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**CASE STUDY**

# CONGENITAL DOUBLE UPPER LIP WITH EUTHYROID GOITER: A SHORT REVIEW WITH A CASE REPORT

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**ABSTRACT**

Lip is an essential feature of the face reflecting ones persona as well as forms an essential part of the mouth. Double lip is a rare anomaly and presents as a hyperplastic tissue on either side of the midline appearing as a Cupid's bow when it affects the upper lip. Double lip at times poses esthetic/functional glitches and can affect a young patient psychologically as a result of a marring effect on smiling. This article reports a 16 year old male patient who presented with double upper lip and euthyroid goiter sans blepharochalism. The patient was referred for cosmetic surgery and followed up without any recurrence or development of blepharochalism for a period of one year.



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## INTRODUCTION

Congenital Double lip (macrochelia / hamartoma) is a rare anomaly affecting the upperlip more than the lower lipand occurs in segregation or as a part of Ascher's syndrome. (Ariyawardana, 2011; Srivastava *et al*., 2011) It is initiated by excessive areolar tissue and non-inflammatory labial mucous gland hyperplasia of the pars villosaand comprises of a fold of excess or redundant hypertrophic tissue on the mucosal side of the lip. (Daniels, 2010; Aggarwal *et al*., 2016) Double lip commonlypresents as two masses of hyperplastic tissue on either side of the midline which can be at times asymmetrical. (Srivastava *et al*., 2011) Cosmetic surgery is usually the treatment of choice when the condition interferes with various functions such as smiling, speech and mastication. (Zhai *et al*., 2015) As the presence of double lip most frequently results in facial disfigurement, patients usually opt for surgery for cosmetic reasons. (Daniels, 2010) As upper lip and eyelid oedema develops at about the same time (> 80%) and as Ascher's syndrome manifests itself before the age of 20 years, prompt recognition of this condition is essential. (De Andrade Santos *et al*., 2008)

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Double lip cases are often undiagnosed or misdiagnosed by the practitioners. The aim of this report is to enlighten the practitioners regarding this rare anomaly as timely identification of this anomaly can avoid unnecessary tests and diagnostic delays permitting earlier planning of surgical treatment as well as anticipation of a full blown Ascher's syndrome later.

### Case report

A 14 year old male patient presented to the department of oral medicine and radiology with an enlarged upper lip. He visited the department as a part of regular dental check-up. According to the patient, the enlarged upper lip was present since birth with no change in appearance or size. There were no functional disturbances such as difficulty in mastication or speech associated with the lesion and there was no history of lip sucking. There was no previous history of trauma or concurrent medical problems. There was no relevant family history. General physical examination revealed a slightly enlarged thyroid gland which was in a euthyroid state. Ophthalmic examination ruled out blepharochalasis. Extra-oral examination revealed a single uniform enlargement of the upper lip. When the patient smiled, a transverse fold of excess tissue with a shallow transverse sulcus between the mucosa and the skin was

abnormally conspicuous. There was uniform sagging of the pars villosa which became prominent when the lip was tensed. (Figure 1) It measured about 4.5 cm X 2 cm. The swelling was of normal mucosal color and was visible only when the lip was stretched. It had a pebbly, grainy appearance. On palpation, the swelling was soft in consistency, non-compressible and mobile. There were no other anomalies visible extra-orally. Intra orally, vestibular depth appeared normal with adequate width of the attached gingiva in maxillary anterior region. No midline diastema could be appreciated and labial frenal attachment was normal. Dental occlusion appeared normal. No other anomaly was visible. A provisional diagnosis of congenital double lip with euthyroid goiter was given asa provisional diagnosis. Differential diagnoses of diffuse enlargement of the lip were considered such as angioedema, hemangioma, lymphangioma, chelitis granulomatosis and cheilitis glandularis. The patient was educated about the condition and advised for cosmetic surgery. The excised lip tissue was sent for histopathological examination which revealed minor salivary gland hyperplasia with areas of hemorrhage and inflammatory cell infiltrate. (Figure 2)

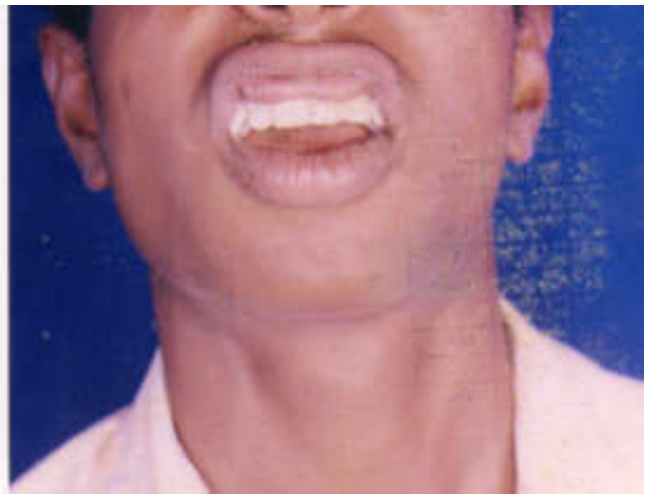

**Fig. 1. Cupid's bow appearance of the double upper lip**

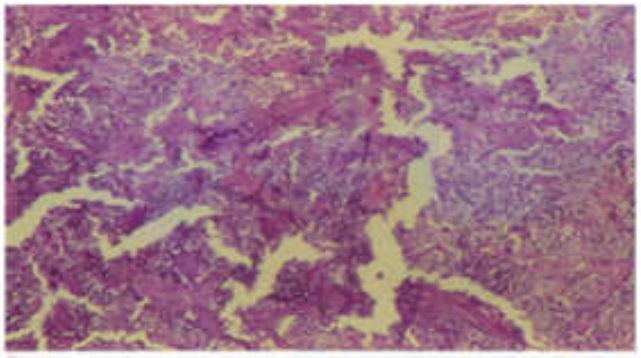

**Fig. 2. Hyperplastic minor salivary glands with areas of hemorrhage and inflammatory cell infiltrate**

## DISCUSSION

Double lip may be either congenital anomaly or acquired deformity. (Ariyawardana, 2011; Srivastava *et al*., 2011) The deformity presenting at birth (developmental anomaly) becomes more conspicuous as the patient grows i.e. after the eruption of permanent teeth. (Srivastava *et al*., 2011; Dhanapal *et al*., 2007) The acquired double lip may develop secondary to trauma or oral habits such as habitual sucking of the lip through the median diastema or between ill-fitting dentures. (Daniels, 2010; Aggarwal *et al*., 2016) Literature reveals a male predilection of 7:1. (Ariyawardana, 2011; Srivastava *et al*., 2011) The double lip is a hypertrophy of the pars villosa and is supposed to grow during second and third month of gestation as a result of persisting exaggerated horizontal sulcus between the pars glabrosa and the pars villosa of the developing lips. This appears as horizontal running duplication situated between innerpars villosa and outer zones pars glabrosa of the upper lip with excess fold of tissue protruding beyond the vermilion border. (Srivastava *et al*., 2011; Daniels, 2010) Pars villosa sags below pars glabrosa when the lip is tensed, producing a characteristic appearance; (Aggarwal *et al*., 2016) i.e during a smile or while talking, a vermillion with transverse furrow appears between the two borders but not seen when the lips are approximated. (Daniels, 2010) The reason being contraction of the muscle orbicularis oris, amplifies the horizontal sulcus, retracting the lip and placing the mucosa over the maxillary teeth producing a double lip/"Cupid's bow appearance". (Aggarwal *et al*., 2016)

When it affects the upper lip, literature describes the appearance of a double lip as a cupid's bow. (Ariyawardana, 2011) Previous reviews revealed double lip in association with various other anomalies. A case reported a 21 year old patient with double upper and lower lips, unilateral eyelid ptosis, hypertelorism, blepharophimosis, broad nose with broad nasal tip and bilateral third finger clinodactyly and a high arched palate. (Dhanapal *et al*., 2007) Another case report consisted of double upper lip associated with hemangiomas and goiter. Congenital upper lip has also been described with bifid uvula, hemangiomas, cheilitis glandularis, cleft lip and cleft palate. (Ariyawardana, 2011; Dhanapal *et al*., 2007) Also, a case of double upper lip in association with cheilitis glandularis was reported in a 14 year old female. (Dhanapal *et al*., 2007) In 1909, Laffer was the first one to describe the combination of blepharochalasis (swelling of the eyelids) and non-toxic goiter. In 1920, Ascher an ophthalmologist from Prague described a rare disease, Ascher' s syndrome (Laffer-Ascher's syndrome) consisting of a triad of double lip, blepharochalasis and euthyroid goiter which may be transmitted as an autosomal dominant disorder. (Aggarwal *et al*., 2016; De Andrade Santos *et al*., 2008) Both upper lip and eyelid oedema frequently appear at about the same timein more than 80% of the casesin thesyndrome. (Daniels, 2010; De Andrade Santos *et al*., 2008) The syndrome without non-toxic goiter is considered as the form efruste or incomplete form of Ascher's syndrome. (Zhai *et al*., 2015) The Ascher's syndrome is often undiagnosed by general practitioner because of its rarity and manifests itself before the age of 20 years. (De Andrade Santos *et al*., 2008) Angioneuroticoedema on both eyelids as well as on the upper lip constitutes the main clinical feature of this syndrome. Diagnosis of Ascher's syndrome is purely clinical.[8] The present case reported with double upper lip which was present since birth, euthyroid goiter which the patient was not aware of sans blepharochalasim.

Previous literature reveals that the presence of double lip and blepharochalasis is essential for the diagnosis of Ascher's syndrome (Daniels, 2010) and therefore a clinical diagnosis of Ascher's syndrome could not be given in the present case. Thyroid enlargement is not associated with any toxic symptoms. It usually presents several years after the initial eyelid and lip oedema,or may only be apparent after scanning with radioactive iodine. (Zhai *et al*., 2015; Daniels, 2010)

Enlargement of thyroid with variable degrees of severity has often been observed in previous case reports.[5]As thyroid enlargement is variable (10-50%) in Ascher's syndrome, it is not considered essential for the diagnosis. (Daniels, 2010) The present case had euthyroid goiter. The progressive nature of Ascher's syndrome is suggested by development of the blepharochalasis at times well after (years after) the double upper lip has been diagnosed. (Daniels, 2010)

There is a temporary swelling of both the upper eyelids which is followed by blepharochalasis (with or without telangiectasia), at times narrowing the palpebral fissure causing a reduction in the visual field. (Zhai *et al*., 2015) Blepharochalasis is a disorder manifested by thinning/atrophy, wrinkling and discoloration of the skin of the eyelids causing ultimate drooping of the affected eyelid. Secondary edema (recurrent) may easily develop as the skin is not adherent to the underlying tissueaggravating the atrophic changes of the skin overlying the eyelids. This causes the elastic skin components to become increasingly flaccid. The excess skin is invariably moreconspicuouslaterally than medially. Besides vision is frequently impaired. (De Andrade Santos *et al*., 2008) Blepharochalasis (> than 80%) is seen in Ascher's syndrome starting at puberty and usually involving both the upper eyelids. Pathologically it is a form of localized angioedema with a decrease in dermal elastin and has three stages: First stage or oedema stage manifests as intermittent painless swelling of the lids. Second stage or atonic ptosis manifests as a result of dehiscence of levatoraponeurosis or lax thin skin falling as redundant folds over lid margin. Third stage or ptosis adiposa manifests as medial fatpad atrophy, orbital fat prolapse and lacrimal gland prolapse. (Ramesh, 2011) Clinical examination of our patient didn't reveal any signs of blepharochalism.

Differential diagnosis of double lip should include vascular tumors like hemangioma, lymphangioma, angioedema, chelitis granulomatosis, cheilitis glandularis. (Dhanapal *et al*., 2007)

Histopathological examination of the excised specimen from the lips shows hyperplastic mucous glands, loose areolar tissue, numerous blood-filled capillaries and perivascular infiltration with plasma cells and lymphocytes. (Daniels, 2010)

The present case also showed similar histopathological features. Though at times double lip may not pose functional problems, these deformities may produce severe psychological anguish to the affected person as a result of the disfiguring effect on smiling. (De Andrade Santos *et al*., 2008) Management is aimed at both aesthetic and functional improvement. (Aggarwal *et al*., 2016) Cosmetic surgery comprises of excising the excess tissue of the eyelids and the lips. Good functional and cosmetic results are normally obtained. (Zhai *et al*., 2015) But surgery should be postponed for atleast one year from the previous attack of eyelid oedema. Indications for surgery being visual acuity, disturbance or ocular complications. (Ramesh, 2011)

The surgery of the double lip involves excision of the excess mucosa and submucosa without involving the underlying muscular layer under local anesthesia using infra-orbital nerve block or general anesthesia. The surgery is carried out using a transverse elliptical incision. (De Andrade Santos *et al*., 2008) Recurrence of the disease is rarely observed. According to some reports, since blepharochalasis may also arise years after the development of the double lip, it is possible that later a full blown Ascher's syndrome may occur in such patients demanding surgical mediation. As a result, it is mandatory that the patient should be on a long term follow-up. (Daniels, 2010) The present case was followed up for about one year without development of blepharochalasis after which the patient was lost to follow-up.

**Conclusion**

Arare case of congenital double upper lip with euthyroid goitersans blepharochalism in a 16 year old boy is presented. This is yet another addition to the paucity in literature of double upper lip cases presenting simultaneously with euthyroid goiter.

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