대한성형외과학회지. 제 38 권, 제 4 호
J Korean Soc Plast Reconstr Surg
Vol. 38, No. 4, 485 - 489, 2011

정중 상구순열의 수술적 교정 치험례

한형민 · 김지예 · 민희준 · 김석원

연세대학교 원주의과대학 성형외과학교실

Surgical Correction of a Median Cleft of the Upper Lip Associated with Enlarged Frenulum and Palatal Masses

Hyung Min Hahn, M.D., Ji Ye Kim, M.D., Hee Joon Min, M.D., Sug Won Kim, M.D., Ph.D.

Department of Plastic and Reconstructive Surgery, Yonsei University Wonju College of Medicine, Wonju, Korea

Purpose: Median cleft of upper lip is defined as any congenital vertical cleft through the midline of the upper lip. It is uncommon, its embryological pathogenesis remains unexplained to date. The authors hereby report a rare case of median cleft of the upper lip associated with enlarged frenulum and palatal mass. This case offers some understanding of the possible embryologic development of this anomaly.

Methods: A 10-month-old boy born by normal vaginal delivery at full-term had a notch in the midline of the upper lip with widened philtrum along with enlarged median frenulum, alveolar cleft, and mass of the hard palate. We performed en bloc resection of the enlarged frenulum and palatal mass and cheiloplasty under general anesthesia.

Results: Histological examination revealed that the frenulum and palatal mass was consisted of fibrous tissue with normal mucous membrane. The postoperative course was satisfactory.

Conclusion: A rare case of median cleft of the upper lip with associated enlarged frenulum and palatal mass was presented with proper surgical management. The surgical technique includes marginal excision of the clefted epithelium and reconstruction of orbicularis oris muscle, in addition to en bloc resection of the palatal mass and frenulotomy.

Key Words: Median cleft of upper lip, Craniofacial cleft, Upper lip, Palatal mass, Enlarged frenulum

Received March 15, 2011
Revised May 2, 2011
Accepted May 23, 2011

Address Correspondence : Sug Won Kim, M.D., Ph.D., Department of Plastic & Reconstructive Surgery, Yonsei University Wonju College of Medicine, Wonju Christian Hospital, 162 Ilsan dong, Wonju 220-701, Korea. Tel: 033) 741-0611/Fax: 033) 732-4022/E-mail: sugwonkim@yonsei.ac.kr

* 본 논문은 2008년 제 65차 대한성형외과학회 학술대회에서 포스터 발표되었음.

I. INTRODUCTION

Craniofacial clefts are rare condition, with the incidence estimated to be 1.4 to 4.9 per 1,000,000 live births.[1-3] In 1974, Tessier established a comprehensive classification of craniofacial clefts based on personal experience with 336 patients. Tessier 0 through 14 cleft locates the median or middle facial cleft, with a varied spectrum of presentation from a simple vermilion notch to a wide cleft with a bified nose and hypertelorism.[1,2] There have been many reports of median cleft of the upper lip with diverse presentation and their surgical management.

Here, we describe a rare case of median cleft lip associated with enlarged frenulum and solid mass of the hard palate with normotelorism and meningoencephalocele. This case does not classified into DeMyer groups, since hypertelorism was not observed in this patient. There has been one prior report of a similar case in 2002 by Asada et al., in which the case with enlarged frenulum and palatal mass was presented.[4] The authors also describe a literature review of previous reports of clinical cases and their possible pathogenesis.

II. CASE

A 10-month-old Korean male presented with midline deformity on his upper lip. The patient was born by normal vaginal delivery at full-term with no complication. The mother did not have any disease during pregnancy. She did not smoke or drink alcohol, nor was she exposed to any environmental risk factors during pregnancy. The family history was noncontributory. The infant's general growth and development didn't show age-appropriate, with the weight below 10 percentile in his age and delayed gross motor development. General physical examination revealed no anomalies of the trunk, extremities, gastrointestinal, cardiovascular, or genitourinary system. On physical examination of the face, there was a median cleft of the upper lip matching to Tessier 0/14. The notching groove started from columellar base,

extending to upper vermilion, dividing the widely separated philtrum in the midline (Fig. 1). The vermilion was separated in the midline by the groove. The patient had a widened and shortened columella. The orbicularis oris muscle fibers were considered to be interrupted completely. The median cleft was accompanied by several other findings. Intraorally behind the vermilion, enlarged frenulum was observed, the diameter was about 2 mm. The frenulum was extended through mucosa above alveolar bone, hard palate, and soft palate. The end of the frenulum was associated with a solid mass which was found in the center of the hard palate. The mass was about 5 × 5 mm and covered with normal mucous membrane. The partial cleft of maxillary alveolus was observed on preoperative computed tomography, but there was no cleft palate nor orbital hypertelorism (Fig. 2).

In addition to midline facial deformity, the patient had maldevelopment of the forebrain. On computed tomography and magnetic resonance imaging of the head, large defect in midline anterior base of the skull was observed. Through the defect, herniation of the meninges occurred forming meningoencephalocele in sphenoethmoidal area (Fig. 2).

The intercanthal distance as well as the shape of the nose was normal. The length and height of the vermilion was in normal limit. The function of the soft palate was normal.

Treatment plan included direct excision of the clefted skin and vermilion with primary repair, reconstruction of the orbicularis oris muscle in the midline, narrowing and lengthening the columella base in V-Y fashion, and the en bloc removal of the palatal mass along with abnormally connected frenulum.

Surgical correction of the cleft was performed at 10 months of age. Oral intubation was used to administer general anesthesia. The incisions were outlined, and white roll, wet-dry border of the vermilion were marked with sterile gentian violet. After infiltration of 1 mL of 2% lidocaine with 1 : 100,000 epinephrine, the grooved skin was excised starting from columellar base in V-shape to labial skin, 5 mm in width and 13 mm in length. The incision extended through vermilion and oral mucosa into abnormally enlarged frenulum. The en bloc resection of enlarged frenulum and palatal mass was performed. The skin and subcutaneous tissue were dissected from the orbicularis oris muscle on both side. Reconstruction

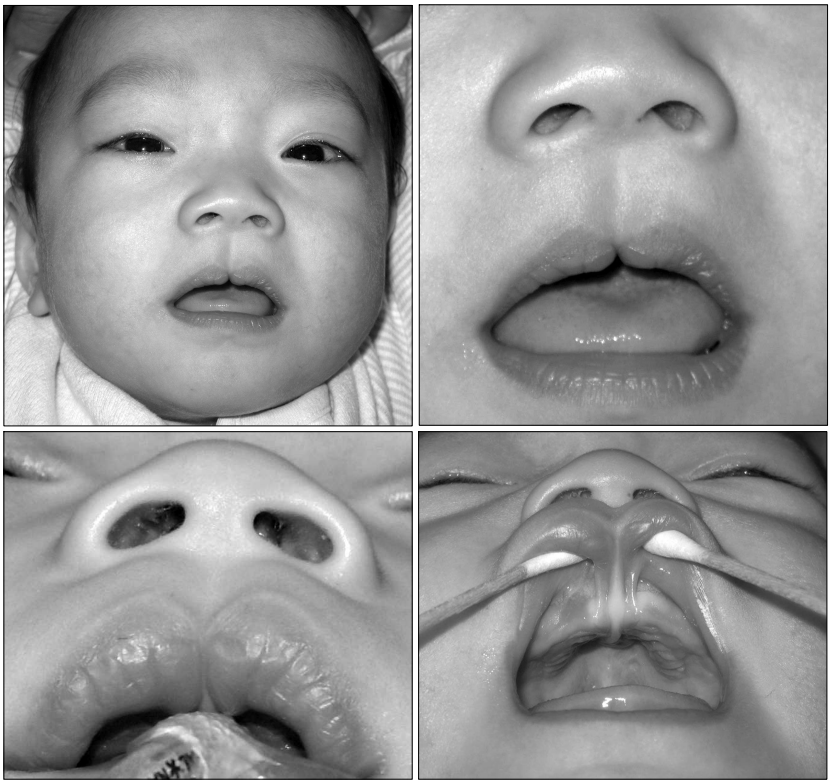

Fig. 1. (Above, left, Above, right, and Below, left) A 10-month-old male with median cleft of the upper lip. Note the central indentation of vermillion, widened philtral column, widened and shortened columella. (Below, right) The intraoral view. Median notch was associated with enlarged frenulum and the mass of the hard palate.

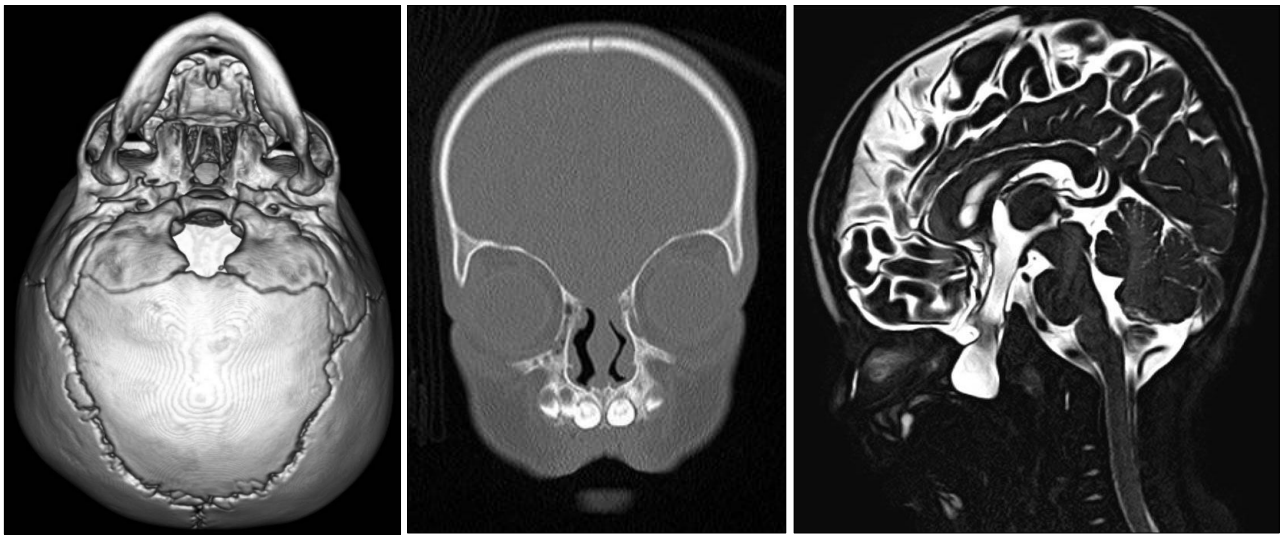

Fig. 2. (Left) Inferior view of three-dimensional computed tomography. Note the midline alveolar cleft. (Center, right) Computed tomography and magnetic resonance imaging. The large basal bony defect in sella area with sphenoethmoidal meningoencephalocele was observed.

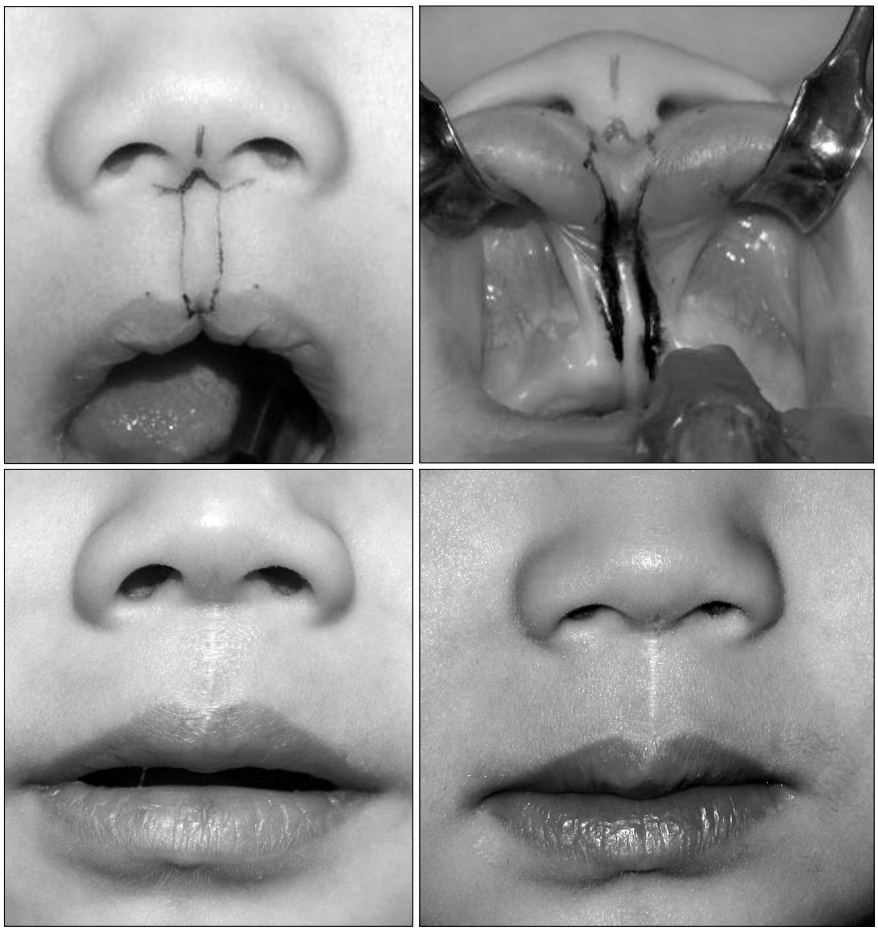

Fig. 3. (Above, left, Above, right) Operative design. Direct excision of clefted skin with en bloc resection of enlarged frenulum and palatal mass. (Below, left, Below, right) Postoperative view at 6 months and 2 years 9 months. Note narrowing of columella and philtral width with acceptable scar. Natural continuity of white roll is achieved.

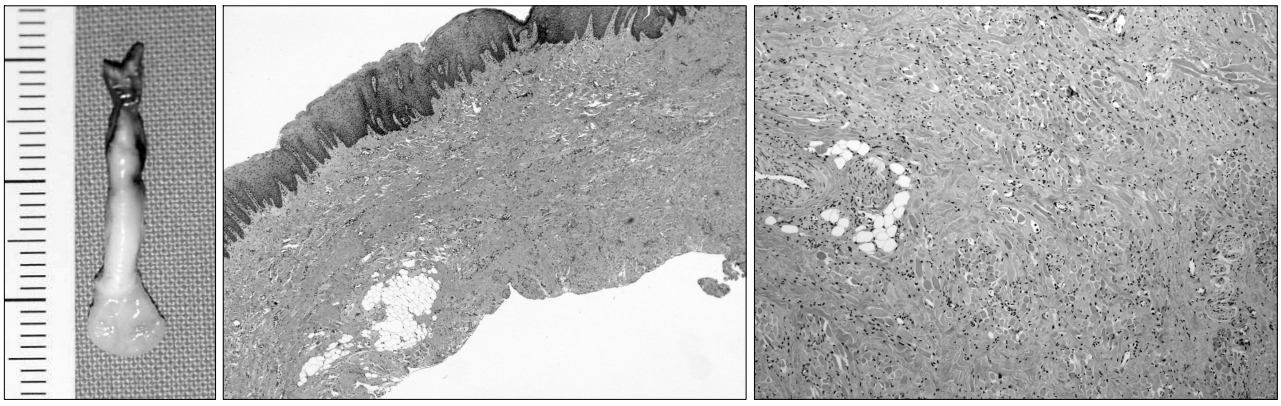

Fig. 4. (Left) Resected enlarged frenulum and palatal mass. (Center) Histologic findings. The mass was diagnosed with mucocele (Hematoxylin and eosin stain, × 12.5). (Right) The resected frenulum was composed of normal skeletal muscle and fibrous tissue (Hematoxylin and eosin stain, × 100).

of the muscle with Z-plasty was performed using 5-0 Vicryl suture. The skin was repaired with 6-0 Vicryl and 6-0 Nylon suture carefully realigning the white roll and red line of the vermilion. The V to Y advancement of columellar base was performed using 5-0 Vicryl and 6-0 Nylon. The median mass in hard palate was resected completely and bilateral mucosa-muscle flap advanced to repair the defect. The soft palate and frenulum were reconstructed with Z-plasty using 6-0 Vicryl. The nonabsorbable sutures were removed on the fourth postoperative day.

Postoperatively, the patient had a satisfactory result with acceptable scar (Fig. 3). Cupid's bow was well aligned and the height of the both sides was equal. The natural philtral dimple and tubercle of the lip were formed instead of notched skin in the midline. The width of columellar base was reduced from 8.5 mm to 6 mm. A good cosmetic result was observed at 5 months postoperatively.

Histological examination revealed that the frenulum consisted of fibrous tissue with normal skin and mucous membrane and the palatal mass was fibrous tissue with mucous cyst (Fig. 4).

III. DISCUSSION

The median cleft of the upper lip is a rare condition among craniofacial anomalies. There have been some reports in the literature depicting different variations of presentation.

The embryology of facial development is still not completely understood. And this is why there exists many explanations and classifications of middle facial clefts. Traditionally, the mechanism of craniofacial cleft formation can be described by either the fusion failure theory or the failure of mesoderm penetration theory. For interpreting the median cleft of the upper lip, Dursy and His fusion of processes theory doesn't help. Rather, Veau's proposal that the lack of neuroectoderm and mesoderm migration and penetration into the bilaminar ectodermal sheets is the cause of clefts can give some basis for explanation of median cleft.[5] This occur during the third week of fetal development when bilateral frontonasal processes become thickened. Besides, there is another explanation that the enlarged frenulum and palatal mass are embryological remnant of the tectolabial frenulum and palatine papilla. During the third month of gestation, persistence of the tectolabial frenulum or continued connection of the palatine papilla with the lip after birth results in the failure of the median upper lip to develop normally.[4]

There have been many debates on how to classify the median cleft according to the various degrees of presentation. In contrast to Braithwaite et al.[6] who classified Median clefts of the upper lip into true and false group, Millard and Williams did not differentiate between true and false median cleft and proposed that any congenital, vertical cleft through the center of the upper lip, no matter what extent, be classified as a median cleft of the lip.[5] They divided median clefts into two forms: agenesis of the median element, which is frontonasal process, and clefts of the median element. Wiemer et al.[7] indicated that median clefts can be divided into those with hypertelorism and those with hypotelorism, thus midfacial malformation can range from frontonasal dysplasia to holoprosencephaly.

The case we presented is classified as cleft of the medial element according to Millard and Williams.[5] This

type of deformity is attributed to anomalous development of the lateral nasal and frontal processes, not merely a failure of fusion of mesial nasal processes. The patient had normotelorism, so the patient is not diagnosed with median cleft face syndrome that DeMyer described in 1967.[8]

The surgical technique we have chosen to correct the median cleft lip is direct excision of the clefted epithelium in the midline of the upper lip. For lengthening of the skin in the center of the Cupid's bow, Millard's combination of an inverted V excision and a 90-degree angle in the excision 2 mm above the mucocutaneous white roll on each side of the cleft can be helpful.[9] This case did not required the lengthening of the skin.

A rare case of median cleft of the upper lip with associated enlarged frenulum and palatal mass was presented with proper surgical management, along with detailed patient review and long-term follow up photographs. The surgical technique includes marginal excision of the clefted epithelium and reconstruction of orbicularis oris muscle, in addition to en bloc resection of the palatal mass and frenulotomy.

REFERENCES

1. Tessier P: Anatomical classification of facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg* 4: 69, 1976
2. Kawamoto HK Jr: The Kaleidoscopic world of rare craniofacial clefts: order out of chaos (Tessier classification). *Clin Plast Surg* 3: 529, 1976
3. Kawamoto HK Jr: Rare craniofacial clefts. In McCarthy JG (eds): *Plastic Surgery*. Philadelphia, WB Saunders, 1990, p 2922
4. Asada K, Ishibashi K, Usui H, Hirano Y: A rare case of median cleft of the upper lip associated with enlarged frenulum and palatal masses. *Plast Reconstr Surg* 109: 1464, 2002
5. Millard DR Jr, Williams S: Median lip clefts of the upper lip. *Plast Reconstr Surg* 42: 4, 1968
6. Braithwaite F, Watson J: A report on three unusual cleft lips. *Br J Plast Surg* 2: 38, 1949
7. Wiemer DR, Hardy SB, Spira M: Anatomical findings in median cleft of upper lip. *Plast Reconstr Surg* 62: 866, 1978
8. DeMyer W: The median cleft face syndrome. Differential diagnosis of cranium bifidum occultum, hypertelorism, and median cleft nose, lip, and palate. *Neurology* 17: 961, 1967
9. Millard DR: Median cleft lip with hypertelotism. In Millard DR: *Cleft Craft*. Boston, Little Brown & Co., 1976, p 745