Surgical management of double lip: A case report

B. Lewandowski[1-2], D. Brzęcka[1], R. Brodowski[1]

1Department of Maxillofacial Surgery of the F. Chopin Clinical Provincial Hospital in Rzeszów, Rzeszów, Poland
2Department of Emergency Medical Services, Medical Faculty of the University of Rzeszów, Rzeszów, Poland

Email: dagna.brzecka@gmail.com

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Abstract

Background Double lip is a rare developmental anomaly that mainly affects the upper lip. It is characterised by the presence of excess hyperplastic tissue that gives an illusion of double organ. It may occur independently or as a component of a set of defects, mainly in Ascher's syndrome, which presentes with blepharochalasis, non-toxic thyroid enlargement and double upper lip. The aim of this article is the presentation of a case report.

Case report The case of a 15-year-old patient with double lip treated surgically for cosmetic reasons is reported. Surgical excision under general anaesthesia was performed, without any complications neither during the surgery nor postoperatively, with no recurrence at the 2-year follow-up. Histopathological analysis showed normal oral mucosa with numerous hypertrophic mucosal glands, capillaries, and lymphocytic and plasmocytic infiltration.

Conclusion Due to the lack of unambiguous, clear recommendations and comparative studies in the literature, the choice of the incision depends on the experience and preferences of the operator. Regular follow-ups after the surgery are crucial; though recurrences are observed rarely, long-term, regular follow-ups are suggested due to the risk of developing additional symptoms characteristic for Ascher's syndrome.

KEYWORDS Ascher's syndrome; Double lip; Hyperplastic tissue.

Introduction

Double lip (labium duplex) is a rare developmental lip anomaly that may affect either one or both lips, more commonly the upper one [Martins et al., 2004]. The majority of authors did not observe any age or sex predilection, however Palma et al. [2009] presented a male to female ratio of 7:1. This defect is characterised by the presence of excess of non-inflammatory hyperplastic tissue in the mucous part of the lip, which gives the illusion of a double organ. Double lip may be either acquired of congenital. In foetal age the lip consists of 2 transverse zones: an outer cutaneous zone (pars glabrosa) and an inner mucosal zone (pars villosa). Congenital double lip is thought to arise as a result of pars villosa hypertrophy and persistence of an exaggerated horizontal sulcus between both parts, which is thought to normally disappear between the second and third month of gestation [Aggarwal et al., 2016; Daniels, 2010; Martins et al., 2004]. The acquired double lip may be developed secondary to trauma [Ali, 2007; Martins et al., 2004] or oral habits such as lip suction between diastema or malfunctioning prosthetic restorations [Martins et al., 2004]. It may also occur independently or as a component of set of defects, mainly in Ascher's syndrome in which it coexists with blepharochalasis and non-toxic thyroid enlargement [Aggarwal et al., 2016; Donato et al., 2017].

Case report

A 15-year-old female patient was referred to the Department of Maxillofacial Surgery of the F. Chopin Clinical Provincial Hospital in Rzeszow (Poland), because of the upper lip defect which was characterised by the presence of excess of tissue in the form of a thick fold on the labial mucosa between the upper and lower lip. According to the patient and her parents the anomaly was present since birth, at first slightly visible, it became gradually more apparent with age. Initially it did not cause visible look changes, but with time it caused a non-aesthetic appearance when smiling (Fig. 1). Clinical examination revealed 1x1.5 cm excess of tissue projecting beyond the lower margin of the upper lip, which was covered with mucosa of proper colour, of soft consistency and wide base "vanishing in lip tissue". A surgical management of the defect included an elliptical incision to remove the excess mucosal and submucosal tissues, leaving the orbicularis oris muscle undamaged was proposed to the patient. After standard blood and urine tests, which revealed normal parameters for this age group and receiving informed written consent of patient's parents, the surgical excision of the excess of tissue unger general anaesthesia was conducted. There were no complications during the surgery. The postoperative wound healed properly. Histopathological report of the excised material showed normal oral mucosa with numerous hypertrophic mucosal glands, capillaries, and lymphocytic and plasmocytic infiltration. Postoperatively the patient was followed-up for 2 years with regular recalls at 1, 3, 6, 12 and 24 months, which did not reveal any recurrence. The patient was very satisfied with the aesthetic and functional result of the surgical procedure.

Discussion and conclusion

The congenital double lip, although present since birth, become clinically evident after eruption of the permanent teeth [Martins et al., 2004]. Mostly not visible at rest, the excessive tissue become apparent during smiling when the lips mucosa lean against the teeth [Daniels, 2010; Martins et al., 2004]. Double upper lip is mostly present with central constriction, due to the attachment of labial frenulum [Hanemann et al., 2004; Srivastava et al., 2011], which is a significant criteria for differential diagnosis [Hanemann et al., 2004]. However, in the

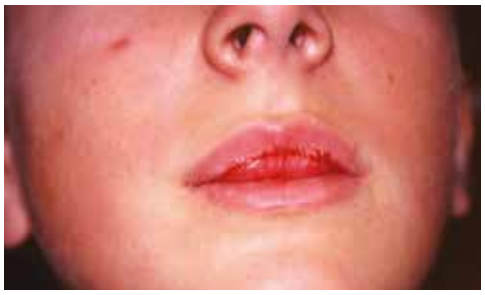

FIG. 1
Preoperative image of a 15-year-old female patient with the excess of tissue mimicking a double lip.

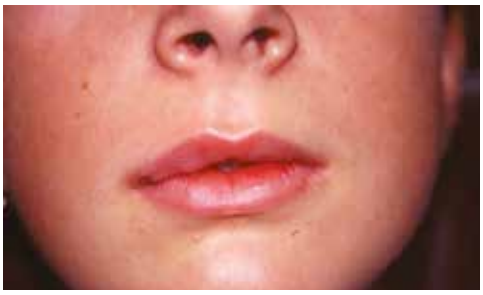

FIG. 2
Postoperative image.

report by Eski et al. [2007] such constriction was observed only in one of the five described cases.

Double lip may occur independently or as a component of a set of defects. In 1920 Ascher described the syndrome that bears his name, which consists of the triad of coexisting defects: double lip, blepharochalasis and non-toxic thyroid enlargement. The exact cause of this syndrome remain unknown but autosomal dominant inheritance is suggested [Daniels, 2010; Eski et al., 2007]. Nowadays blepharochalasis is thought to be present in approximately 80% of cases and is characterised by recurrent eyelid oedema that gradually leads to formation of redundant folds over the lid margins. It mostly starts at puberty, secondary to previously diagnosed double upper lip, which suggests the progressive nature of Ascher's syndrome. Non-toxic thyroid enlargement is present in 10–50% of the cases and is not necessary for the diagnosis of the syndrome [Ali, 2007; Donato et al., 2017]. In our clinical case the patient was not diagnosed with any additional defects, which suggests that double lip occurred independently. However, further follow-ups are needed, due to the risk of developing a full-fledged syndromic Ascher's disease in the future. Double lip has also been described in the literature in association with other congenital abnormalities. Parmar and Muranjan [2004] observed double lip coexisting with hypertelorism, eyelid ptosis, blepharophimosis, broad nose, clinodactyly and high-arched palate, suggesting a new, previously unrecognised syndrome. Hanemann et al. [2004] reported a case of recurrent double lip associated with enlargement of thyroid gland and numerous oral and facial haemangiomas.

In the differential diagnosis of double lip several possible lip anomalies should be included, such as cheilitis glandularis, cheilitis granulomatosa (Miescher's syndrome), mucocele, angioedema, haemangioma, salivary gland tumours as well as sarcoidosis [Daniels, 2010; Hanemann et al., 2004; Martins et al., 2004; Srivastava et al., 2011]. Cheilitis granularis is a rare inflammatory disorder of the lip that is associated with a relatively high incidence of squamous cell carcinoma, which is why great attention should be taken during examination of patients with lip anomalies [Atzeni et al., 2009; Martins et al., 2004]. The aforementioned lip pathologies usually do not present with central constriction characteristic for double lip associated with the attachment of the frenulum [Ariyawardana, 2011; Daniels, 2010; Hanemann et al., 2004]. Nowadays, in the differential diagnosis of double lip, magnetic resonance imaging (MRI) may also be used [Atzeni et al., 2009].

The treatment of choice in a double lip management is the surgical excision of the excess tissue that impairs speech, mastication or leads to developing pathological oral habits [Martins et al., 2004]. The majority of patients ask for surgery for cosmetic reasons. Specifically in young patients, the unaesthetic appearance may lead to low self-esteem and social exclusion [Ariyawardana, 2011]. The surgery is aimed at both aesthetic and functional improvement [Aggarwal et al., 2016]. Excision of the pathological tissue may be performed under general anaesthesia [Daniels, 2010], as in our case. However, local anaesthesia is mostly preferred, in such case infraorbital nerve block is suggested to avoid tissue distortion that may be present after local infiltration [Aggarwal et al., 2016; Srivastava et al. 2011]. During the surgery the excess of mucous and submucous tissue is excised leaving the muscle layer intact. According to the literature, in patients without the central constriction a simple elliptical incision is advisable [Ali, 2007; Ariyawardana, 2011; Hanemann et al., 2004], while in the presence of the constriction the authors propose two elliptical incisions combined with Z-plasty in the central part [Eski et al., 2007; Srivastana et al., 2011]. Nevertheless, in the case report by Srivastava et al. [2017] despite the presence of central constriction, only two elliptical incisions on both sides of the frenulum attachment were performed with satisfactory aesthetic outcome. Due to the lack of unambiguous, clear recommendations and comparative studies in the literature, the choice of the incision depends on the experience and preferences of the operator. According to Daniels [2010] patients that are planned for orthodontic or prosthetic treatment in the aesthetic zone should have the excision of excessive tissue deferred until completion of orthodontic treatment and proper stabilization of teeth. Regular follow-ups after the surgery are crucial. Recurrences are observed rarely. Palma et al. [2009] described only one case of recurrent double lip. No recurrence was observed within the period of two years in the patient described in our case. The authors suggest long-term, regular follow-ups due to the risk of developing additional symptoms characteristic for Ascher's syndrome in the future.

References

- Aggarwal T, Chawla K, Lamba A, Faraz F, Tandon S. Congenital double lip: a rare deformity treated surgically. World J Plast Surg 2016;5:303-307.
- Ali K. Ascher syndrome: a case report and review of the literature. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;103:26-28.
- Ariyawardana A. Congenital double upper lip: review of literature and report of a case. J Investig Clin Dent.2011;2:212-215.
- Atzeni M, Ceratola E, Zaccheddu E, Manca A, Saba L, Ribuffo D. Surgical correction and MR imaging of double lip in Ascher syndrome: record of a case and a review of the literature. Eur Rev Med Pharmacol Sci 2009;13:309-311.
- Daniels J. Congenital double upper lip: A case report and review of the literature. Saudi Dent J 2010;22:101-106.
- Donato C, Melo D, Santos J. Do you know this syndrome? Ascher's syndrome: clinical findings of little known triad. An Bras Dermatol 2017;92:729-730.
- Eski M, Nisanci M, Aktas A, Sengezer M. Congenital double lip: review of 5 cases. Br J Oral Maxillofac Surg 2007;45:68-70.
- Hanemann J, Oliveira D, Gomes M, dos Anjos M, Sant'ana E. Congenital double lip associated to hemangiomas: report of a case. Med Oral 2004;9:155-156.
- Martins W, Westphalen F, Sandrin R, Campagnoli E. Congenital maxillary double lip: review of the literature and report of a case. J Can Dent Assoc 2004;70:466-468.
- Palma M, Taub D. Recurrent double lip: literature review and report of a case. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2009;107:20-23.
- Parmar R, Muranjan M. A newly recognized syndrome with double upper and lower lip, hypertelorism, eyelid ptosis, blepharophimosis, and third finger clinodactyly. Am J Med Genet A 2004;124:200-201.
- Srivastava A, Chaudhri S, Kumar A, Srivastava S. Management of Double Lip. J Clin Diagn Res 2017;11:7-8.
- Srivastava A, Parihar A, Soni R, Shashikanth M. Chaturvedi T. Surgical Management of a Rare Case of Congenital Double Upper Lip. Case Rep Med. 2011.