CLINICAL REPORT

Isolated Congenital Midline Upper Lip Sinus in A 5-year Old Child: A Rarity

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Introduction

The congenital sinus affecting lip is extremely uncommon [1], and when present, they are usually located at the commissure of the lip or at the midline of lower lip. They can occur as isolated anomaly or in association with other developmental deformities like cleft lip, cleft palate or both.

Holbrook [2], first described congenital midline sinus of the upper lip and since then only around 40 cases have been reported in literature affecting upper lip [3]. Because of its rarity, etiology is still unclear, though some embryological hypotheses had been put forward. The congenital lip sinuses involving lower lip have been reported earlier in India [4]. We are reporting a case of an isolated congenital midline upper lip sinus in a 5-year old male Indian child.

Case Report

A 5-year old male child, accompanied by his grandfather, came to our OPD with complaint of small depression just below the base of columella in the midline of philtrum (Fig. 1). This depression was since birth and on squeezing, a whitish discharge use to come out of it. There was no history of any external swelling or pain. The patient had no other congenital malformation and his family

history did not reveal any known congenital abnormalities.

On close examination of upper lip, a small pit of around 0.9 mm diameter was noted in the midline of philtrum just below the base of the columella. The probing was done, and probe could be passed a few millimeter away from the frenulum. There was no internal opening, but a very small painless swelling was noted lateral to frenulum, which on applying pressure yielded whitish discharge (Fig. 2). The discharge was sent for culture that came out to be sterile. Thus a diagnosis of congenital midline sinus of upper lip was established.

Pre-operatively, our surgical team was not much aware about the rarity of the disease, thus besides culture of the discharge only routine blood investigations were done and no radiological imaging was performed.

Intra-oral excision of the sinus was performed under general anaesthesia. The tract of the sinus was localized by introducing a metallic probe through opening and sublabial incision was given around 5 mm lateral to the frenulum. The tract was followed the along the probe and excised totally with some skin surrounding the opening, and measured around 8 mm. Intra-oral suturing was done with absorbable suture and two sutures were applied externally using prolene 6-0, which were removed on 5th post-operative day. At 10 months of follow-up, there was barely visible scar over philtrum without any recurrence of the sinus.

Discussion

Congenital sinuses of the lip, also known by names like pits or dimples, are extremely rare malformations [1]. De Murquay was the first to report and describe congenital lip

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Fig. 1 Sinus just below the base of the columella

Fig. 2 Whitish discharge coming out from the sinus

pits in 1845 [5]. When present, lower lip is mostly involved by congenital sinuses. The affection of upper lip is very rare with only around 40 cases reported in English literature, occurring either in midline or paramedian position. Holbrook [2], was first to describe congenital sinus of upper lip. Lower lip congenital sinuses have earlier been described in India [4], but only very few cases of midline congenital upper lip sinus had been reported [6].

These congenital anomalies occur either as an isolated defect or in association with other developmental deformities, including cleft lip, cleft palate or both [7]. There are cases of lower lip sinuses inherited as an autosomal dominant trait with variable penetrance [5]. Van der Woude (1954) studied 5 pedigrees and found that the association of lower lip sinuses with cleft lip and palate is based on single dominant gene. He recognized a syndrome that has since been known as Van der Woude syndrome [8].

Aoki et al. [9], reviewed all 31 reported cases of upper lip sinus through 2011 and classified them into

three types. Type 1 (13 cases) was a midline sinus without accompanying anomalies, type 2 (9 cases) were midline sinuses with accompanying anomalies, type 3 (9 cases) were lateral sinuses with or without accompanying anomalies. There had been report of coexistence of midline congenital upper lip sinus with Pierre-Robin syndrome [10].

Normal morphogenesis of oro-facial tissues includes a complete, step-by-step sequence of cell migration and of interaction between group of cells; but the mechanism responsible for dysmorphogenesis are still unclear [11]. There are reports suggesting that the midline upper lip cleft and midline upper lip sinus differs in etiopathogenesis.

Thus the exact etiology of midline upper lip sinus remains obscure and debatable and several theories had been proposed in this regard. Some have suggested abnormal fusion of facial prominences or merging of mesoblasts [12], but the most accepted theory is that of epidermal burrowing and invagination [13], resulting in formation of embryonic nasal pits and it may be postulated that midline upper lip sinus can also result from this mechanism.

The treatment, most common indicator for which is an unaesthetic cosmesis, consist of simple surgical excision of the entire sinus tract, as any remaining sinus tract can result in recurrence or cyst formation. The histopathology of most of the sinuses showed, to be lined by squamous epithelium having sebaceous glands, salivary gland sand hair follicles.

Conclusion

Though the etiology of congenital midline upper lip sinus is still not very clear and ours case is a new addition to the existing list of already reported cases, there might be a number of cases which can go unreported due to lack of awareness about the rarity of this disease and absence of any threatening symptoms.

Compliance with Ethical Standards

Funding The authors declare that this study has received no financial support.

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Approval All procedures performed on patient in study were in accordance with the ethical standards of institutional committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was taken from the patient for use of clinical photographs.

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