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Congenital midline sinus of the upper lip: A case report and review of literature

Bareqa I. Salah^{a,*}, Baeth Al-Rawashdeh^b, Zaid R. Al-Ali^c, Mohamad Mahseeri^d, Zayed Al-Zu'bi^c

^a Jordan University Hospital, Department of General Surgery, P.O. Box 755, Tla-Ali, Jordan

^b Jordan University Hospital, Department of Special Surgery, 11942, P.O. Box 13046, Amman, Jordan

^c Jordan University Hospital, Department of Oral and Maxillofacial Surgery, Jordan

^d Jordan University Hospital, Department of General Surgery, Jordan

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ABSTRACT

INTRODUCTION: Congenital pits of the lip are uncommon and may be associated with conditions such as Van der Woude syndrome. Isolated lip pits are extremely rare developmental defects.

PRESENTATION OF CASE: A 7 year old Caucasian girl presented to plastic surgery clinic in Jordan University academic hospital complaining of an upper lip pit that has been present since birth. It was associated with a single episode of whitish discharge. On examination, an isolated upper lip midline sinus was found. She is otherwise fit and healthy. Surgical excision was curative with good cosmetic outcome.

DISCUSSION: The prevalence of lower lip sinuses has been estimated to be about 0.00001% of the white population. Upper lip sinuses are even more uncommon. To date, there have been several case reports of upper lip sinuses and fistulas. A total of 55 cases was found upon reviewing the English literature. No similar cases were reported in Jordan. Different presentations are mentioned in this review. Several embryologic theories are presented.

CONCLUSION: Upper lip sinus formations are rare developmental events, and the pathogenesis of these lesions still needs further evaluation. Simple surgical excision is the treatment of choice.

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1. Introduction

Congenital perioral sinuses, pits or dimples can be seen in lower lips of patients with cleft lip and palate or Van der Woude syndrome. However, it is very rare to find isolated congenital upper lip sinuses and fistulae [1]. We report a very rare case of congenital midline sinus of upper lip with fluid discharge that was managed in Jordan University academic hospital. No previous cases have been reported in Jordan. A review of reported cases in the past 20 years has been conducted. Etiology and classification of this congenital anomaly will also be discussed. This work has been reported in line with the SCARE criteria [2].

2. Case report

A seven year old girl previously healthy presented to plastic surgery clinic with an upper lip midline pit. It was associated with one episode of whitish discharge. This was present since birth with no history of pain, recurrent swelling or infection as her mother states. She is a school student and comes from a middle class Caucasian family. Family history was unremarkable.

Upon physical examination, she had a normal growth pattern and normal laboratory blood results. An opening of 2 mm diameter was found at the midline of an otherwise normal upper lip, 10 mm inferior to collumela of nose and 4 mm superior to white roll of upper lip (Fig. 1). No similar openings were found on the mucosal surface of the lip, indicating a blind-ending sinus which was confirmed by inserting a fistula probe. No other congenital anomalies were found in either lips such as cleft lip or palate, no fluid discharge from the opening noticed upon applying pressure on the lip. Given the history and examination, she was booked for a routine sinus surgical excision under general anesthesia in school's Spring break.

Intraoperatively, methylene blue dye was injected into the sinus opening, then a small vertical elliptical skin incision was made around the opening. Sinus tract was excised using extraoral approach. Closure of skin and subcutaneous was done

* Corresponding author.

E-mail addresses: bareqahebishan@gmail.com

(B.I. Salah), drbraw@gmail.com (B. Al-Rawashdeh),

alali.zaid@hotmail.com (Z.R. Al-Ali), mohamadp1987@hotmail.com (M. Mahseeri),

zaid_alz3bi@hotmail.com (Z. Al-Zu'bi).



Fig. 1. Pit in the midline of the upper lip.



Fig. 2. Inserted probe confirms the presence of sinus tract.



Fig. 3. Dissection of sinus tract.

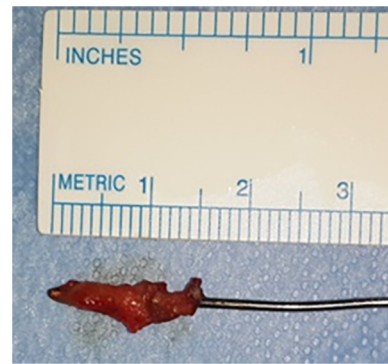


Fig. 4. Excised sinus tract.

(Figs. 2 and 3). Histopathological examination of the specimen revealed a sinus tract lined by benign stratified keratinized squamous epithelium. It is surrounded by dermal structures and subcutaneous tissue containing skeletal muscle (Fig. 4). Postoperative recovery was unremarkable. She was followed up on a monthly basis for the first 3 months then every 3 months for 12 months with no reported complications.

3. Discussion

Perioral sinuses are very rare presentation, with prevalence of 0.001% for lower lip sinus [3]. Midline upper lip sinus is even more rare condition that is infrequently reported in the literature, with a total of 55 cases reported in English literature. The first case was reported by Lanneloque et al back in 1879 [4]. Nagasao et al. [5] reported 33 cases through 1995. We reviewed the literature for new cases in the past two decades 1996–2016, we found 22 new cases including our reported case (Table 1).

Nagasao found that majority (18/32) of the cases were females. But due to small number of cases, this conclusion is not precisely accurate. Also, majority of cases have been reported from South Asian countries. This may indicate race predilection for Asian race group [5].

Upon reviewing the literature in the past two decades we found 22 new cases of upper lip midline sinus. The clinical presentation ranged from asymptomatic and cosmetic concerns (8/22) to recurrent upper lip swelling (11/22) and intermittent discharge (12/22). Fluid discharge could be clear, whitish or pus in nature. The vast majority of cases were blind-end tract (20/22) and only 2 cases were fistulae (Table 1).

According to Aoki et al who developed a classification system of upper lip sinuses, three categories were introduced depending on associated anomalies and position of the presenting sinus. (a) Type I: midline sinus without accompanying anomalies; (b) Type II: midline sinus with accompanying anomalies; and (c) Type III: lateral sinus with or without accompanying anomalies [6].

Unlike lower lip sinuses, which are commonly associated with cleft lip, alveolus and palate [7] we found that most of the upper lip midline sinus cases were not associated with any other congenital anomalies (type I) (37/55). When other anomalies were present (type II), median cleft lip (7/55) and frenal anomaly (3/55) were the most common associated ones. Other reported anomalies were: Pierre-Robin syndrome, unilateral cleft lip, dorsum of nose sinuses, asymmetrical nasal opening, preauricular fistula, notch of vermilion, intra oral fibroid polyp, midline alveolar process cleft and grooving, bifid uvula, notching of posterior nasal spine and ankyloglossia (Table 1).

The etiology is not fully understood until now due to scarcity of reported cases. The literature provides three theories on its embryopathogenesis: The invagination theory proposes that upper lip

Table 1
Upper lip midline sinuses and fistulae reported between 1997 and 2017.

Num.	Author, Year	Age	Sex	Country	Signs	Type	Associated anomalies
1.	Asahina, 1997 [8]	5	F	Japan	Intermittent swelling, mucus discharge	Sinus	None
2.	Shigihara, 1997 [9]	2	F	Japan	Asymptomatic	Sinus	Intraoral fibroid polyp
3.	Licht, 1998 [10]	16	M	USA	Mucus discharge	Sinus	Median cleft lip
4.	Illing, 1999 [1]	13	F	UK	Asymptomatic	Sinus	None
5.	Al-Qattan, 2000 [11]	12	F	KSA	Recurrent cellulitis	Sinus	
6.	Sumitomo, 2002 [12]	22	F	Japan	Asymptomatic	Sinus	
7.	Sancho, 2002 [13]	2	F	Spain	Intermittent clear fluid discharge, recurrent swelling	Sinus	Bifid maxillary frenulum, midline cleft of primitive palate
8.	Sancho, 2002 [13]	7	F	Spain	clear fluid discharge, intermittent swelling	Sinus	
9.	Charrier, 2006 [7]	8m	F	France	Intermittent swelling and discharge	Sinus	None
10.	Tuncali, 2006 [14]	4	M	Turkey	Asymptomatic	Sinus	Midline grooving of maxillary alveolar process
11.	Tuncali, 2006 [14]	4	M	Turkey		Sinus	Microform midline clefting of upper lip, partial median clefting of maxillary alveolar process, bifid uvula, notching of posterior nasal spine
12.	Sen, 2006 [15]	47	F	Turkey	Recurrent swelling, pain, discharge and erythema	Sinus	None
13.	Nakano, 2010 [16]	5	F	Japan	Recurrent swelling and pus discharge	Fistula	
14.	Salati, 2012 [17]	6	F	KSA	Thick discharge	Fistula	
15.	Aoki, 2012 [6]	1	F	Japan	Intermittent swelling and discharge of white fluid	Sinus	None
16.	Jung, 2012 [18]	14	F	Korea	Asymptomatic	Sinus	None
17.	Fok, 2014 [19]	11	F	Singapore	Recurrent swelling and abscess	Sinus	None
18.	Anicete, 2014 [20]	2	F	Singapore	Whitish discharge	Sinus	None
19.	Bakshi, 2015 [21]	10	F	India	Recurrent swelling and pus discharge	Sinus	None
20.	Hili, 2016 [22]	7	F	UK	Asymptomatic	Sinus	None
21.	Xu, 2016 [23]	2	M	China		Sinus	None
22.	Our study, 2016	7	F	Jordan	Whitish discharge	Sinus	None

sinuses are formed by failure of ectodermal invagination of the nasal placodes during the frontonasal process [19]; the merging theory, the formation of upper lip fistula is explained by the paucity of mesenchymal cell volume, these mesenchymal cells give rise to swellings and furrows on the developing face, and the paucity of these cells may lead to formation of upper lip fistula [21]; and failure in the forward growth of the frontonasal process or a failure in the complete fusion between the maxillary and the frontal process could explain the formation of lateral fistula but not the median fistula [13].

Management of this condition was agreed upon by most authors. Excision of the sinus tract was associated with best results regarding recurrence and symptoms. Histopathological examination revealed the tracts are aligned with stratified squamous epithelium in all cases, and some showed sebaceous glands, hair follicles, mucous glands, salivary glands, or even hyaline cartilage [21].

4. Conclusion

Upper lip midline pits and sinuses are extremely rare. Lip pits can present as asymptomatic irregularities of the lip or with recurrent discharge or infection. The mechanisms involved in congenital upper lip sinus formation are incompletely understood. Surgical excision is generally curative with good outcome.

Conflicts of interest

No conflict of interest.

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Ethical approval

Ethically approved by the Jordan University Hospital's ethical committee.

Consent

Written informed consent was obtained from the patient's father for publication of this case report.

Author contribution

Dr. Bareqa Salah – Surgeon operator.
Dr Baeth Rawashdeh – Surgeon operator.
Dr Zaid Al-Ali – Data collection and analysis.
Dr Mohamad Mahseeri – study design and writing.
Dr Zayed Al-Zu'bi – study design and writing.

Registration of research studies

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Guarantor

Dr Bareqa Salah.

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