uneventful (Figure 4). A written consent was obtained from the patient for case presentation.

DISCUSSION

Double lip can be recognised easily during the routine clinical examination of general practi-



FIGURE 4: View of the improved upper lip following 2 years after surgery.

(See color figure at <http://dishekimligi.turkiyeklinikleri.com/>)

tioner. Double lip is a rare congenital or acquired anomaly consists of a fold of bilateral excess or redundant hypertrophic tissue on the mucosal side of the lip and it occurs often bilaterally on the upper lip as accessory one, but sometimes affects both lips and also referred to as macrocheilitis or hamartoma.¹⁻⁸ Although this anomaly occurs at birth, it usually becomes evident after the permanent teeth eruption. Also the congenital form mostly involves the upper lip.^{3-6,9,10} In the foetus during the second or third month of gestation the mucosa of the lip is divided into two transversal zones; an outer cutaneous zone (pars glabrosa) that is smooth and similar to the skin and an inner mucosal zone (pars villosa) which is similar to the oral mucosa.^{1,2,4,6} Hypertrophy and thickening of the pars villosa creates a groove which separates the two embryonic layers.^{1,3,8} As a matter of fact double lip is the result of hypertrophy of the pars villosa and the persistence of the sulcus between the pars glabrosa and the pars villosa of the lip.^{1,3,8,9} Moreover it has been suggested that the original double lip may be the result of a chronic trauma enhanced by a reactive process after habitual pulling of the mucosa through a diastema or “sucking-in” of the tissue between the teeth of maloccluding dentures.^{6,8,9} Constant suction by the teeth on the hyperplastic tissue is thought to be efficient factor in the increase of its size. The double lip deformity may develop in association with Ascher’s syndrome which is a rare entity and characterized by a double upper lip, blepharochalasis and nontoxic thyroid enlargement only in 10% to 50% of the patients.⁶ Additionally, Costa-Hanemann et al. found that the congenital double lip was associated with hemangiomas.⁹ Clinically double lip must be differentiated from other types of chronic enlargements. The differential diagnosis of double lip should include vascular tumours, lymphangioma, angioedema, cheilitis glandularis, cheilitis granulomatosa, Miescher syndrome, mucocele, salivary gland tumours, inflammatory fibrous hyperplasia, sarcoidosis, and plasma cell cheilitis.^{2-4,6,9,11} Surgical intervention becomes necessary for cosmetic reasons or when the condition interferes with

speech and mastication.^{1-4,6,9} Several surgical techniques have been described to repair double lip: electrosurgical, triangular excision and W-plasty which was advised by Guerrero-Santos and Altomirano can be mentioned among these.^{1-3,5} A simple elliptical surgical excision of the excess tis-

sue is the treatment of choice in most of the cases. Simple excision through an elliptical incision was advocated by Reddy and Kotewera.⁸ In our case a simple surgical excision through the transverse elliptical incision was the treatment of choice. We obtained good result with this kind of surgery.

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