Congenital Oral Synechiae: A Case Report and Review

Buche A. R.[1*], Borade V. R[2], Suman Neha[3], Chintale S.[4], Jaiswal S. A.[5]

{[1]Resident, [2,3,4]Assistant Professor, [5]Professor and Head}

Department of ENT, Government Medical College, Solapur, Maharashtra, INDIA.

[*]Corresponding Address:

draniketbuche@yahoo.in

Case Report

Abstract: Congenital oral synechiae are rare entity out of all congenital malformation, typically occurring between theupper and lower alveolar ridges or between the tongue and marginsof the palate or maxilla .Neonatal respiratory distress and feeding difficulty are the most common clinical presentation of an oral synechiae and it often require prompt intervention.There may be residual limitationof mandibular excursion following division of the band. Here we are reportingcase of congenital oral synechiae which was operated successfully.

Keywords: Congenital, oral synechiae, cleft palate, feeding difficulty.

Introduction

Oral synechiae is aextremely rare congenital anomaly which was documented in literature by very few authors. These are broadly divided intolateral synechiae and medial synechiae out of which lateral synechiae are more common. Early diagnosis and appropriate treatment can avoid serious complications in neonate like breathing difficulty secondary to upper airway obstruction and feeding difficulties. These oral synechiaesometimes may be a part of some syndromes like congenital alveolar synechiae syndrome, Vander Woudesyndrome , popliteal pterygium syndrome, and oromandibular–limb hypogenesis spectrum or it may be associated with other anomalies like hydrocephalus,cleft of the secondary palate, bifid tongue, patent ductusarteriosus, hypospadius, and bifid scrotum. (1-6)

Case Report

A 2 day old full term female neonate who was 4th child of 27 year old female out of non consanguineousmarriage, referred to ENT department with complaint of inability to breast feed with some abnormal tissue connecting upper and lower jaw. A normal vaginal delivery with birth weight of 2.740kg with no history of noisy breathing or cyanosis spell. There was no prenatal historyof maternal infection, gestational diabetes, exposure to alcohol, tobacco, drugs, medications or toxins. There was no family history of oromaxillary or facial anomaly or any other congenital anomaly in other siblings.Oral examination suggestive of two lateral fibrous band with median free space and fibrous band connecting floor of mouth with inner or medial alveolar margin of palate. Right sided bandwas thin and broader compared to that on left side. Palate and visualised tongue was normal. *(Figure 1)* Rest of ENT examination was within normal limit. In systemic examination she had soft systolic murmur rest systemic examination is within normal limit. Patient was evaluated by paediatrician for any other associated congenital anomaly with all other routine blood and radiological investigation, which found to be within normal limit except 2D echo cardiogram showing small muscular ventricular septal defect (VSD) with left to right shunt and patent foramen ovale with left to right shunt,no patent ductusarteriosus (PDA) with normalatria, ventricle and valves showing congenital cardiac anomaly. So surgery to release intraoral fibrous band was planned under general anaesthesia and release of fibrous band done with scissor and bipolarelectrocautery *(Figure 2)*. Complete oral examination done after resection of band and oral cavity found to be normal. *(Figure 3)* Perioperative and post operative period was uneventful. Patient was started on breast feed on 1st post operativeday.

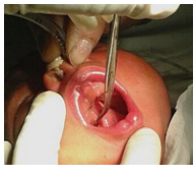

Figure 1

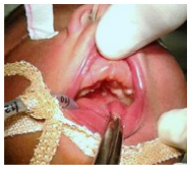

Figure 2

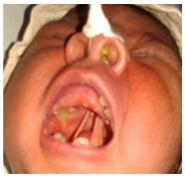

Figure 3

Discussion

The first case of oral synechiae was reported by Illera in 1875 but in 1957 it was Hayward and Every who documented first case of lateralsynechiae with cleft palate. Out of around 60 documented cases of oral synechiae till date 52 were of lateral and other were of median variety.(7) These are of various degrees and can be classified in to five types like synechiae by cord like adhesion of the alveolar mucosa of upper and lower jaw(alveolar synechiae) ; synechiae by membranous adhesion on hard palate and floor of mouth excluding

tongue (lateral synechiae);synechiae partially involving hard palate and tongue; synechiae widely involving soft palate and tongue so that continuity is interrupted between oral cavity and pharynx ; and synechiae by membranous adhesion between hard palate and lower lip. [8] Gartlan *et al* stated that if oral bands are consisted of soft tissue only,it can be termed as synechiae and it involves bone then it can be called as syngnathia. He also divided oral synechiae in 2 categories: 1) posterior intraoral bands near base of tongue representing remnants of buccopharyngeal membrane 2) anterior intraoral band which are ectopic membrane subclassified into subglossopalatal band,glossopalatalankylosis or syngnathia. But in contrast to this Gorlin *et al* argued that synechiae in anterior oral cavity are derived from vestiges of buccopharyngeal membrane because the tongue developed behind the buccopharyngealmembrane. [1] Though exactetiology is not known many theories are given to explain etiology of intraoral synechiae like genetic,teratogenicormechanical insult during critical stage of embryonal development of alveolar ridges, tongue and palatal shelves[9]; while Longacre gave theory of persistantbuccopharyngeal membrane ; Kruger stated that the mechanical effect of tongue may give rise to adhesion between mucosa of palate and floor of mouth and adhesion in cases of cleft palate and floor of mouth is result of obstruction by tongue [10];according to Mathisadhering epithelial rudiments during development of palate may give rise to oral synechiae. [11]Intraoral synechiae can involve various sites, including the alveolar ridges (Dalal and Davison, 2002; Tanrikulu *et al*., 2005), tongue (Kalu and Moss, 2004), the lingual base (Chandra *et al*., 1974), floor of the mouth (Nakajima *et al*., 1979; Zalzal *et al*., 1986), and along the midline of the secondary palate (Chandra *et al*., 1974; Dalal and Davison, 2002; Kalu and Moss, 2004).Oral synechiaewere rarely seen in isolation but these were most of the time associated with some other congenital defect or some syndromes. Donepudi *et al*. reported two cases of cleft palate lateral synechia (CPLS) syndrome in a single family and described surgical management of the synechia. [12] Albert K. Oh reported subglossopalatal synechia in association with cardiac and digital anomalies. [13] Snijman *et al*. reported congenital fusion of gums in a 3 week old child along with temporomandibular joint ankylosis. [14]Nakajima *et al*. (1979) reported an infant born with a large subglossopalatal membrane, velar cleft, ankylosis of the distal interphalangeal joints of the fifth fingers, absence of the distal phalanx of a fifth toe, and patent ductusarteriosus. A male infant with a subglossopalatal membrane, soft palatal cleft, bilateral hypoplastic small fingers,third-degree hypospadias, and a bifid scrotum, has also been described (Zalzal *et al*., 1986). Other syndromes associated with oral synechiae are Vander Woude syndrome which is caused by mutations in interferon regulatory factor 6(IRF6) (Kwang *et al*., 2002); Popliteal pterygium syndrome, which includes intraoral synechiae (Nauman and Schulman, 1961; Gartlan *et al*., 1993; Knoll *et al*., 2000), is also caused by mutations in IRF6 (Kwang *et al*., 2002) ;and Murphy *et al* documented 2 cases ofcleft palate and congenital alveolar synechiae syndrome (previously defined by Verdi and O'Neal, in 1984). Respiratory distress and feeding difficulties are the major causes of morbidity in infants with intraoral synechia (Chandra *et al*., 1974; Nakajima *et al*., 1979; Zalzal *et al*.,1986; Bagatin and Boric, 1990; Pillai *et al*., 1990; Gartlan *et al*., 1993; Denion *et al*., 2002; Kalu and Moss, 2004). In our patient congenital oral synechiae had complaint of feeding difficulty but no evidence of respiratory distress. It was associated with congenital heart disease without any other anomaly. There are different methods have been mentioned in the literature to excise these bands using surgical instruments ,silk ligatures, scissors , electrocautery , and with scalpel. Schonauer *et al*. reported endoscopic release of intra oral fibrous bands in a patient with popliteal pterygium syndrome. [16] In our patient we excise oral synechiae with the help of scissors and electrocautery and 1 month post operative follow up there was no readession. Considering rarity of case this case make interesting reading for all.

Conclusion

Early diagnosis and appropriate treatment with complete assessment of patient for any other congenital anomaly is advisable in the patient of oral synechiae

References

1. Gartlan MG, Davies J, Smith RJ. Congenital oral synechiae. Ann OtolRhinolLaryngol. 1993;102:186–197.
2. Chandra R, Yadava VNSY, Sharma RN. Persistent buccopharyngealmembrane. PlastReconstr Surg. 1974;54:678–679.
3. Bagatin M, Boric V. Congenital intraoral epithelial bands: report of twocases. J Oral Maxillofac Surg. 1990;48:309–310.
4. Pillai KG, Kamath VV, Kumar GS, Nagamani N. Persistent buccopharyngeal membrane with cleft palate. Oral Surg Oral Med Oral Pathol.1990;69:164–166.
5. Nakajima T, Takahashi M, Tateno S. Subglossopalatal membrane. PlastReconstr Surg. 1979;63:574–576.
6. Zalzal GH, Bratcher GO, Cotton RT. Subglossopalatal membrane. ArchOtolaryngol Head Neck Surg. 1986;112:1101–1103.
7. Hayward JR , Avery JK. A variation in cleft palate.J Oral Surg. 1975:15:320-4
8. Ogino A, Onish K, Maruyama Y. Congenital oral synechia associated with cleft palate: Cleft palate medial synechia syndrome? Eur J PlastSurg 2005;27:338-40.

9. Haydar SG, Tercan A, Uçkan S, Gürakan B. Congenital gumsynechiae as an isolated anomaly: A case report. J ClinPediatr Dent 2003;28:81 -3.
10. Kruger GO. Textbook of oral surgery. 6thed. Jaypee Brothers: Mosby 1990. p. 462.
11. Mathis H. Case report of patient with difficulty in eating due to congenital syngnathia. German Dent J 1962;16:1167-71.
12. Donepudi SK et al (2009) Cleft palate lateral synechia syndrome: an opportunity for unique surgical closure.Int J PediatrOtorhinolaryngol. 73(6):861–866.
13. Oh AK (2008) Subglossopalatal synechia in association with cardiacand digital anomalies. Cleft Palate Craniofac J 45(2):217–221.
14. Snijman PC et al (1966) Congenital fusion of the gums. Am J DisChild 112(6):593–595.
15. Verdi GD, O'Neal B (1984) Cleft palate and congenital alveolar synechia syndrome. PlastReconstrSurg 74(5):684–686.
16. Schonauer I et al (2002) Endoscopic release of intraoral synechiaein popliteal pterygium syndrome. Ann Plastic Surg. 49(5):550–552.