

Isolated Congenital Alveolar Synechiae: Review of Literature and Case Report

— A Case Report —

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Abstract

선천성 치조점막 유착에 대한 문헌고찰 및 증례보고 —증례보고—

강릉대학교 치과대학 구강악안면외과학교실, [†]구강병리학교실, *GSR 인도 하이데라바드 GSR 센터,
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상악과 하악이 선천적으로 붙어서 태어나는 선천성 유합증은 드문 선천성 기형으로 단순히 점막이 붙은 점막유합증에서 악골이 붙은 골유합증까지 다양하게 나타난다. 이중 상악골과 하악골의 골자체가 붙는 골유합증은 아주 드물어서 현재까지 26증례만 보고되고 있는데, 보고된 대부분의 증례는 편측에만 발생하는 불완전형으로 알려져 있다.

7세 된 여아환자가 인디아의 GSR 병원에 입이 벌어지지 않는다는 주소로 내원하였는데 환아의 턱은 완전히 움직이지 않았으며, 2-3 mm 정도 벌어지는 앞니부위에서는 2.5 cm 폭경의 3.0 mm 두께의 단단한 치조점막이 관찰되었다. 전기메스로 전방부의 부착성 섬유밴드를 잘라준 후 즉각적인 개구정도는 16 mm 정도까지 가능하여 구강으로의 기관삽관이 가능하였다. 삽관후 양쪽 후방부 협측점막의 두꺼운 밴드들을 모두 제거하여 개구량을 33 mm까지 증진시킨 후 수술을 종결하였다. 환아의 보호자에게 거즈 블록과 설압자를 이용하여 개구 연습을 능동적으로 시키도록 강조하여 교육하였으며 술후 16개월 경과시까지 특별한 합병증이나 개구량 감소는 관찰되지 않았다.

독립적으로 발생한 선천성 치조점막 유합증 환자에서 비정상적으로 커져있는 과두와 설골이 관찰되었는데, 설-하악 구조의 비정상적인 발육에 기인하여 지속적인 비정상적 운동으로 인한 이차적인 치은과 협점막의 섬유성 부착이 생긴 것으로 추측되었다. 이에 마취과와의 효율적인 협진으로 기관절개술 등의 부가적인 마취방법 없이 효과적으로 치료할 수 있었다. (JKDSA 2007; 7: 22~26)

핵심용어: 선천성 유합(Congenital fusion), 악골유합(Syngnathia), 악골점막 유합(Synechiae), 구강 기관 삽관 (Orotracheal intubation)

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* This work was co-supported by Korea Research Foundation Grant (KRF-2005-013-E00036) and a grant of the Korea Health 21 R&D Project, Ministry of Health & Welfare, Republic of Korea (A050547).

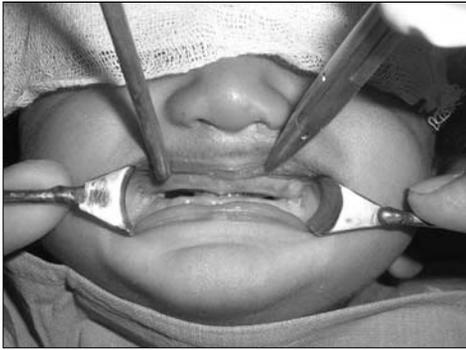


Fig. 1. Thick fibrotic bands in anterior alveolar lesion.



Fig. 2. Coronal CT scanning showing enlarged both condyle head (arrows) of mandible but no bony ankylosis bilaterally.



Fig. 3. Removing fibrotic bands state.



Fig. 4. Performing successful orotracheal intubation.

Congenital fusion of the maxillary alveolar and mandible is a very rare disease. Most of cases are occurred either as a single mucosal band called a synechiae or as a complete bony fusion called a synostosis (Choi et al, 2004; Daniels, 2004; Tanrikulu et al, 2005; Ugurlu et al, 2005; Verdi et al, 1984).

We present a case of isolated congenital alveolar synechiae in a 7-month-old girl, and details of the operative and anesthetic management are presented with literature review.

CASE REPORT

A 7-month-old girl suffering from restricted mouth opening was brought by her family to the GSR facial plastic surgery clinic with a chief complaint of limited mouth opening. Physical examination showed that her

general condition was good without other abnormalities. The baby had been born vaginally and no other anomaly than alveolar synechiae found during or after her birth. She was the family's first child and there was no history of cleft palate or other anomaly in close relatives. Her mother had undergone a healthy pregnancy and there was no account of any illness, trauma or drug use, and so on. Otherwise, no useful information was obtained from her family's medical history.

Patient's mandible was completely immobile and her mouth opening was only 2–3 mm because of anterior intermaxillary fibrous adhesions about 2.5 cm wide and 3 mm thick in the anterior alveolar lesions (Fig. 1). She could eat through a small gap in the posterior part of the mouth, and no other examination of the oral cavity, palate, and tongue could be achieved.



Fig. 5. Excised state of posterior fibrotic bands on posterior buccal mucosa.

On the serial axial and coronal CT sections of maxilla, both mandibular condyles were shown as the hypertrophic morphology with no evidence of bony ankylosis in both sides. However, any bony erosion or destruction was not detected, excluding that fluid density was increased in middle ears bilaterally due to prominent adenoid (Fig. 2). In addition, her hyoid bone was prominently located in the sublingual area on CT image.

After consultation with the pediatrician, we planned this operation with careful consideration to get over the intubation problem. Deep conscious sedation with intravenous ketamin in the dosage of 1.0 mg/kg of body weight was done, and the injection of a low dosage of lidocaine to the anterior adhesion regions. After cutting the adhesive fibrotic bands by bovie (Fig. 3), active mouth opening increased immediately up to 16 mm. Skillful anesthesiologist performed orotracheal intubation successfully (Fig. 4), and after intubation procedure, thick fibrotic bands on both posterior buccal mucosa were all excised (Fig. 5). At the end of these procedures, more than 33 mm mouth opening was achieved immediately (Fig. 6).

After the mouth was opened, an intraoral examina-



Fig. 6. Confirming more than 33 mm mouth-opening.

tion revealed that palatal vault and uvula were found in normal shape. And the mobility of the tongue looked fine. Parents were educated firmly to continue her mouth opening exercises by using a gauze block and wood blades. No post-operative complications and recurrence were observed during 16 months follow-up period.

DISCUSSION OR COMMENTS

Congenital fusion of maxillary and mandibular alveolar margins is extremely rare. There are approximately 26 cases of syngnathia reported in the literature up to date. In the early stages, this disease can be easily treated, but in late stages, it frequently involved with temporomandibular joint ankylosis. After first reporting of a congenital bony temporomandibular ankylosis by Burket et al. in 1936, very few cases of congenital fusion of maxilla and mandible have been reported (Daniels, 2004; Tanrikulu et al, 2005; Verdi et al, 1984).

Isolated congenital alveolar synechiae is a very rare disease, most of the reported cases of maxillomandibular fusion were alveolar fusion in the lateral side,

but the case of isolated fibrous interalveolar synechiae was also very rare. Our case is not associated with other anomalies, although many reported cases were associated with other facial anomalies such as cleft palate, mandibular hypoplasia or mandibular cleft, bifid or absent tongue, and a persistent buccopharyngeal membrane. Congenital alveolar synechiae with various syndromes such as Van der Woude (VDW) and cleft palate lateral alveolar synechiae syndrome (CPLASS) is generally observed together, and is concomitant with other anomalies in the maxillofacial or other regions of the body (Gassner et al, 1979).

Various experimental studies have been performed to study the embryologic basis for congenital alveolar synechiae, but the cause of this fusion is unknown yet. Gartlan et al. speculated that major congenital synechiae of the oral cavity constitute a clinically confusing spectrum of abnormalities, and they proposed two categories on the basis of clinical data such as abnormalities secondary to persistence of the buccopharyngeal membrane and abnormalities secondary to the formation of ectopic membranes. An ectopic membrane results from abnormal fusion and can be subclassified as a subglossopalatal membrane, glossopalatal ankylosis or syngnathia. In our case, the cause of the synechiae seems to be an abnormal secondary to the formation of ectopic membranes (Gartlan et al, 1993).

Embryologically, the alveolar ridges, tongue and palatal shelves are in contact with each other during the 7th to 8th week of development, and the ensuing palatal closure depends on downward contraction of the tongue. When the tongue protrudes from the mouth as a result of downward mandibular movements, this can prevent the alveolar ridges from fusing. During these critical development period, any genetic, teratogenic, or mechanical insults may lead to close, quiescent contact between oral structures with consequent abnormal fusion. Persistence of the buccopharyngeal membrane, amniotic constriction bands in the region of the developing branchial arches, abnormality of the stapedial artery, environmental insults

such as trauma in pregnancy, teratogenic drugs such as meclozine, and large doses of vitamin A have been also known as the postulated causes.

We did not find any family history or prenatal and postnatal problems of this patient, so any unknown cause of genetic or developmental origin might be not happened to the patient. In our case, the fibrous adhesions in anterior alveolus and posterior buccal gingival made initial 2–3 mm mouth opening, so it can be opinioned that these situations might prevent the formation of a cleft palate and TMJ ankylosis during the phases of embryonic development.

The general standardization of congenital maxillo-mandibular interalveolar fusion treatment is not established because of the rare occurrence of this disease. Successful surgical corrections during late post natal life also have been reported. But, early surgical approach is usually recommended because of the high risk of aspiration pneumonia and feeding handicaps during patient's childhood, and of (not) disrupting of teeth eruption and alignment. If the patient are not treated during his early childhood, growth retardation or psychological diseases will be happened because of malnutrition and difficulty in communication.

In our case, anterior mucosal adhesions were separated easily with excision after the administration of deep sedation and local anaesthesia. This simple division of the anterior alveolar synechiae was necessary to allow mouth opening for orotracheal intubation. After performing the safe intubation, thick fibrotic bands on both posterior buccal mucosa can be all excised. These all procedures must be considered as a very important and necessary sequences for ensuring airway during initial mouth opening procedures. More than 33 mm mouth opening was confirmed immediately, and active mouth opening exercises were educated to the patient's parents firmly and continuously during postoperative 6 months. We recommended a gauze bite block and wood blades instead of a silicon block, since the patient was too small to burden the rigid silicon block.

In the present case, the cause of the synechiae

seems to be an abnormality secondary to the formation of ectopic membranes. The adhesions were separated with excision after the administration of local anaesthesia. Simple surgical division of the adhesions was necessary to allow normal feeding, to avoid upper airway obstruction and to allow normal mandibular function and growth.

Recurrence of maxillomandibular fusion between the alveolar arches after operation have also been reported in several literature, but most of their cases were synostosis cases without active mouth opening exercises. No post operative complications and recurrence were observed during 16 months follow up period in our case.

However, contrast to other cases reported in the literature, the present case characteristically disclosed enlarged both condyles with hypertrophic cartilaginous cap in CT observation. Although there appeared no bony fusion in the condyle joint, those enlarged condyles might be a cause of mandibular dysfunction.

Furthermore, the rest position of hyoid bone for the present patient who complained of mandibular immobilization was deeply approximated to the sublingual space, and the body of hyoid bone was thickly ossified in the CT findings. Taken together, the malpositioned hyoid bone may cause weak mouth-opening activity, and the hypertrophic bilateral mandibular condyles may affect her mandibular movement more difficult. Because there was no evidence of familiar history and other systemic medical abnormalities, we thought this isolated congenital alveolar synechia was arisen from some insults of developmental or environmental origin, which aggravated the harmonious development of neuro-muscular skeletal complex of linguo-mandibular structure.

In the point of anesthesiologic view, tracheostomy

can be needed especially in the congenital synostosis patient for performing an osteotomy to relieve the fusion between maxilla and mandible. But only in the congenital synechia case, tracheostomy can be avoided by doing deep sedation first and consequently doing orotracheal intubation after detaching anterior mucosal adhesions such as in this case. By doing these serial approaches, lots of complications arisen from tracheostomy procedure in young child can be prevented.

So we reported an extremely rare case of alveolar synechia without other congenital abnormalities and recommended the suitable treatment procedures including anesthetic techniques.

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