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SURGICAL CORRECTION OF HEMIFACIAL MICROSOMIA REPORT OF A CASE

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Abstract

Hemifacial microsomia is characterized by underdevelopment of the TMJ, mandibular ramus, and associated muscles of mastication.

The Maxilla and malar bones on the affected side frequently are underdeveloped. The contiguous parotid gland may be hypoplastic.

Preauricular sinus tracts and tags may exist, along with underdevelopment of the associated external ear, and affected facial nerve and muscles of facial expression may also show dysfunction.

Children exhibiting the more classic signs will be identified at birth. Little is known about the etiology of hemifacial microsomia.

We have corrected surgically a 22-year-old woman with hemifacial microsomia. We have performed leveling Le Fort I osteotomy with iliac bone graft on the maxilla, reverse-L osteotomy and iliac bone graft on the right mandibular ramus, vertical ramus osteotomy on the left side, onlay bone graft on the right mandibular body, and augmentation genioplasty.

The postoperative course was uneventful and restoration of facial asymmetry was achieved.

INTRODUCTION

Hemifacial microsomia is a variable, progressive, asymmetric defect involving the skeletal, soft tissue, and neuromuscular components of the first and second branchial arches.

With an incidence of 1 in 5,600 live births in the USA, it is the second most common congenital facial deformity after cleft lip and palate[1,2].

Other identifiable craniofacial syndromes have anomalous characteristics similar to hemifacial microsomia. Pierre Robin syndrome (glossoptosis-micrognathia), Treacher Collins syndrome (mandibulofacial dysostosis), Goldenhar's syndrome (oculoauriculovertebral dysplasia), and Hallermann-Streiff syndrome (dyscephalia mandibulo-oculo facialis) all exhibit deformities of the first arch[3,4].

Hemifacial microsomia is characterized by underdevelopment of the TMJ, mandibular ramus, and associated muscles of mastication.

The maxilla and malar bones on the affected side frequently are underdeveloped.

Asymmetric mandibular growth is the earliest skeletal manifestation of hemifacial microsomia and plays important role in progressive deformity of the ipsilateral and contralateral facial skeleton[5].

The deficiency in soft tissue bulk, hypoplasia of the first and second branchial arch muscles, and facial nerve palsy also play a role in the progressive skeletal distortion.[1,5].

The parotid gland may be hypoplastic.

Preauricular sinus tracts and tags may exist, along with underdevelopment of the associated external ear.

Children exhibiting the more classic signs will be identified at birth and the deformities are more often unilateral[5].

Initially the most disturbing esthetic features of the child with hemifacial microsomia are the deformities of the external ear[6]. The auricle may be missing or grossly mishappen. Accessary tags and sinus tracts may be present along the orotragal line.

The full-face view best demonstrates the asymmetry, with the chin deviated toward the hypoplastic side. Underdevelopment of both the skeletal and soft tissue components accentuates the aymmetry. The mouth may be enlarged and skewed toward the affected side (macrostomia)[6].

The profile view emphasizes the mandibular retrusion. The retrusion is primarily the result of the unilateral hypoplasla, but the growth pattern of the opposite, relatively normal hemimandible may contribute to the appearance of retrusion.

The dental occlusion is altered as a result of the asymmetric growth of the jaws. Insufficient length of the dental arch causes crowding and impaction of teeth, particularly in the mandible. Deficient alveolar bone height in both the mandible and maxilla is evident on the affected side, but the development of the individual teeth in shape and number is unaffected.

The relative retrusion of the mandible and its asymmetric development contribute significantly to the malocclusion.

In attempting corrective measures, it is essential to regard the malocclusion as secondary to the more fundamental skeletal deformity.

Early surgical correction of the facial asymmetry has a number of potential benefits in addition to the positive emotional effect. Longacre[7] stressed early and repeated augmentation of the deficient skeletal areas, using split ribs. The subperiosteal regeneration of ribs in the growing child provides a continual source of bone.

Hovell[8] and Osborne[9] have advocated early and, if necessary, repeated surgery to lengthen the hypoplastic mandible during the time of facial growth. In treatment planning, both advocated creating an open bite on the hypoplastic side in order to establish space for alveolar growth and tooth eruption. Whereas Osborne recommends surgery by age 6 years, Hovell suggests delay until 8 years of age.

Converse et al[10] advocate early surgery and suggest the period of mixed dentition (age 8 or 9 years) as the most practical time for the initial surgery. There are usually sufficient permanent teeth by age 9 to provide an aid to intermaxillary fixation. A second surgical stage in late adolescent should be anticipated to correct any disparity of growth following the first operation and to attain adequate facial contour.

Although conventional orthodontic tooth movement is of little value in young patients with hemifacial microsomia, efforts to guide skeletal growth and stimulate the affected areas are indicated. Harvold[11] advocates the use of activators to guide eruption of teeth and prevent midline shift until the time of operation. This approach has a stimulatory effect on muscle development and serves to prevent canting of the occlusal plane.

For older patients, the goal of presurgical orthodontics should be to align the teeth as ideally as possible to their own arch in anticipation of surgical realignment of the skeletal parts. This approach usually makes the occlusion worse temporarily. At the time of surgery, it is often advisable to create an open bite posteriorly on the affected side, into which the teeth are extruded orthodontically, afterward.

In those of moderate-to-severe hemifacial microsomia in which treatment has been delayed until after completion of facial growth, correction of both the maxilla and mandible.

In Korea, there are few case report[12] on hemifacial microsomia, so we present another case and discuss

the characteristics and corrective measures for this deformity.

REPORT OF A CASE

A 22-year-old woman visited the Dept. of Oral & Maxillofacial Surgery, Pusan National University Hospital for correction of facial asymmetry on May 2, 1988. Her mother said that the birth had been abnormal but there was no unusual event during the gestation period. At birth, hypoplasia of the right mandibular area, maxilla, zygoma, and external ear was noticed and went on worse. The past history revealed that the condition was congenital, with no history of maternal or intrauterine problems.

Generalized undergrowth and underdevelopment was noticed (142cm in height, 40kg in weight).

Generalized dental caries due to poor oral hygiene made several teeth extracted at the local dental clinic.

The face had an obvious asymmetry characterized by a flatness of the right side and deviation of her mandible to the right (Fig. 1, 2). Occlusal analysis revealed a tilted occlusal plane that was higher on the right than the left, a discrepancy between the dental and facial midlines, and multiple dental caries and right posterior cross bite (Fig. 3).

There was normal range of mandibular motion, without any dysfunction of the temporomandibular joints, even though the right condyle was malformed. The panoramic radiograph (Fig. 4) showed a diminutive right ramus and temporomandibular joint. The posteroanterior cephalogram (Fig. 5) showed underdevelopment of the right zygoma, a tilted maxillary occlusal plane, and discrepancy between the maxillary dental midlines and that of the face. The lateral cephalogram (Fig. 6) revealed mandibular retrusion.

This involved a three-dimensional movement of total maxilla by Le Fort I osteotomy, with downward displacement by 6mm to level the occlusal plane and bony defect was filled a part of corticocancellous bone from iliac crest (6cm in length, 3cm in width) and commercially-packed xenogeneic bone (Osteo-

Fig. 1. 22-year-old woman demonstrates facial asymmetry and and shift of the chin to the right side.

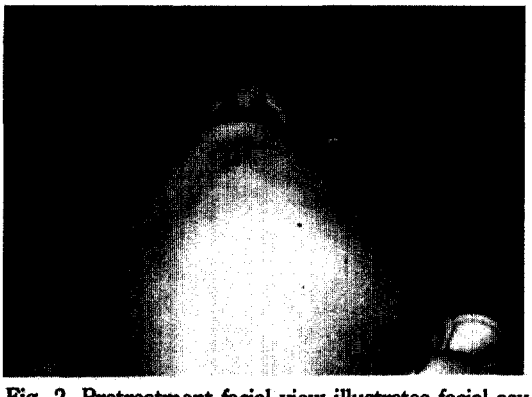

Fig. 2. Pretreatment facial view illustrates facial asymmetry with a flatness of the right side.

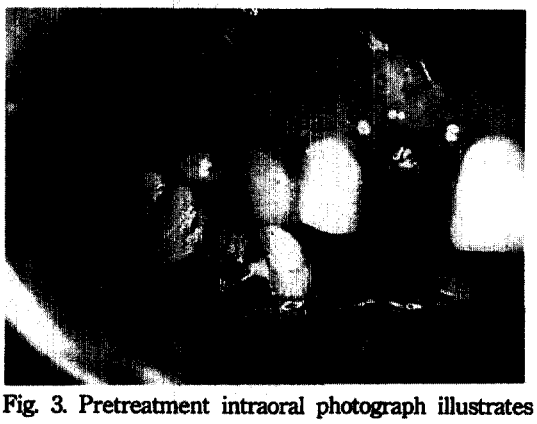

Fig. 3. Pretreatment intraoral photograph illustrates posterior cross bite of the right side.

vit®). The mandible was then corrected with ramal osteotomies (left EVRO and right reverse L osteotomy with iliac bone graft) to allow it to occlude with the repositioned maxilla, and for downward displacement and lengthening of right ramus. To fill out the deficient right mandibular body region, onlay grafting with iliac bone was added. Augmentation genioplasty was performed for anterior displacement by 6mm and filling the bony defect with iliac bone and Osteovit®.

Fig. 4. Panoramic radiograph illustrates a diminutive right ramus and temporomandibular joint.

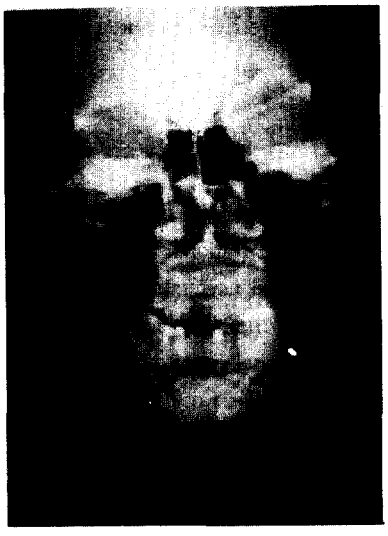

Fig. 5. Posteroanterior cephalogram illustrates underdevelopment of the right zygoma, a tilted occlusal plane and mandibular symphysis deviated to the affected side.

Fig. 6. Lateral cephalogram illustrates mandibular retrusion and short ramus.

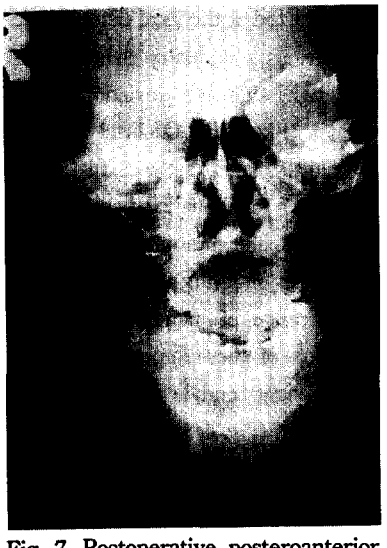

Fig. 7. Postoperative posteroanterior cephalogram.

The postoperative radiographs show the surgical result (Fig. 7, 8, 9). Postoperatively, facial appearance was greatly improved (Fig. 10) and the occlusal plane became horizontal (Fig. 11).

She was discharged on June 1, 1988, and was taken on prosthodontic treatment thereafter.

Fig. 8. Postoperative lateral cephalogram.

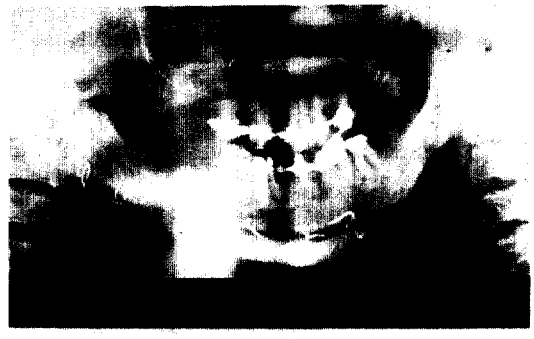

Fig. 9. Postoperative panoramic view.

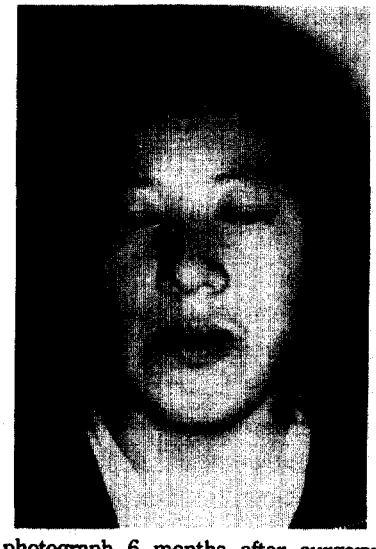

Fig. 10. Facial photograph 6 months after surgery illustrates the soft tissue changes as a result of a Le Fort I maxillary osteotomy with iliac bone graft, bilateral ramus osteotomies, and a genioplasty.

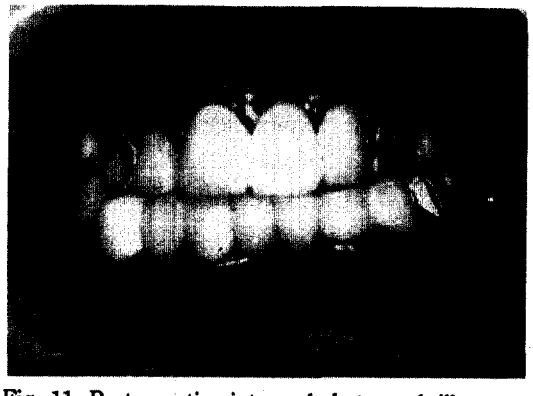

Fig. 11. Postoperative intraoral photograph illustrates horitontal occlusal plane. (after prosthodontic treatment)

DISCUSSION

Little is known about the etiology of hemifacial microsomia. Inferences drawn from descriptive analyses of the clinical examples, fetal material, and a small number of animal studies from the basis for the present theories. Stark and Saunders[13] supported the theory of mesodermal deficiency based on the presence of generalized hypoplasia of the soft and hard tissue. McKenzie[14], convinced that inadequate blood supply was the key factor, attributed the reduced regional development to a malformation of the exter-

nal carotid artery system. Pursuring the vascular theory further, Poswillo[15] suggested vessel wall rupture with hematoma formation as the causative agent. He has described an animal phenocopy in which hemorrhage from the developing stapedial artery produces a hematoma in the area of the first and second branchial arches. Between the third and fifth weeks in utero, the first aortic arch disappears, and the external carotid systems develops. This vascular rearrangement from branchial arch vessels to the carotid systems forms the weakest link in the developmental chain, since it takes place at a time when an anomalous development is most likely to occur[16]. The size of the hematoma and resultant tissue destruction explain the morphology and variability of hemifacial microsomia[17].

The normal mandible grows downward and forward in relation to the cranial base by programmed bone deposition and resorption on periosteal and endosteal surfaces. Increase in vertical height of the ramus is the result of bone deposition on the posteroinferior surface and resorption on the anterior surface. Resorption along the anterior border of the ramus also contributes to the length of the mandibular body. Resorption on the medial surface and deposition on the lateral surface account for the shape and width of the mandible in the transverse plane[17]. In hemifacial microsomia, three-dimensional mandibular growth on the affected side is impaired and the mandible becomes short, retrusive, and narrow[17].

The maxilla normally grows inferiorly (downward) and anteriorly (forward) as a result of bone resorption on the superior (nasal) and anterior surfaces and deposition of bone on the inferior (palatal) surface. The nasomaxillary region therefore downward and forward, away from the cranial base. In the case of hemifacial microsomia, mandibular hypoplasia inhibits normal downward (vertical) growth of the maxilla and midface[17,18]. It prevents the progressive separation of the orbit from the piriform aperture and maxillary alveolus. The result is a short maxilla with an occlusal plane that is tilted upward toward the abnormal side ; the orbit may be inferiorly displaced[1,2,5].

The skeletal defect of hemifacial microsomia is classified by the anatomy of the mandibular ramus and temporomandibular joint[1,19,20]. Type I skeletal deformity consists of a 'mini-mandible' and temporomandibular joint ; all structures are present, normal in shape, but small. Type II skeletal deformity consists of a small and abnormaly shaped mandibular ramus with a hypoplastic, anteriorly and medially displaced temporomandibular joint. Type III hemifacial microsomia is characterized by complete absence of the mandibular ramus and temporomandibular joint.

The soft tissue defect consists of a decrease in bulk of subcutaneous tissue ranging from mild to severe ; the muscles of mastication and of facial expression are hypoplastic. The ear anatomy varies from normal to complete absence of the external ear. There is a conductive hearing loss due to hypoplasia of the ear ossicles, which are first and second branchial arch derivatives. Some patients have cranial nerve abnomalities, usually consiting of facial nerve palsy and/or deviation of the palate toward the affected side with motion. Rarely, a patient may have a sensory deficit in the fifth cranial nerve distribution[21,22].

Controversy exists concerning the appropriate timing of reconstructive procedures involving bone in relation to the growth pattern. The reasons for delay are : (1) possible untoward effects of the surgery on subsequent growth and (2) the difficulty in predicting the final facial form. Kazanjian[23] proposed soft tissue repair during childhood but preferred delaying osseous surgery until after maturity. Obwegeser[24] advocates delay of both the soft tissue repair and surgery on the facial skeleton until facial growth has ceased. Conversely, the reasons for early correction are as follows[6] : (1) creating interocclusal space may promote a more normal eruption pattern of the teeth and stimulation of alveolar bone development on the affected side, (2) soft tissue development on the affected side is stimulated, and (3) there are psychologic advantages to the child and parents in observing esthetic and functional improvement.

Three-dimensional correction of the end-stage adult deformity of hemifacial microsomia consists of an operation to level the maxilla and piriform aperture, to make the mandible symmetrical and to place the temporomandibular joint in its proper location (coronal plane). Abnormalities in mandibular and maxillary width (transverse plane) are corrected orthodontically or at the time of operation. In the sagittal plane, the maxilla and mandible are mobilized in the direction dictated by the relationship of these structures to the cranial base.

The first step in planning the operation is to determine the proper location for the temporomandibular joint[5]. The next step in correction of the end-stage adult deformity is to place the maxilla in its correct position by a Le Fort I osteotomy. It is important to choose the correct fulcrum for maxillary repositioning. If there is vertical maxillary exess, the fulcrum of rotation of the maxilla is on the abnormal (short) side, leveling the occlusal plane without midface elongation. If the vertical length of the midface is normal, then the fulcrum of rotation is in the midline, so midface length does not change. If the midface is short, the fulcrum is on the normal side to provide maximum midface lengthening while leveling the occlusal plane. Once the maxilla is repositioned, bilateral mandibular osteotomies are required to rotate the lower jaw into its correct relation to the maxilla.

In instances in which the ramus of the mandible is missing and no mandibular articulation is discernible, a reconstruction of the temporomandibular joint may be justified[25]. At the present time, autogenous grafting is the only practical biologic approach to reconstructing the temporomandibular joint. Possible donor sites include the metatarsal bones, costochondral junctions, head of the fibula, and sternoclavicular joint [26,27]. In order to establish a suitable articulation, the graft must be fixed securely to the body of the mandible and ramus and placed against the zygomatic portion of the temporal bone. In adults, Obwegeser[24] has advocated reconstruction of the hypoplastic temporal and malar bones prior to mandibular grafting. In children, there is some evidence to suggest that joint function stimulates morphogenesis, and any necessary onlay grafting of the malar and temporal bones would appear best delayed until facial growth is completed.

Improvement of facial asymmetry may be attained by camouflage procedures[28]. But in those instances of moderate-to-severe hemifacial microsomia in which treatment has been delayed until after completion of facial growth, correction of both the facial asymmetry and the malocclusion may require surgery in both the maxilla and mandible[6].

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Hemifacial Microsomia의 외과적 교정 1례

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국문초록

Hemifacial microsomia는 편측의 측두 하악 관절과 하악지의 발육장애 및 저작근의 발육장애를 특징으로 하며 종종 상악골과 협골의 발육장애도 동반한다. 또한 관련된 외이의 발육장애 및 외이도의 부재, 이하선 및 도관의 부재, 안면 신경 및 안면 표정근의 기능 저하를 보이기도 한다.

전형적인 것은 출생시에 이미 알 수 있는데 이러한 변형은 주로 편측에 나타나며 하악골의 후퇴, 안면 비대칭, 부정 교합등이 존재하게 된다.

이 선천성 기형의 원인은 불명이며 그 치료로는 외과적으로 변형을 교정하는 것이다.

저자들은 22세된 여자 환자에서 hemifacial microsomia를 관찰하고 악교정 수술에 의해 만족할 만한 안면의 대칭 및 기능의 회복을 얻을 수 있었기에 문헌고찰과 함께 보고하는 바이다.