

Surgical correction and MR imaging of double lip in Ascher syndrome: record of a case and a review of the literature

M. ATZENI, E. CERATOLA, F. ZACCHEDDU, A. MANCA*, L. SABA**, D. RIBUFFO

Section of Plastic Surgery, Department of Surgery; *Section of General Surgery, Department of Surgery; and **Department of Radiology, Cagliari University Hospital, Cagliari (Italy)

Abstract. – A double-lip is an infrequent anomaly which may occur either isolated or as a component of Ascher's syndrome. Apart from a deformity that may interfere with speech and mastication, surgery may be indicated for cosmetic reasons. We present a case of a male patient with an acquired double lip and blepharochalasis. In order to make the differential diagnosis a magnetic resonance imaging (MRI) was performed. The mass was removed by a transverse elliptical incision under local anesthesia. No post-operative problems occurred and the cosmetic result was good.

Key Words:

Ascher syndrome, Double lip, MR imaging.

Introduction

A double-lip (DB) is an infrequent anomaly which may occur either isolated¹ or as a component of Ascher's syndrome²⁻⁵. It is caused by abnormal non inflammatory labial mucosa gland hyperplasia of the pars villosa⁶ and it may require the surgical correction for aesthetic reasons. In this paper, we report a case of a male patient with an acquired double lip and blepharochalasis that was seen for cosmetic reasons. In order to make the differential, a diagnosis magnetic resonance imaging (MRI) was performed. He had never informed of the possibility of a surgical correction.

Case Report

A healthy 33 year old man been presented to our Department showing swelling of both sides of the upper lip (Figure 1). He did not present with functional abnormalities of speech or mastication and was wearing a full upper denture. He had not

lip sucking habit. The overlying mucosa was intact and appeared normal. On palpation the swelling was soft in consistency, mobile and fluctuant. The patient presented blepharochalasis and Ultrasonography (US) didn't show a thyroid enlargement. After written informed consent was obtained from the patient, a MRI was performed to study the mass (Figure 2). The mass was removed by transverse elliptical incision under local anesthesia, without resecting the maxillary labial frenum in order to avoid the modification of lip form. Surgical findings confirmed MRI results.

No post-operative problems occurred and the cosmetic result was good (Figure 3). Histologic examination of the excised material revealed section of labial mucosa covering numerous minor salivary glands and few muscle fibers (Figure 4).

Discussion

DB is an infrequent anomaly more commonly affecting the upper lip^{1,7} in the form of 2 masses of hyperplastic tissue on either side of the midline caused by excessive areolar tissue and non inflammatory labial mucosa gland hyperplasia of the pars villosa⁶. It sometimes can occur unilaterally in one or both the lips^{1,2,8}.

DB may be either congenital^{7,9,10} or acquired, secondary to trauma. The congenital form has been associated to the persistence of the horizontal sulcus between the pars glabra and the pars villosa of the lip^{5,11,12}. However, it has been suggested that the original DB may be enhanced by a reactive process after a "sucking-in" of the tissue between the teeth, or maloccluding dentures^{11,13}. Moreover, DB may occur either isolated¹ or as a component of Ascher's syndrome²⁻⁵. Some Authors² reported that the first case of DB



Figure 1. Initial aspect of the bilateral upper double lip of a 33 years old man.



Figure 3. Same patient. A 12-month follow-up photograph showing good esthetic result (He becomes overweight in these months).

and blepharocalasis was reported in 1902, but the first association of these findings with thyroid enlargement was notes in 1920 by Ascher¹⁴. However, goitre may be evident in only 10% to 50% of patients⁵⁻¹⁵.

The association of congenital double lip with bifid uvula², cleft palate¹⁶ and cheilitis glandularis has been described^{17,18}.

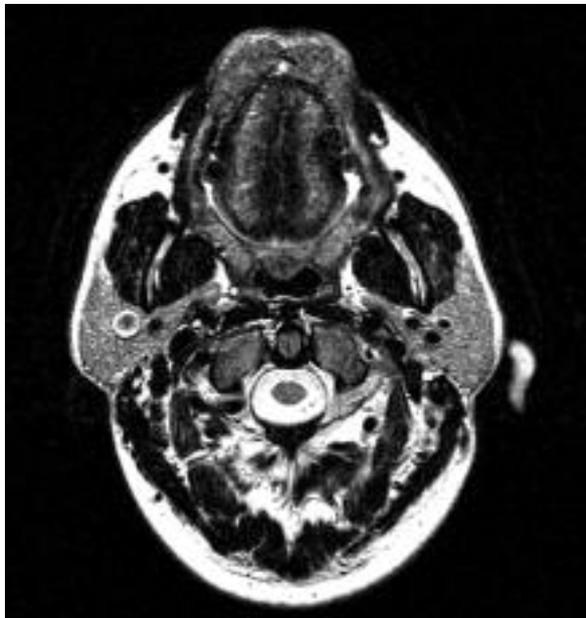


Figure 2. MR images depict the upper lip of the patient. Transverse fast spin-echo T2-weighted MR images (5,000/102; echo train length, seven; matrix size, 256 × 256; field of view, 150 × 200; section thickness, 3 mm; two signals acquired were obtained; acquisition time, 4 minutes); in the upper lip the presence of hyperplastic tissue with homogeneous iso-intense signal is clearly visible.

Parmar and Muranjan¹⁹ have reported a case of a 21-year-old male with double upper and lower lip, hypertelorism, unilateral ptosis, blepharophimosis, broad nose with broad nasal tip, highly arched palate and bilateral third finger clinodactyly. Also a case of congenital DB associated to hemangiomas was reported⁸.

The differential diagnosis should include vascular tumors, lymphangioma, angioedema, cheilitis glandularis, cheilitis granulomatosa, Miescher syndrome, mucocele, salivary gland tumours, inflammatory fibrous hyperplasia, sarcoidosis and plasma cell cheilitis². In particular we believe is important to make a differential diagnosis with cheilitis glandularis, because it has been associated with an increased risk of the development of the squamous cell carcinoma¹⁷.

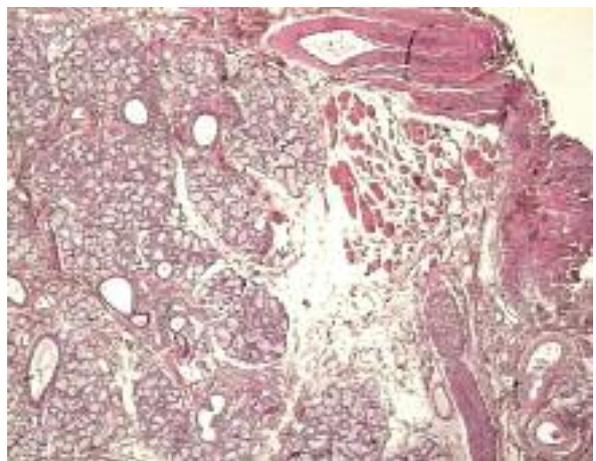


Figure 4. Microscopic aspect (hematoxylin and eosin, original magnification 20 ×).

In more than 80% of cases, Ascher's syndrome manifests itself before the age of 20⁴. In the present case, because the patient reported the increasing of the lip enlargement and blepharochalasis since he was older than 20 years old and because the absence of the goitre we preferred to propose him a MRI in order to determinate the nature of the swelling and to get a differential diagnosis.

Apart from a deformity that interferes with the mastication and the speech, a surgical treatment may be indicated for cosmetic reasons. Various surgical techniques to correct the DB have been described: w-plasty^{10,22}, "midmoon"²³ incisions, electrosurgical excision²⁴, triangle incision²⁵ or an elliptical excision on each side, combined with a vertical midline Z-plasty to release the central constriction²¹. However, several Authors suggest an elliptical incision^{1,7,26}.

Recurrence has not been observed in congenital forms¹², but sometimes it can happen in acquired ones². In our case, no recurrence occurred in a year of follow-up.

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