ORIGINAL ARTICLE

MIDLINE CLEFT OF LOWER LIP: REVIEW AND CASE REPORTS

[1]ADESINA OA
[2]EFUNKOYA AA
[2]OMEJE KU
[2]SULEIMAN AR

SUMMARY

This paper aimed to review previous articles on midline lower lip cleft and to present the three case reports seen at the University of Maiduguri Teaching Hospital, Maiduguri, Borno State, Nigeria.

Midline clefts of the lower lip are rare. They may occur as isolated cases or may be associated with other anomalies such as congenital heart defects, hand and foot anomalies, cleft palate, Van der Woude syndrome, Robin sequence, and ectopic salivary gland.

Key Words: *Midline cleft, Lower lip cleft, case reports.*

INTRODUCTION

Midline cleft of the lower lip is a rare congenital anomaly first described by Couronné in 1819 and was classified by Tessier as No 30 facial cleft.[1] Midline cleft of the lower lip is not associated with any identified hereditary factors or gender predilection.[1] Clinical presentation of these patients varies from a mild notching of the lower lip to complete midline cleavage of the inferior face, including bifurcation of the mandible, tongue and neck.[2] There is occasional, associated deformities of soft tissue structures derived from the lower branchial arches seen in these patients, there may be cleavage of the neck and manubrium sterni.[3]

Midline cleft may occur as an isolated cases or may be associated with other anomalies such as congenital heart defects[4,5], hand and foot anomalies, cleft palate, Van der Woude syndrome, Robin sequence, and ectopic salivary gland.[6-8]. The severity of the cleft has been related to the period of development of the mandibular process that is affected. Hypoplasia during the early embryonic period could result in severe median cleft involving the mandible which may extend into the neck; while disturbances later on in the embryonic period may result in less severe median clefts.[2]

[1] Adesina OA, BDS, FMCDS, Department of Oral & Maxillofacial Unit, University of Maiduguri Teaching Hospital, Maiduguri, Borno State, Nigeria.
[2] Efunkoya AA, BDS, FMCDS, Department of Oral & Maxillofacial Unit, Aminu Kano Teaching Hospital, Kano State, Nigeria.
[2] Omeje KU, BDS, FMCDS, Department of Oral & Maxillofacial Unit, Aminu Kano Teaching Hospital, Kano State, Nigeria.
[2] **For Correspondence:** Dr Suleiman Abdul Rasheed, BDS, Department of Oral & Maxillofacial Surgery, Aminu Kano Teaching Hospital, PO Box 3452, Kano, Nigeria
Email: rasheedleoras@gmail.com Tel: +2347030776567

Received for Publication: July 15, 2016
Revised: August 14, 2016
Accepted: August 15, 2016

There is no consensus on the nature and timing of the corrective surgery due to the rarity and variation in severity of the condition. The majority of the authors propose correction of the soft tissue structures as soon as possible, and mandibular bone grafting at later stages of life.[9]

The purpose of this article was to review previous literature on midline cleft of the lower lip and to present three cases seen at the University of Maiduguri Teaching Hospital, Maiduguri, Borno State, Nigeria.

CASE REPORTS

CASE 1

A 2-year-old girl presented at the oral and maxillofacial unit, University of Maiduguri Teaching Hospital, Maiduguri, Borno State, Nigeria with a history of facial disfigurement and nasal regurgitation of fluid since birth. She was the fifth child of a healthy 37-year-old mother. Her pregnancy was uneventful and the baby was delivered by spontaneous vaginal delivery at 36th gestational week. The mother who was the informant gave a history of no febrile illness, or exposure to any teratogenic drugs during the pregnancy period. There was no relevant information obtained from the family history, which was considered contributory.

Clinical evaluation revealed normal contour of the upper face and head. The upper lip was intact however, there was a midline cleft of the lower lip, which was obvious. Palpation of the mandible did not reveal any discontinuity in the mid symphyseal region or abnormal mobility of any region. There was a cystic mass in the midline extending from the floor of the mouth across the mandibular symphysis and extending into a skin tag at the chin (Fig 1). Intraorally the tip of the tongue

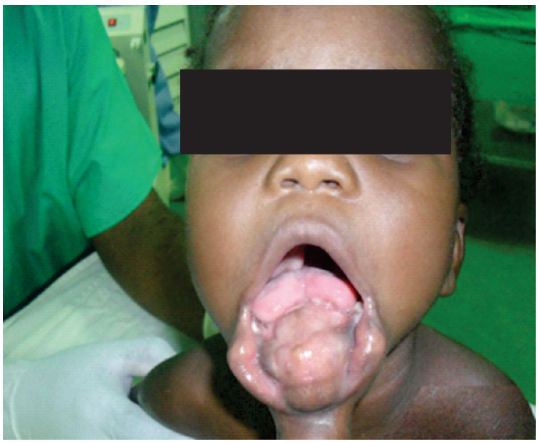

Fig 1: Median cleft of lower lip and ankyloglossia

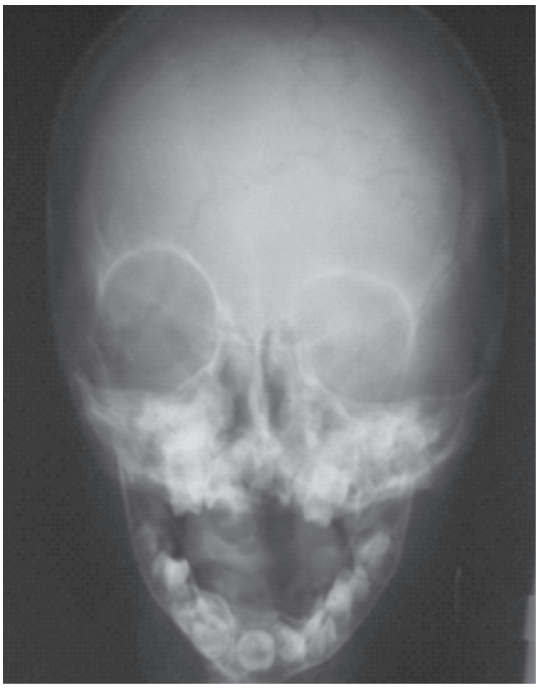

Fig 2: Radiogragh shows molar like teeth in symphyseal region of the mandible

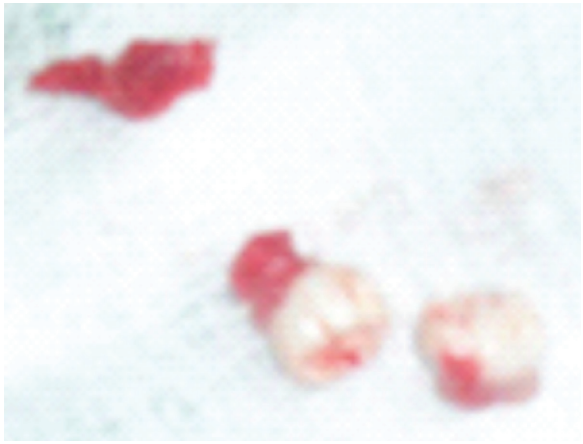

Fig 3: Molar like teeth extracted from the cyst in the symphyseal region of the mandible

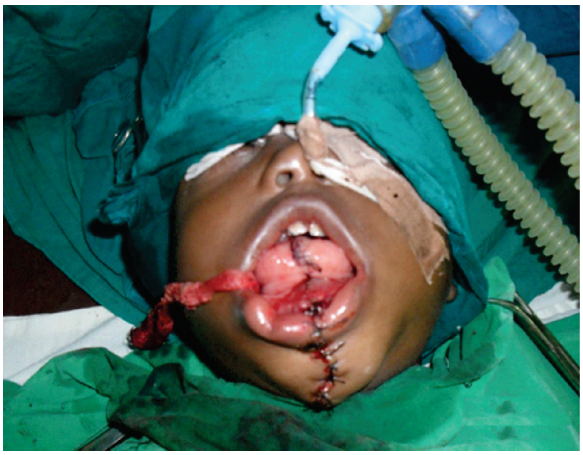

Fig 4: Repair of the tongue

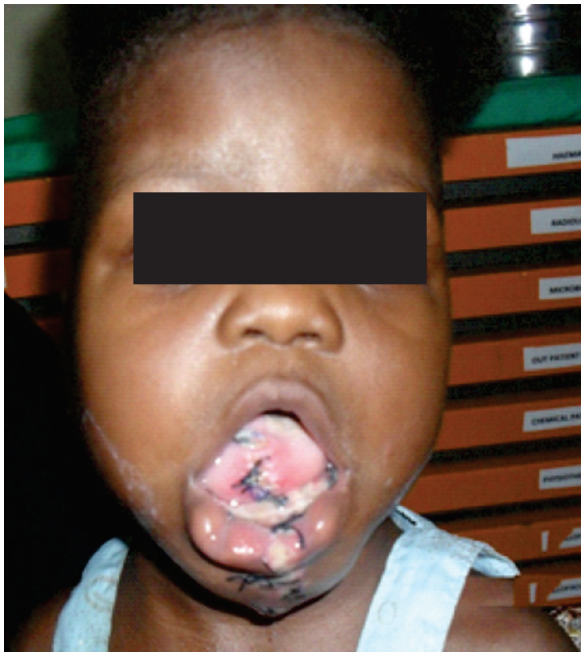

Fig 5: Appearance of patient on 5th post-operative day

was bifid and fixed to the floor of the mouth, the incisors were missing and there was an obvious palatal cleft.

Radiological investigation revealed no defect of the mandible at the midline. Radiopacities simulating molar- like tooth structures and bony projections were seen at the symphyseal region (Fig 2). Lower incisors were not visible radiologically. Relevant basic laboratory tests were within normal ranges. Diagnosis of midline cleft of the lower lip and tongue and palate with associated dermoid cyst was made. Patient's treatment was planned in two stages, repair of lower lip, tongue and dermoid cyst enucleation in the first stage with palatal repair at a later period.

Surgical treatment was carried out under general anaesthesia, routine preparation of patient was done including institution of throat pack. The cystic mass was enucleated while the molar- like teeth were removed

and the bony process excised (Fig 3). The tongue was released, and a frenectomy of the lingual frenulum with repair of the tongue was performed (Fig 4). The lower lip defect was also repaired in layers with a combination of 3/0 vicryl and 3/0 braided silk suttures (Fig 5).

The excised specimen revealed a cystic lesion consisting of thick fleshy covering, with associated the molar like teeth and a bony tissue. Histopathological examination revealed a cyst lined by keratinizing squamous epithelium with attached pilosebaceous structures suggestive of dermoid cyst.

CASE 2

A 50-year-old female patient presented to the maxillofacial clinic with a complaint of deformity of the lower lip. There was no positive history of cleft lip/palate in the family. Examination revealed cleft of the lower lip involving the vermillion and white roll, the upper lip and nose appeared intact. Intraorally the cleft was noted across the vermillion. The alveolus, lingual frenum, tongue and palate were intact. Relevant basic laboratory investigations were within normal range. A diagnosis of midline cleft of the lower lip was made.

Surgical treatment, which was performed under local anaesthesia, included a "V" shaped excision of the cleft with direct-layered closure of the lip using a combination of 3/0 vicryl and 3/0 braided silk sutures.

CASE 3

A 30-year-old male cattle rearer presented to the maxillofacial clinic with a complaint of deformity of the lower lip. There was no positive history of cleft lip/palate elicited in the family. Clinical examination revealed cleft of the lower lip involving the vermillion and white roll, the upper lip and nose appeared clinically normal. Intraoral examination showed that the cleft was across the vermillion. The alveolus, lingual frenum, tongue and palate were clinically normal. Relevant basic laboratory investigations were within normal range. Surgical treatment, which was performed under local anaesthesia, included a "V" shaped excision of the cleft with direct-layered closure of the lip using a combination of 3/0 vicryl and 3/0 braided silk sutures.

DISCUSSION

Developmental anomalies of structures derived from the upper half of the first branchial arch are common, giving rise to deformities such as cleft lip or cleft palate.[10] However, abnormal or incomplete development of structures derived from the lower half of the first branchial arch is rare.[11] These anomalies may arise from failure of fusion of the first pair of branchial arches or a failure of mesodermal penetration into the midline of mandibular part of the first branchial arch presenting as a complete or incomplete cleft of the lower lip, mandible, and tongue with occasional associated deformities of soft tissue structures in the neck derived from the lower branchial arch.[12]

Proposed pathogenesis of median clefts of the lower lip and mandible include, hypoplasia of the mandibular processes either during the early embryonic period or during late embrayonic periods. Hypoplasia in the early embrayonic period results in severe mandibular clefting unlike hypoplasia during the late period which results in less severe median clefts. The cleft of the lower lip, ranges from a notch of the mucosa to a complete cleft of the lip. This clefting may be related to the insufficient outgrowth of the lip, most probably due to incomplete merging of the mandibular processes. The bifid tongue may be explained by the persistent intermandibular groove, similar to a merging defect of the tongue. The mandibular cleft is also thought to result in impairment of the outgrowth of the osteogenic centers of the definitive mandible, resulting in absence of its symphysis.[2]

There is a broad variation in the severity of this deformity, ranging from a simple notch in the mucosa to complete cleavage of the lower lip involving the tongue, chin, mandible, neck, and manubrium sterni. The tongue involvement may range from a bifid anterior tip with ankyloglossia and adhesion to the mandibula cleft margins, up to complete tongue hypoplasia. The incisor teeth are frequently missing at the mesial mandibular margins. The hyoid bone and thyroid cartilage may be cleaved or completely absent.[11] Other malformations, which may be present, include congenital heart deformities, as well as cleft palate.[8] The severity or extent of the cleft has been related to the period of development of the mandibular process that is affected. Hypoplasia during the early embryonic period could result to severe clefting of the mandible extending into the neck, while disturbances during the late embryonic period may result in less severe median clefts.

Dermoid cysts are unusual neoplasms that often present in childhood, with the orbit being the area most commonly affected in the head and neck region.[13] The causes of both epidermoid and dermoid cysts include failure of surface ectoderm to separate from underlying structures, sequestration and implantation of surface ectoderm.[14] Most congenital dermoid and epidermoid cysts probably arise due to an embryologic accident during the early stages of development, between 3 and 5 weeks of gestation. Enclosed ectodermal cysts can occur when the surface ectoderm fails to separate completely from the underlying neural tube. Alternately, they may result from abnormal sequestration or invagination of surface ectoderm along the embryologic sites of dermal fusion that form the eyes, ears, and face.[14] The association with the midline cleft remains a subject of on going debate.

In the case presented there was cleft of the lower lip, missing incisors, bifid tongue with ankyloglossia, cleft palate with a dermoid cyst. However the absence of defect in the mandible makes the diagnosis to be an incomplete median cleft.

Concerning treatment, the rarity and variation in severity of the condition may be responsible for the lack of a consensus on the nature and timing of the corrective surgery. The majority of the authors propose correction of the soft tissue structures as soon as possible, so as to limit feeding and/or speaking problems and mandibular bone grafting in later stages of life. The timing of treatment of mandibular clefts by most surgeons is when the child is about 8 to 10 years to avoid damaging developing tooth buds.

Armstrong and Waterhouse have suggested that reconstruction should be done after the age of 10 years to avoid damaging developing tooth buds. They recommended a protocol of deferment of second surgery to age of 10 years in cases with no gaps between the mandibular halves and an early attempt to stabilize the mandible with bone grafts or reconstruction plates in cases with feeding, breathing difficulties or where the mandibular segments are mobile.[10] Seyhan freshened the edges of mobile bone segments and fixed with stainless steel wires in a 10 months old child with good result.[7,15] Sherman and Goulian[8] reported one-stage reconstruction using rib graft at 20 months age.[5] However at the 1973 cleft congress in Copenhagen, a case of complete midline cleft was presented with the mandibular defect being grafted at three months of age and the lip was repaired at six months and growth and development still progressed normally.

We recommend correction of Soft tissue deformities of the lower lip, tongue and neck as early as 3 months of age. Bone graft with a rib graft or iliac crest should follow when the baby is under 2 years of age (before speech development). However earlier attempt should be made to stabilize the mandible with either bone graft or reconstruction plate in patients with respiratory or feeding problems.

REFERENCES

1 Couronné (of Rouen) A. Observation d'un bec-de-lièvre, compliqué de fentes à la voûte palatine. Ann Clin Soc Med Prat Montepellier. 1819; (2s)6: 107.

2 Freitas RS et al. Tessier number 30 clefts: surgical correction of a rare malformation. Rev Bras Cir Craniomaxilofac 2010; 13(1): 12-17.

3 Constantinides CG, Cywes S. Complete median cleft of the mandible and aplasia of the epiglottis. SA Med J 1983; 64: 20: 293-94.

4 Oostrom CAM, Vermeij-Keers C, Gilbert PM, et al: Median cleft of the lower lip and mandible. Case reports, a new embryologic hypothesis, and subdivision. Plast Reconstr Surg 1996; 97: 313-20.

5 Sherman JE, Goulian D: The successful one-stage surgical management of a midline cleft of the lower lip, mandible and tongue. Plast Reconstr Surg 1980; 66: 756-59.

6 Gardner DG, Kapur RN, Jordan RE. A cleft of the mandible in the lateral incisor-cuspid region. J Oral Surg 1973. 35: 649-52.

7 Richieri-Costa A, Pereira SCS. Autosomal recessive short stature, Robin sequence, cleft mandible, pre/postaxial hand anomalies, and clubfeet in male patients. Am J Med Genet 1993. 47: 707-09.

8 Lekkas C, Latief BS, Corputty JEM. Median cleft of the lower lip associated with lip pits and cleft of the lip and palate. Cleft Palate Craniofac J 1998. 35: 269-71.

9 Almeida LE, Ulbrich L, Togni F. Mandible Cleft: Report of a Case and Review of the Literature. J Oral Maxillofac Surg 2002; 60: 681-84.

10 Leela Krishna Guttikonda, Koteswara Rao Nadella, Vijayalakshmi Uppaluru, Rama Mohan Kodali, and Ranganadh Nallamothu Nonsyndromic Mandibular Symphysis. Case Rep Dent. 2014; 2014: 682163. Published online Mar 3, 2014.

11 Monroe CW. Midline cleft of the lower lip, mandible and tongue-with exion contracture of the neck: case report and review of the literature. Plast. Reconstr. Surg. 1966; 38(4): 312-19.

12 Renato da Silva Freitas, Gilvani Azor de Oliveira e Cruz, Nivaldo Alonso, Antonio Jorge Forte, Luciano Busato, John Persing; Tessier number 30 clefts: surgical correction of arare malformation. Craniomaxilofac 2010; 13(1): 12-17.

13 Pryor SG, Lewis JE, Weaver AL, Orvidas LJ Pediatric dermoid cysts of the head and neck Otolaryngol Head Neck Surg. 2005 Jun; 132(6): 938-42.

14 Orcun Toptas, Ismail Akkas Mustafa Tek, Fatih Ozan, and Cetin Boran Intraosseous Epidermoid Cyst Associated with Impacted Mandibular Wisdom Teeth: An Uncommon Entity. J Clin Diagn Res. Jul 2014; 8(7): ZD31–ZD32. Published online Jul 20, 2014. doi: 10.7860/JCDR/2014/9413.4630

15 Seyhan T, Kylyur H. Median cleft of lower lip: Report of two newcases and review of literature. Ann Otol Rhinol Laryngol 2002; 111: 217-21.