OPEN

PRS GLOBAL OPEN

CASE REPORT

Craniofacial/Pediatric

Midline Mandibular Cleft Repair: A Case Report

Wael Fawzy Ismaiel, MD, PhD*
Mohamed Hussein Abdelazim, MD*
Ahmed Younes, MD*
Abdelgawad M. Hadeya, MD*
Mahmoud Ahmed Amin, MD†

Summary: Midline clefts of the lower lip and mandible are extremely uncommon. Couronne reported the first case in 1819, and fewer than 80 cases have been discovered in the literature to date. This case report describes a female infant who was born having a full midline cleft of the mandible and lower lip, as well as neck flexion contracture that extended from the lower lip to the manubrium sterni. A fenestrated type of acrylic splint was applied over the mandibular arch and fixed by circum-mandibular wiring. Fixation was done using a 0.5 interosseous wire passing through the prepared hole and tightened over a previously harvested rib bone graft.
(Plast Reconstr Surg Glob Open 2022;10:e4283; doi: 10.1097/GOX.0000000000004283; Published online 25 April 2022.)

Midline clefts of the lower lip and mandible are very uncommon. They are thought to have resulted from the failure of mesodermal penetration and fusion of paired mandibular processes.[1]

Regarding craniofacial clefts, Paul Tessier established a numbering system in 1976, with the mandible midline cleft being designated as number 30 and characterized by a branchiogenic midline syndrome, intermandibular dysplasia, and a mandibular cleft. There were 14 caudal extensions among them.[2]

Mandibular midline clefts can occur in conjunction with lip midline clefts. Lower lip clefts have been reported to be associated with neck and midline cervical cord flexion contracture.[3]

CASE REPORT

The patient's history revealed that upon birth, the parents noticed the cleft lip and cleft mandible.

At the age of 2 months, the patient had achieved a successful lip repair. Over the next 4 years, the parents have sought many centers to find a solution for such a deformity, with no medical or surgical advice of benefit.

We evaluated the patient clinically and radiologically (Fig. 1). We detected a large irregular defect of the mandible and deformed lower incisors. A detailed workup was conducted to screen any associated anomalies.

*From the *Otorhinolaryngology Department, Faculty of Medicine, Al-Azhar University, Damietta, Egypt; and †Plastic Surgery Department, Faculty of Medicine, Al-Azhar University, Damietta, Egypt.*

Received for publication January 27, 2022; accepted March 7, 2022.

DOI: 10.1097/GOX.0000000000004283

After consulting a plastic surgeon, we decided to repair the cleft. The father of the patient provided written informed consent to perform this surgery.

METHODS

Preoperative Preparation

Preoperative preparation of the patient was done by the fenestrated type of acrylic splint for the mandibular arch. The surgery was performed under general anesthesia, using nasotracheal armored intubation.

Operative Technique

The cleft line was exposed through an intraoral approach via a vestibular mucosal incision extending from the right first premolar to the left first premolar, exposing the cleft line and both mental nerves. Division of the lower lip through the midline, for more exposure of the mandible, was done to facilitate the surgery. A fenestrated type of acrylic splint was applied over the mandibular arch and fixed by circum-mandibular wiring.

Refreshment of both bony edges of the mandibular cleft was carried out followed by refreshment of the anterior surface of the mandible between both mental nerves, using a small round bur (Fig. 2), followed by a hole in the mandibular lower border on both cleft sides to be able to fix and insert the cancellous bone graft that was harvested from the rib. Afterward, fixation was done using a 0.5 interosseous wire passing through the prepared hole and tightened over the bone graft (Fig. 3). Finally, closure of the incision was done with Virile 3-0.

Postoperative Follow-up

The fenestrated type of acrylic splint was removed 1 month after the operation. A 3D radiography was

Disclosure: *The authors have no financial interest to declare in relation to the content of this article.*

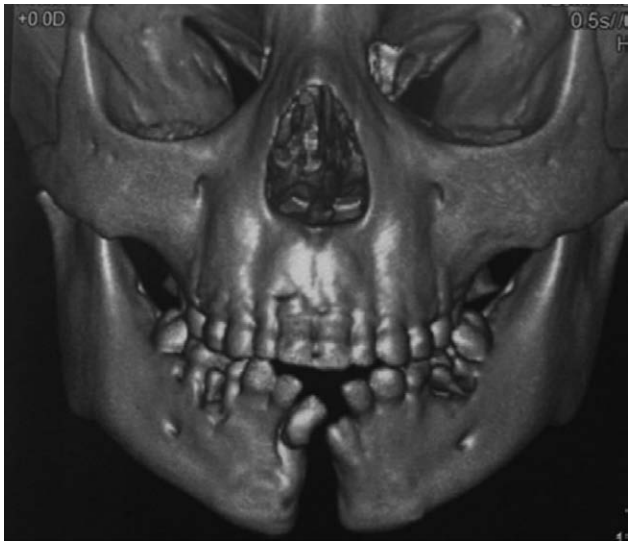

Fig. 1. High-resolution CT scans axial and coronal cuts with 3D radiography demonstrate the site of the defect.

performed to demonstrate the complete closure of the cleft by the age of 7 (Fig. 4).

DISCUSSION

The most frequent types of orofacial developmental anomalies are those affecting the upper lip and face. Midline mandibular cleft is a very unusual abnormality, with fewer than 75–80 cases identified globally, deeming it valuable to emphasize whenever it happens.[2]

The majority of the researchers believe that the problem develops as a result of the first branchial arch failure to fuse or due to insufficient midline mesodermal penetration. This also explains why severe cases lack the manubrium sterni, strap muscles, and hyoid.[2]

Two mandibular processes develop within this first branchial arch, separated in the midline by a groove. During the late embryonic phase, these mandibular processes combine and do not fuse (crown-rump length: ≥17 to ≤60 mm). During the same embryonic stage, the alveolar process and the lip are formed together with the anlage and the outgrowth of one membrane bone center

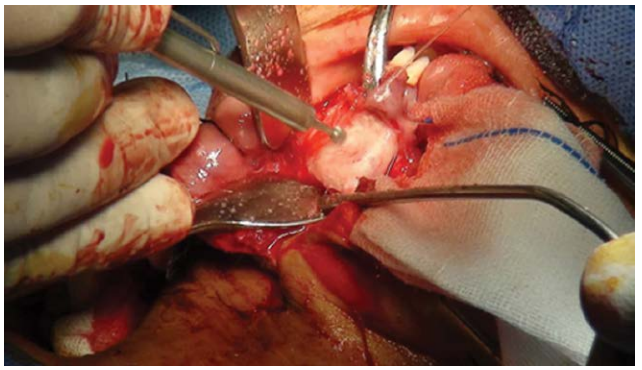

Fig. 2. Refreshment of the anterior surface of the mandible between both mental nerves, using small round bur.

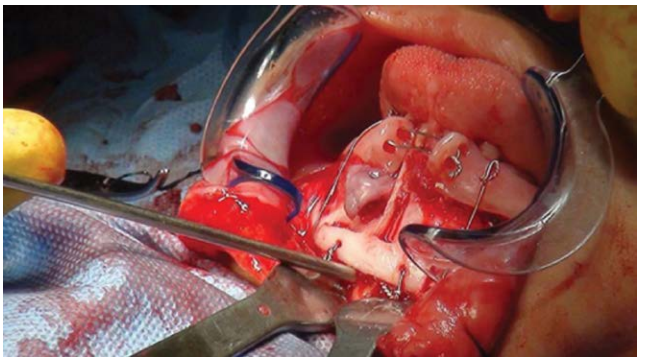

Fig. 3. Surgical reconstruction of the mandibular cleft.

in each mandibular process. As a consequence, the mandible with its symphysis is formed[3] Moreover, it is hypothesized that, during the early embryonic stage, hypoplasia of the mandibular process results in the severest mandibular cleft extending into the neck. Median clefts of less severity occur in the late embryonic stage.

As a result, the clinical manifestations might be minor or severe. At the vermilion border, a simple notching may be present. Chin and lower lip can be bifid, with no bone gap in certain situations. The anterior tongue aspect may be divided and connected with a fibrous band to the mandibular cleft borders. Armstrong reported ankyloglossia and bifid uvula.[4]

Armstrong and Waterhouse propose that staged reconstruction is the most frequently used surgical protocol worldwide.[4] The mandible is reconstructed using rib and iliac bone grafts, as well as titanium or bioresorbable miniplates and miniscrews.[1] Seyhan and Kýlýnr[5] successfully restored the margins of the mobile mandibular segments in a 10-month-old infant, and secured them using stainless steel wires. The child's most recent follow-up, at the age of 7 years, has revealed normal neck extension and a stable mandible with normal speech and normal occlusion.

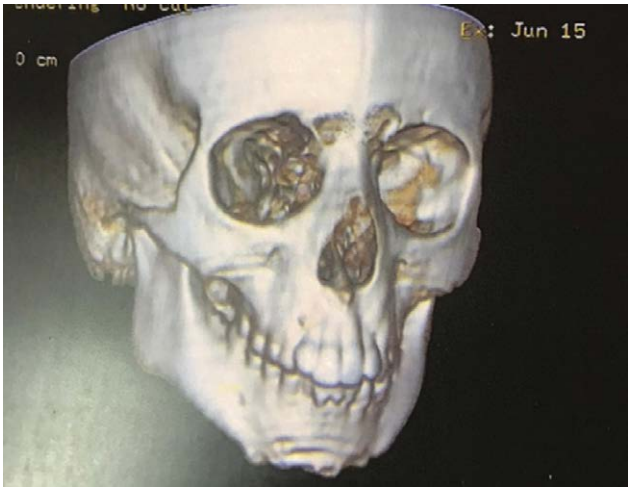

Fig. 4. Postoperative 3D radiograph showing complete repair of the cleft at 7 years old.

CONCLUSIONS

Mandibular midline clefts and the accompanying clinical abnormalities are very rare congenital facial malformations. Neck flexion contracture and hypoplastic neck muscles were seen in this patient. This study records a well-defined management strategy, effective surgical repair, and long-term follow-up of the patient.

Wael Fawzy Ismaiel, MD, PhD
Otorhinolaryngology Department
Faculty of medicine, Al-Azhar University
Damietta 11651
Egypt
E-mail: dr_mallah@hotmail.com;
dr_mallah@domazhermedicine.edu.eg

REFERENCES

1. Rao AY. Complete midline cleft of lower lip, mandible, tongue, floor of mouth with neck contracture: a case report and review of literature. *Craniomaxillofac Trauma Reconstr.* 2015;8:363–369.
2. Schutte BC, Murray JC. The many faces and factors of orofacial clefts. *Hum Mol Genet.* 1999;8:1853–1859.
3. Prakash A, Parelkar SV, Oak SN, et al. Giant epignathus with midline mandibular cleft: insights in embryology and management. *Ann Maxillofac Surg.* 2012;2:56–59.
4. Armstrong AP, Waterhouse N. Tessier 30 median mandibular cleft: case report and literature review. *Br J Plast Surg.* 1996;49:536–538.
5. Seyhan T, Kýlýnr H. Median cleft of the lower lip: report of two new cases and review of the literature. *Ann Otol Rhinol Laryngol.* 2002;111(3 Pt 1):217–221.