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Median alveolar cleft and palatal mass without a median upper cleft lip

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ABSTRACT

Median cleft is rare among facial clefts, including bilateral and unilateral clefts. Median upper cleft lip and median alveolar cleft correspond to Type 14 and Type 0, respectively, in Tessier's classification system. Some authors have reported surgical procedures for median cleft. In the case of median alveolar cleft, bone grafting to the cleft side and orthodontics are generally applied, similarly to bilateral or unilateral cleft. Median alveolar cleft is usually accompanied by median upper cleft lip, the degree of which differs in each case. The symptoms include, but are not limited to, median lip defect, wide philtrum, and vermilion notch. However, an isolated alveolar cleft is extremely rare. We encountered a patient with an isolated alveolar cleft who did not have a light median upper cleft lip, such as a wide philtrum or vermilion notch. We herein report this case and describe its treatment.

1. Introduction

In comparison to bilateral and unilateral cleft, median cleft is a rare deformity. Davis and Fogh-Anderson reported that the median cleft was observed in 0.73% and 0.43% of cleft lips, respectively [1,2]. Another study reported that the frequency of rare craniofacial clefts was 1.4–4.9 per 100,000 live births [3]. Various classifications are used for median clefts. Veau described three varieties of median cleft: a notch, a median cleft extending to the columella, and a defect due to lack of development of the whole medial element [4]. However, Millard proposed that vertical cleft through the center of the upper lip, regardless of the extent, should be classified as a median cleft lip [5]. The median cleft is distinguished into two types: true median cleft and false median cleft. True median cleft is accompanied by other abnormalities, such as polydactyly and hypertelorism. False median cleft was proposed that was associated the deformities of median cleft lip, nasal malformation, and orbital hypotelorism [6]. The false type is a kind of holoprosencephaly and it is difficult for patients to achieve long-term survival [7]. In this case report, we describe the treatment of a case of median alveolar cleft with a palatal mass and without an upper cleft lip.

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1.1. Case report

A 4-year-old girl with median alveolar cleft visited the department of Plastic and Reconstructive Surgery in Kyoto University Hospital. She had the large space between the central maxillary incisor and a palatal mass in the cleft (Fig. 1A and B) without a median upper cleft lip (Fig. 2A). She had a past history of epilepsy, but no other diseases or abnormalities. The patient sometimes complained of feeling uncomfortable because of the mass. We planned to perform combination therapy with corrective orthodontics and surgical bone grafting.

The patient underwent surgery at 9 years of age. The median distance between the incisors was narrower than at 4 years of age after applying corrective orthodontics (Fig. 2B). The surgical operation was carried out under general anesthesia; 1% lidocaine with 1:100,000 epinephrine was injected into the gingiva and mucosa around the alveolar cleft. First, the palatal mass was excised. Then, the mucosa-periosteum flaps on both the lip and palatal side were elevated to expose the maxillary bone in the cleft. A pocket and space surrounding the periosteum was made to transfer iliac cancellous bone (Fig. 2C). The incised mucosa at the median line was sutured completely and mucosal grafting was required on the palatal side (Fig. 2D). There were no complications (e.g., infection or wound dehiscence) on the oral or iliac sides. The macro and microscopic findings of the mass are shown in Fig. 3A and B. The lesion was covered with squamous epithelium. Furthermore, collagenous fiber and an increased number of vessels were observed between the superficial epithelial side and the deep side, including the small salivary gland (Fig. 3B).

Bone formation after grafting was almost complete. Postoperative 3D-computed tomography showed efficient bone formation between the incisors in comparison to preoperative imaging (Fig. 5A and B). The alignment of the teeth remained very good (Fig. 4).

2. Discussion

Median cleft is a rare facial cleft. Tessier described the classifications of facial, craniofacial, and latero-facial clefs. Median upper cleft lip and median alveolar cleft correspond to Types 14 and 0, respectively, in Tessier's classification system [8]. Usually, an alveolar cleft is accompanied by median upper cleft lip. Liao described alveolar bone grafting in the treatment of incomplete median cleft lip. In all 6 of Liao's patients, alveolar cleft was accompanied by a median cleft (e.g., vermilion notch or median cleft lip extending to the mid part of the philtrum) [9]. Freitas reported the surgical correction of 32 Tessier Type 0 cases. Five cases involved isolated median alveolar cleft without median upper cleft lip [10]. However, it is not clear whether they included light symptoms, such as median vermilion notch. Thus, alveolar cleft without a median upper cleft lip is extremely rare. In our case, the patient had no symptoms (e.g., vermilion lip or a wide philtrum).

Median clefts are classified into the true and false types. However, Millard insisted that any congenital vertical cleft through the upper lip, regardless of the extent, should be classified as a median cleft of the lip. Millard further classified two groups [1]: agenesis of the frontonasal process associated with cerebral anomalies and [2] less severe cleft of the median element [5]. The former and latter approximately correspond to the false and true types, respectively. In a review 75 cases, DyMyer described five levels of holoprosencephaly; false median cleft corresponds to Facies 4 [7]. Indeed, Millard's classification of median cleft is easier to understand than the true or false types. This is why DyMyer focused on holoprosencephaly rather than median cleft.

Generally, alveolar cleft requires surgery such as bone grafting in order to align the teeth and induce the eruption of permanent teeth. Of course, dental orthodontics are important both before and after bone grafting. Liao performed cancellous bone grafting for 6 patients with median alveolar cleft. In all cases Abyholm Type 1 bone formation was achieved at the cleft [9,11]. We evaluated the present case using 3D computed tomography. The height of the bone formation in the cleft was efficient and corresponded to Abyholm's Type 1. The timing of bone grafting in cases of median alveolar cleft is controversial. In Liao's report, 2 of 6 patients underwent grafting at 5 years of age, and other 4 patients underwent grafting at 11 years of age [9]. In our case, we performed bone grafting after applying corrective orthodontics. Although time was required to apply the corrective orthodontics, we believe that the timing of surgery was appropriate.

Some authors have described the presence of a mass with median cleft. The mass often exists at the midline cavity, such as in cases of median upper cleft lip and median alveolar cleft [12,13]. It appears that the mass fills the cavity of the cleft. The histological findings

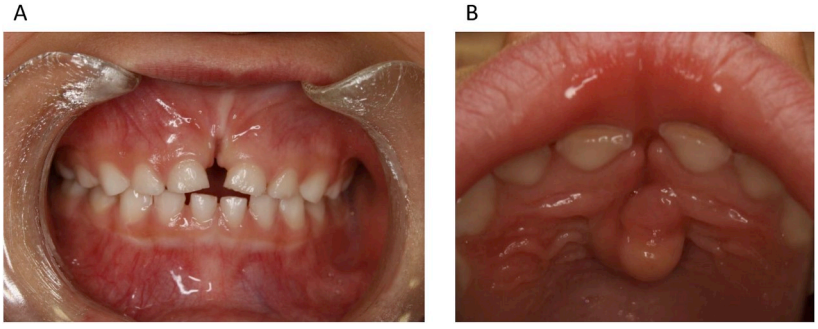

Fig. 1. The median alveolar cleft and palatal mass at four years of age. A gap between the central teeth was recognized (A). The mass at the median hard palate is shown (B).

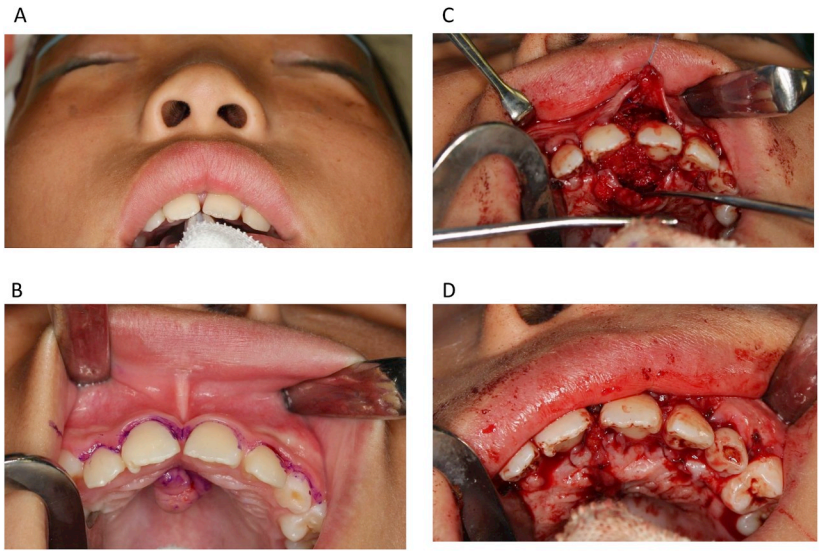

Fig. 2. An intra-operative photograph at 9 years of age. The median cleft was not recognized (A). The gap between the central teeth became narrower after corrective orthodontics. The design for excision of the palatal mass and the incision of the muco-periosteal flaps (B). Bone grafts were transferred from iliac cancellous bone (C). The flaps were put back. The median line was completely closed and the mucocutaneus graft was transferred to the mucocutaneous defect at the right palate (D).

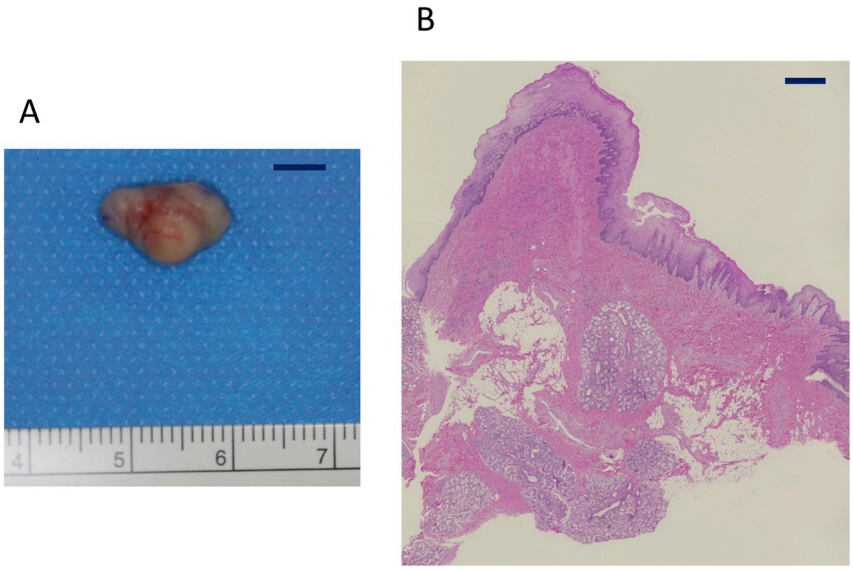

Fig. 3. Pathological examinations. The macroscopic findings. Bar indicates 5 mm (A). The H-E findings. The lesion was covered with squamous epithelium. Collagenous fiber and an increased number of vessels were observed between the superficial epithelial side and the deep side, including the small salivary gland. Bar indicates 500 μm.

were different in the two reports. One was a teratoid polyp and the other was merely a skin mass [12,13]. Our case was similar Nakamura's case in that it appeared like normal tissue. The mass may have existed due to the presence of the bone defect.

3. Conclusion

We reported an extremely rare case of isolated alveolar cleft without median upper cleft lip. A good result was gained by bone grafting and corrective orthodontics. An examination of the filling mass with cleft revealed that it was not a tumor, rather, it was mostly composed of normal tissue.

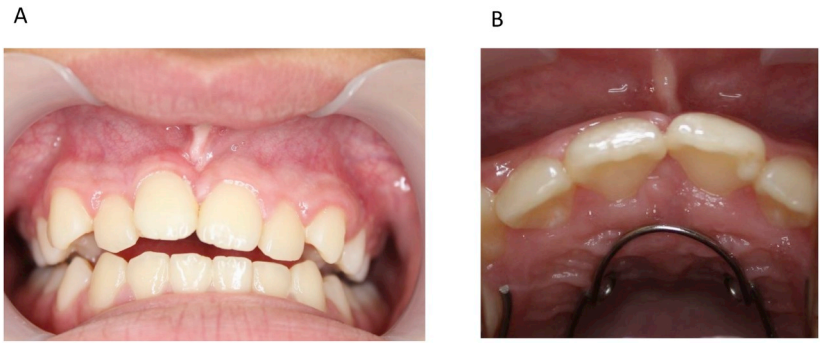

Fig. 4. Postoperative photographs. There was no gap between the central teeth and the tooth alignment was improving (A). The palatal mass was completely removed (B).

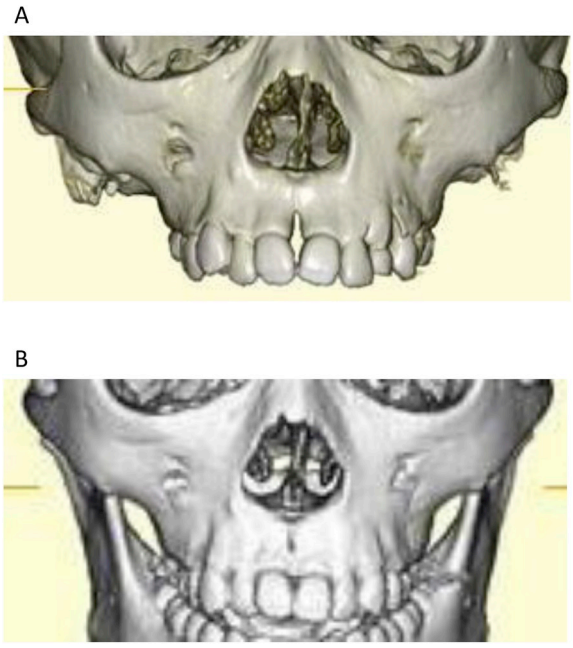

Fig. 5. The 3D computed tomography finding. Pre-operative imaging showed a median alveolar cleft (A). Post-operative imaging at 1 year after surgery. There was no gap between the central teeth. The alveolar cleft was treated completely (B).

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