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Isolated Congenital Lip Sinuses Mimicking Vermillion Cleft of the Lower Lip: Report of a Rare Case and Review of Management Strategies

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Author's contribution

The sole author designed, analyzed and interpreted and prepared the manuscript.

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Case Study

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ABSTRACT

Isolated congenital lip sinus (ICLS) is one of the rarest congenital deformities seldom reported in the literature. The clinical presentation is extremely variable ranging from isolated single lip pit in the midline to deep sinuses, rarely presenting as fistulous tracts opening into the lower lip or alveolus. The management often includes surgical excision but may be associated with secondary aesthetic deformities that may range from obvious scars to mucous retention cysts. The aim of this paper is to report one such case of ICLS and discuss in detail best management strategy and also describe their prognostic significance.

Keywords: Congenital lower lip sinuses; van der woude syndrome; isolated congenital lower lip pits; cleft lip and palate.

1. INTRODUCTION

Congenital lip sinus / Pits is a rare condition, often reported only as a component of craniofacial syndromes. Lip pits and commissural pits are commonly associated with Van der Woude's Syndrome but rarely they may be associated with popletial pterygium syndrome,

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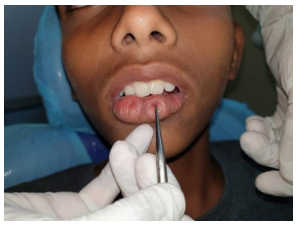
Peutz Jeghers syndrome, Syngnathia and Cremer's syndrome [1]. Occurrence of isolated non syndromic congenital lip pits is an extremely rare condition that has seldom been reported in the literature with 1 in 1lac being the current prevalence rate [2]. Clinical presentation of ICLS is extremely variable ranging from simple microform /Formae Frustae variants to more complex variants that may extend into the labial mucosa and/or into the alveolus [3]. Recurrent inflammation around the lip sinuses and the associated esthetic deformity are the primary reasons that warrant surgical intervention. Meticulous planning and execution of the surgical technique are essential in order prevent secondary lip deformities. The aim of the current paper is to describe a case of this rare anomaly and its management strategy with emphasis on the prognostic significance of the same.

2. CASE REPORT

A 14 year old male child from consanguineous parentage presented to our outpatient clinics with a complaint of unesthetic appearance of the lower lip. History revealed congenital swelling on the lower lip with exudation of fluid from the same. The pre natal and antenatal history was uncomplicated with history of vaginal delivery of the child at full term. There was no history of congenital anomalies in the family. General physical exam and detailed systemic evaluation of the patient was normal with no other systemic abnormalities. Patient was suggested to undergo genetic evaluation, however it was deferred due to financial constraints.

Clinical exam of the face revealed Bilateral para-median lip sinuses with increase in the thickness of the lower lip and hypotonia of the same. Salivary secretion from the base of the pits was evident with no evidence of any other congenital facial deformities. The depth of the lip pits was around 11 mm on the right and 9 mm on the left (Image 1,2) The sinuses were blind ended with no evidence of opening of the same on the labial mucosa or the alveolus. The patient was planned to undergo excision of the pits under local anesthesia. Bilateral Elliptical incision were placed around the lip pits parallel to the vermillion creases of the lower lip and dissection was carried out till the base of the lip sinuses and sinuses with the associated minor labial salivary glands were excised. The vermillion mucosa was released and pars marginalis component of orbicularis oris was dissected around the site of excision and the muscle was approximated in horizontal direction to avoid whistling deformity and reduce the bulk of the lower lip. This was followed by mucosal closure. Histological evaluation which revealed a tract lined by stratified squamous epithelium with thick fibrous connective tissue. Multiple minor salivary glands were seen opening at the base of these crypts with no evidence of pathological changes. Patient was asymptomatic after follow-up of 6 months with no evidence of retention cyst in the lower lip. The surgical site healed well with no aesthetic complications (Image 3).



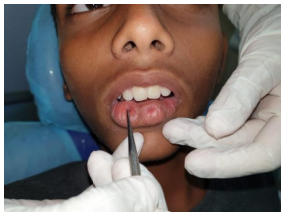


Image 1 and Image 2. Pre-operative image showing the lip sinuses

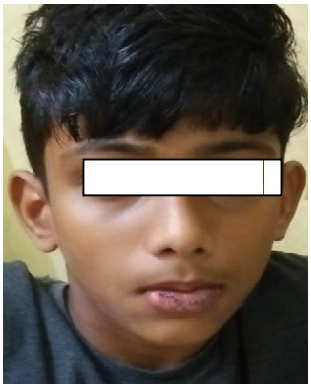


Image 3. Post-operative frontal profile

3. DISCUSSION

ICLS/ ICLP are one of the rarest congenital facial deformities that have seldom been reported in the literature. This condition was first described by Demerquay in the year 1845 [4] and since then less than 500 cases have been reported so far [2]. Congenital lip pits are often described as a part of craniofacial syndromes as mentioned previously. Andre Van der Woude identified the high prevalence of lower lip sinuses in association with bilateral cleft lip and referred to this condition as Vander-woude syndrome (VWS)/ Cheilognathouranoschisis [5]. He was also the first to describe the pedigree associated with the familial transmission of the genetic mutations associated with the same. Later,

Bocian et al. in the year 1987 described Iq32 and q41 were the primary loci of mutations associated with VWS with microdeletions in Ip34 being the second loci [6,7]. Though autosomal dominant mutations have been described pertaining to the incidence of non syndromic ICLS there is no definite evidence for the same [8,9].

Embryologically, ICLS have been considered to be a failure in the process of development of mandibular arch with epithelial invagination (Sulci) into the ectomesenchyme. Persistence of the residual epithelial tissue at the base of the sulci, beyond 6 weeks of intra uterine life leads to formation of the lip sinuses. [9] These sulci tend to deepen with age with hypertrophy of the surrounding connective tissue leading to thickening of lower lips predisposing aesthetic compromise. Rarely these sinuses may present with a transverse orientation which will clinically mimic a double lip [9]. Considering that the development of the lower lip, palate and the upper lip occur simultaneously in the intra uterine life, any terratogenic insult during this phase may interfere with the development of all the foresaid structures, clinically presenting as Van der Woude syndrome [10].

Rintala et al. described a comprehensive classification of the congenital lower lip pits and the forms of facial Clefts they are associated with. They found that, typical bilateral lip sinuses are the most common variants and are commonly associated with bilateral cleft lip [3] Histologically, these sulci are lined by stratified squamous epithelium similar to the vermillion mucosa with multiple minor salivary glands opening into the base of the sulci. Hypertrophy of connective tissue around the base of the sulci is evident which causes unusual thickening of the lip [11].

Management strategy in these cases should involve genetic counselling and parental counselling followed by surgical repair [8,12]. Parental counselling is extremely important as the progeny of these patients have a higher propensity to develop congenital facial deformity mainly the facial clefts.8 Crevenka et al. in their study reported that parents with ICLP/ICLS have 15 -29% chances of having progeny with facial clefts compared to parents with isolated cleft lip (10%) [8,12].

Surgical excision with incision parallel to the vermillion creases with muscle repair is essential to correct the esthetic deformity associated with this condition. Conservative management strategies including the use of chemical/heat Cauterisation, marsuplisation have been proposed with minimal success rates [11]. Meticulous planning is essential in order to avoid secondary deformities which may include bad scars, whistling deformity, mucosal entrapment cysts, paramedian tissue depression and abnormal vermillion contour [9].

The common complication associated with surgical excision is the formation of mucous entrapment cysts. This is attributed to incomplete excision of the sinus tract with the base of the tracts being left behind in the lip musculature. These cysts present as swelling at a later date which may warrant revision surgery. Bad scars and whisteling deformity are due to the incorrect direction of excision with inadequate muscle repair. These can be overcome by placing direction of excision parallel to the vermillion creases and by repairing the pars marginalis component of the orbicularis oris [11]. Rarely V-Y advancements/ Z plasty may be needed to close the defect to achieve good esthetic appearance [13,14] We were able to achieve satisfactory outcome in our case by meticulous closure of the orbicularis oris with no additional measures.

4. CONCLUSION

Isolated congenital lip sinuses are one of the rarest congenital conditions that have seldom been reported in the literature often warranting surgical excision for esthetic reasons. Parental counselling is extremely important considering the greater propensity for the incidence of facial clefts in the progeny.

CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Author has declared that no competing interests exist.

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