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SHORT COMMUNICATION

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Mini-microform cleft lip with complete cleft alveolus and palate: A case report

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Abstract

Cleft lip and cleft alveolus are caused by incomplete fusion of the frontonasal and maxillary prominences. However, milder forms of cleft lip are rarely accompanied by cleft alveolus. Here, we report a rare case of mini-microform cleft lip with complete cleft alveolus and cleft palate. No findings suggestive of cleft lip were evident on initial examination. However, three-dimensional facial measurements confirmed the presence of cleft lip despite no evidence of orbicularis oris muscle (OOM) rupture on ultrasonography. Collapsed nostril, as observed in this case, is usually associated with OOM rupture. However, it can also be caused by skeletal abnormalities, such as cleft alveolus. Three-dimensional facial measurements and ultrasonography can assist in accurate diagnosis when visual examination is ambiguous.

KEYWORDS
cleft alveolus and palate, facial measurements, microform cleft lip, orbicularis oris, ultrasonography

1 | INTRODUCTION

Cleft lip and/or cleft palate are frequently occurring external anomalies and have shown higher incidence rates in the Japanese population than that in other ethnic groups.[1-3] Embryologically, incomplete fusion of the facial prominences; specifically, incomplete fusion of the frontonasal and maxillary prominences could lead to cleft lip and/or cleft alveolus.[4,5] A complete cleft lip is caused by a complete failure of fusion between the frontonasal and maxillary prominences, whereas incomplete fusion in some areas can result in an incomplete cleft lip. Milder forms of incomplete cleft lip are the microform cleft lips or congenital "healed" lips,[6] and they are classified by Yuzuriha and Mulliken into minor-form, microform, and mini-microform according to the extent of lip dysjunction.[7] However, such milder forms of cleft lip are rarely accompanied by cleft alveolus, and no case of mini-microform cleft lip, which is the mildest form of cleft lip, accompanied by cleft alveolus has been reported.[7] Embryologically, cleft lip and cleft alveolus originate from the failure of fusion between the same prominences. However, the reason for cleft alveolus not accompanying cleft lip is unclear.

Milder forms of cleft lip, such as mini-microform cleft lips, are difficult to diagnose solely based on surface morphology. Even in cases of the slightest suspicion of incomplete fusion between the prominences, surgery should be considered as the patient may develop deformities of the lips and nose in the future.[8] Milder forms of cleft lip can be diagnosed by identifying asymmetries in nose width and/or nostril morphology, rather than by observing deformations of the upper lip alone. In recent years, three-dimensional (3D) measurement technology has been used for diagnosis when visual examination alone could not be used to analyze this asymmetry. Furthermore, analysis of the orbicularis oris muscle (OOM) for evidence of rupture can substantiate the presence of incomplete fusion.[9]

Here, we report a rare case of mini-microform cleft lip with complete cleft alveolus wherein 3D measurements of the lips and nostrils and analysis of the OOM using ultrasonography were performed to aid diagnosis.

2 | MATERIALS AND METHODS

Ethical approval for the publication of the case was granted by the Research Ethics Committee at the University of Toyama; all procedures were performed according to the provisions of the Declaration of Helsinki (as revised in Fortaleza, Brazil, October 2013). The male patient was born at 37 weeks of gestation by vaginal birth. The birth weight was 2.714 g, and the Apgar score was 9/10. Family history was unremarkable. A cleft palate was observed after birth without other congenital disease, and the patient was referred to our department at the age of 4 days. Initial examination revealed left-sided cleft alveolus and cleft palate continuous with the cleft alveolus, but no signs of left-sided cleft lip were evident. Hence, we decided to monitor the patient's clinical course (Figure 1). No feeding problems were evident, and the patient underwent palatoplasty at the age of 1 year and 2 months. Thereafter, orthodontic treatment was initiated, and computed tomography (CT) showed complete cleft alveolus, curved nasal septum, and deviation of the subnasale toward the healthy side (Figure 2). The patient underwent secondary bone grafting at 9 years of age, and CT after 1 year showed continuity between the alveolar bone segments and sufficient bone formation at the base of the nostrils (Figure 2). When the patient was 11 years old, 3D measurements of the patient's lip and nose morphology and detailed examinations of the form of the OOM were performed to assess the presence of cleft lip. Written informed consent was obtained from the parent of the patient for publication of this Case report including all the patient data.

2.1 | Three-dimensional facial measurements using 3D photogrammetry imaging

The patient's face was imaged using Vectra H1 (Canfield Scientific, Parsippany, NJ), and a 3D image was constructed using the images obtained by the camera and Vectra imaging software. The points of measurements on the constructed 3D image were the alar base (al), crista philtra superius (cphs), crista philtra inferioris (cphi), subnasale (ns), and cheilion (ch), as suggested by Jodeh and Rottgers.[10] The alar-base width (al-sn), philtrum length (cphs-cphi), and lateral lip length (ch-cphi) (Figure 3).

2.2 | Evaluation of the OOM

We used ultrasonography (SonoSite M-Turbo, Fujifilm Sonosite; Bothell, WA) to observe the course of the OOM fibers in the white roll of the lips. We placed the probe on the white roll of the healthy side to observe the course of OOM fibers between the skin and alveolar ridge. The continuity of the muscle layer was verified, and the probe was moved to the diseased side to determine the presence of any ruptures in the muscle layer (Figure 4).

3 | RESULTS

1. The philtrum length (cphs-cphi) was lesser on the left side, and the alar-base width (al-sn) was greater on the left side. The lateral lip length (ch-cphi) was greater on the left side. The distances between the labrale superius and cphi were same on both left and right sides (Table 1).
2. Ultrasonography confirmed the continuity of the OMM, and no ruptures or discontinuities were evident.

4 | DISCUSSION

4.1 | Possible new type of cleft

Cleft lip and cleft alveolus are caused by incomplete fusion of the frontonasal and maxillary prominences. Hence, the two clefts are embryologically the same.[1-5] Clinically, several cases of cleft lip without cleft alveolus have been reported, but no case of complete cleft alveolus without cleft lip, even microform cleft lip, has been reported. Nagase et al. showed 102 patterns of cleft lip and/or cleft palate in Japanese

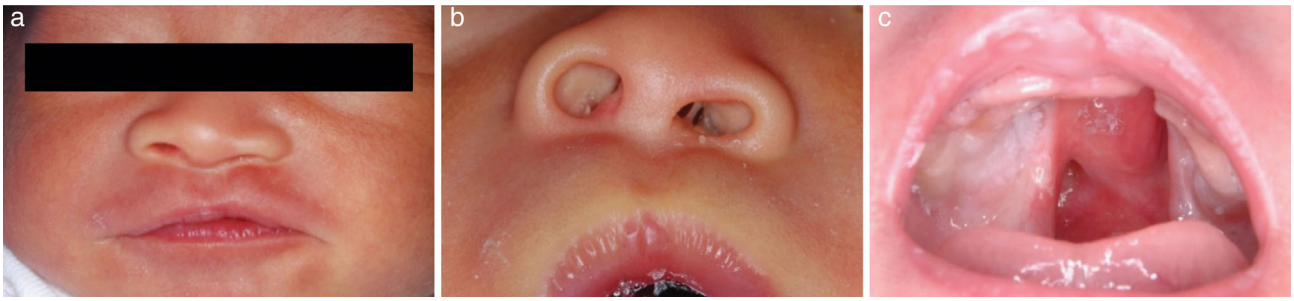

FIGURE 1 Age: 8 days (first visit). A, No obvious depression or cleft from the upper lip to the nose. B, No apparent difference in nostril width. A difference in nostril position can be seen. C, Cleft alveolus and cleft palate

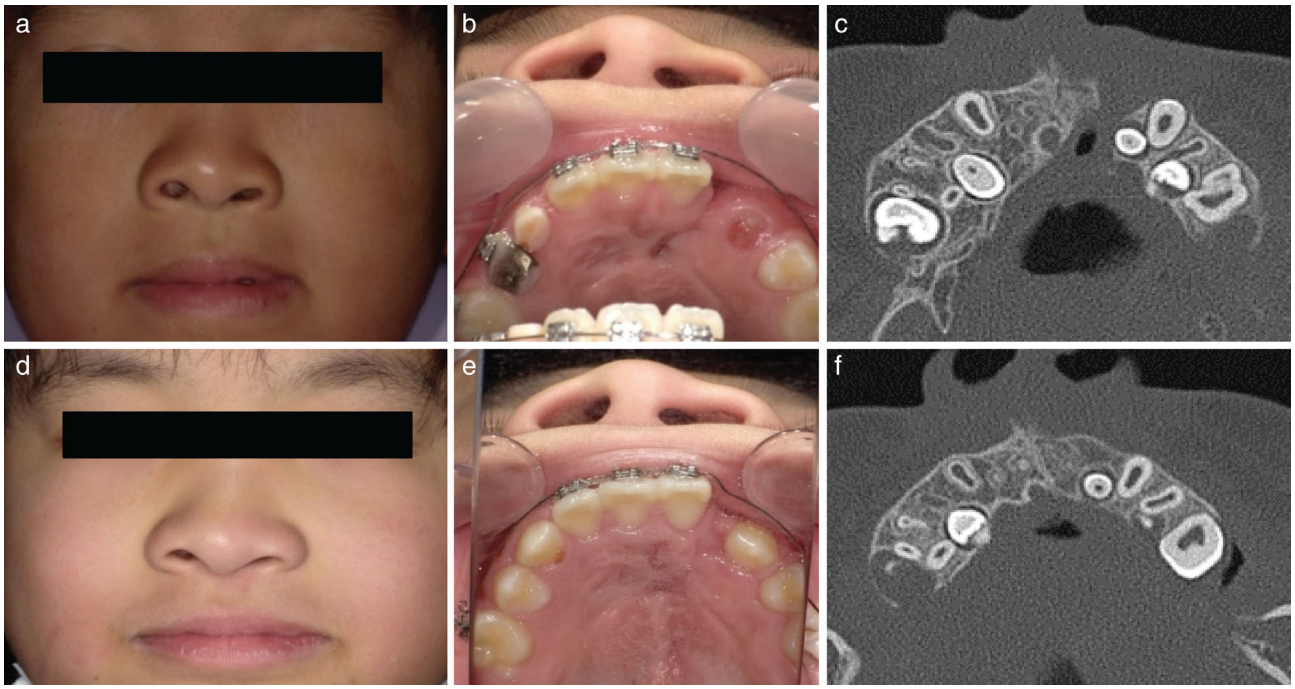

FIGURE 2 Before secondary bone grafting (age: 9 years). A, No obvious depression or cleft from the upper lip to the nose. B, Cleft alveolus on the left side. C, Computed tomography scan: An alveolar bone defect is seen on the left side. After secondary bone grafting (Age: 11 years). D, No obvious depression or cleft from the upper lip to the nose. E, No disruption in the continuity of the alveolus. F, Computed tomography scan: Continuous alveolar bone is seen

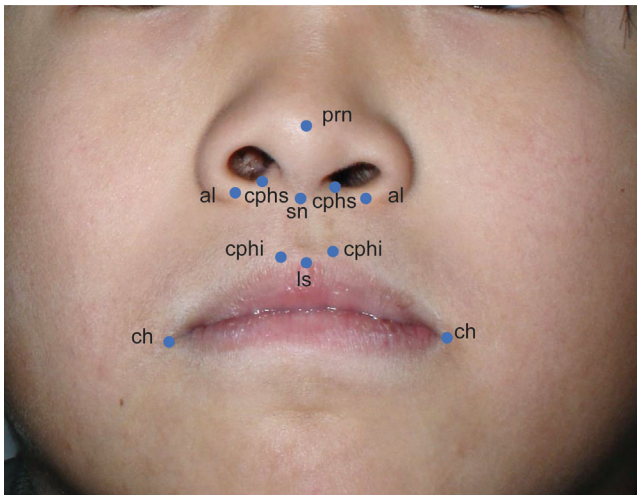

FIGURE 3 Anthropometric anatomic landmarks and distances. *al*: alar base; *ls*: labrale superius; *ch*: cheilion; *cphs*: crista philtri superius; *cphi*: crista philtri inferioris; *prn*: pronasale; *sn*: subnasale

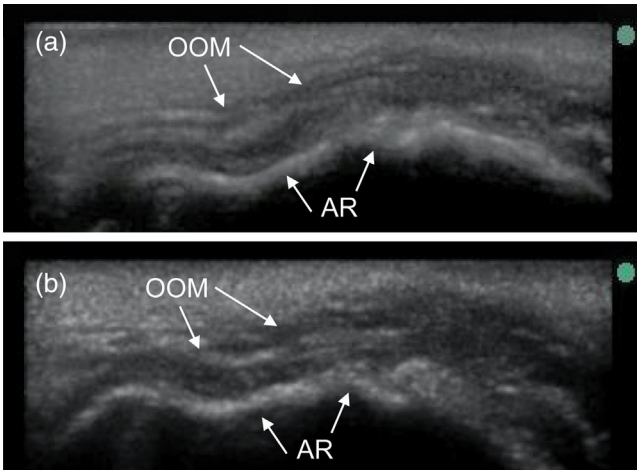

FIGURE 4 Ultrasound images of the orbicularis oris muscle. A, Right (normal) side; B, left side. AR, alveolar ridge; OOM, orbicularis oris muscle

patients using 17 classification codes.[11] However, even they did not report any case of cleft alveolus without cleft lip, minor cleft lip, or cleft palate. Hence, we performed facial measurements and OOM analysis to determine the presence of cleft lip. Nagase et al. reported that cases with minor cleft lip are rarely accompanied by cleft alveolus, and no case of the mildest form of cleft lip, the mini-microform cleft lip, accompanied by cleft alveolus, has been documented. The only case with coexisting cleft palate had a velar-type cleft palate.[7] However, our patient showed a new cleft type: complete cleft alveolus accompanied by cleft palate with a mini-microform cleft lip. Furthermore, familial occurrence of microform has been reported, and Suzuki et al [15] reported the involvement of the *BMP4* gene. Although there was no obvious family history in this case, it seems necessary to investigate the genetic involvement in this new cleft type in the future.

During development of the primary palate, the epithelial tissue in the area where the maxillary and medial nasal segments come into

TABLE 1 Three-dimensional facial measurements

	cphs-cphi			al-sn			ch-cphs		
	R (mm)	L (mm)	L/R	R (mm)	L (mm)	L/R	R (mm)	L (mm)	L/R
Control	17.80	17.86	1.00 ± 0.04	12.33	12.40	1.01 ± 0.07	25.40	24.70	0.98 ± 0.04
Case	11.64	13.8	0.84	13.37	11.63	1.15	25.58	22.26	1.15

Abbreviations: al, alar base; ch, cheilion; cphs, crista philtri superius; cphi, crista philtri inferioris; sn, subnasale; L, left side; R, right side; L/R, left side/right side.
left side is shorter than right side; L/R > 1: left side is longer than right side; L/R < 1: left side is the same length as right side; L/R=1.

contact with each other is depressed, and fusion is completed when the depressed epithelium is gradually pushed out by the proliferation of mesenchymal tissue. Therefore, if the mesenchymal tissue does not proliferate and epithelial depression progresses, cleft lip occurs.[8] Since the epithelial depression progresses from the surface of the face to the oral cavity, it can be inferred that it is highly unlikely that a complete cleft alveolus will develop in the case of an incomplete cleft lip or microform with a slight epithelial depression. The embryological results suggest that the cleft type of this case may be very rare.

4.2 | Cause of collapsed nostril in microform clefts

Collapsed nostrils have been reported as a characteristic of minor cleft lip.[8] Such collapsed nostrils are assumed to be caused by some abnormalities in the soft tissue, such as OOM rupture.[8] In this patient, neither OOM rupture nor microform cleft lip was evident during the initial examination at birth. However, as the cleft alveolus was clearly evident, we thoroughly examined the patient's left-right asymmetry by 3D facial measurements. Camison et al reported the accuracy of 3D facial measurements using a Vectra H1[12] and suggested this technique to examine the postsurgical scar formation, relapse, and asymmetry in patients with cleft lip and/or cleft palate.[10,13,14] Bagante et al[14] measured the facial features of unaffected children using the Vectra and reported that the length of the white roll of the lip on the right (left) side and the lateral lip length to the Cupid's bow on the right (left) side are almost similar in unaffected children. This may mean that the left/right ratio is close to 1 in unaffected children, but further study is needed to determine what value is necessary for the diagnosis of microform cleft lip. However, in this case, the left/right ratio of the length of the white roll of the lip was 0.84 and that of the length to Cupid's bow was 1.15, which clearly shows a difference between the left and right sides. Facial measurements of this patient revealed that the white roll was shorter and the nostril was wider on the diseased side, which prompted us to analyze the presence of microform cleft lip. As several reports have suggested that ultrasonography can be used to observe the presence of OOM ruptures, even in cases of microform clefts where the cleft lip is not evident by visual examination,[8,9,15] we used this method to examine the patient's OOM. We verified the presence of muscle continuity without any evidence of ruptures. Clinically, there was no disorder in the movement of the OOM, and the findings at the time of bone grafting did not show any OOM abnormally attached around the columella base and the nasal alar base. The findings of our report showed that collapsed nostril, as seen in this case, which is a characteristic of OOM rupture, can also result from a skeletal abnormality such as cleft alveolus, rather than being an exclusive feature of soft tissue abnormalities, such as OOM rupture.

We encountered a rare case of mini-microform cleft lip with complete cleft alveolus and cleft palate. We obtained 3D lip-nostril measurements and analyzed the OOM for evidence of rupture using ultrasonography. Though the presence of a collapsed nostril suggested the presence of microform cleft lip, no OOM rupture was observed. This study showed that a collapsed nostril in cases of microform cleft lip accompanied by cleft alveolus may be caused by skeletal abnormalities such as cleft alveolus, rather than by soft tissue abnormalities, such as OOM rupture.

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DISCLOSURE OF INTEREST

None.

DATA AVAILABILITY STATEMENT

All data generated or analyzed during this study are included in this published article.

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