228

200

UNIQUE CRANIOFACIAL DEFORMITIES OF EXTREME SEVERITY AND INDIVIDUAL SOLUTIONS OF TREATMENT

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Abstract

The purpose is to present 5 patients with unusual severe craniofacial deformities.

During the last 14 years several hundred patients with craniofacial deformities have been treated. Five very severe and rare cases of deformities were selected: 1) A patient with a systemic fibrous dysplasia and extreme distortion of the face. 2) A patient with extreme neurofibromatosis I (NF I), probably caused by nuclear exposure. 3) A patient with otocephaly with separation of the oral and pharyngeal cavity with fusion of the masticatory muscles and missing of the lower jaw (agnathia) and tongue. 4) A patient with amniotic bands within bilateral oblique facial clefts. 5) A patient with extreme hyperteleorbitism and a cleft palate 50 mm in width, caused by an encephalocele which had developed into the oral cavity and contained the pituitary gland.

Treatment for all patients was considered dangerous or impossible in other departments. The surgical corrections of the 5 patients were very individual and have been performed in multiple stages. A satisfactory functional and aesthetic outcome could be achieved.

Today, craniofacial deformities of even extreme severity can be treated successfully.

Introduction

During the last 14 years about 1000 patients with major craniofacial deformities (cleft lip and palate patients excluded) were treated in the Department of Craniomaxillofacial Surgery. Every surgeon will consider a different degree of malformation as severe depending on his or her experience.

We present 5 patients with very special pathologies which we would consider severe malformations.

Case Reports

Case 1 (Systemic fibrous dysplasia with total distortion of the face)

This patient was seen in our department when she was 14 years old. The first swelling in the face was noticed by her family when she was 2 years old. Then the maxilla and mandible were constantly and symmetrically expanding in a way that after 12 years her orbits were fully covered by the enlarged protruded maxilla. The eyes were still present but dislocated cranially with a constant upward gaze and the patient had reception only to bright light.

At the age of 6 and 7 years two surgical attempts had been made elsewhere to reduce the tumorous masses. However, the disease process could not be stopped.

Clinical and radiological work up showed a massively enlarged mandible and midface with changes of the bones typical for fibrous dysplasia. The cortical bone was at large diminished or absent. The spongiosa was in some areas very translucent with pseudocysts and showed in other parts as sclerosis. The teeth were displaced in all directions and were at large dysplastic. All bones of the cranium were involved with irregular enlargement, obstruction of the orbits with compression of both eye balls and compression of the nose. The oral cavity was totally displaced and enlarged.

Histology showed the typical features of fibrous dysplasia with replacement of bone by fibrous tissue, generative changes with old bleedings, pseudocysts and giant cells.

As the disease is polyostotic the child had multiple deformities of the extremities and suffered every year several pathological fractures. This resulted in the inability to walk.

Up to now the patient underwent two major operative corrections: At the age of 14 years a subtotal

resection of the midfacial bones was performed. At the same time the hard palate was repositioned 1.5 cm cranially and the orbital floors were reconstructed using lyophilised cartilage. Altogether about 500 cc of dysplastic bone was resected from the midface. In a second procedure two years later the mandible was subtotally resected. Only the lingual cortex was left in place. Reconstruction was performed again with lyophilised cartilage.

The most striking result after the first operation was that the patient could see normally after the first operation, disturbed only by a minor degree of diplopia. The patient is now 16 years old and one has to anticipate further enlargement of the non-resected bones.

Case 2 (Neurofibromatosis I, probably caused by nuclear exposure)

The family of this patient lives in Kazakhstan and the mother was exposed to nuclear radiation during the first five months of pregnancy. At birth the patient showed multiple areas with changes of pigmentation and developed within the first six months large neurofibromas in the face and over the whole body. The tumour masses were growing and became lobuled and finally "flaps" covering the face which did not look human anymore. The patient is blind. The diagnosis of neurofibromatosis type I was made. The patient and his mother were investigated genetically and it was speculated that two mutations occurred at the same time. A mutation analysis is underway.

In one long session the tumorous masses were extensively reduced and plastic reconstruction was undertaken.

Case 3 (Otocephaly syndrome)

This patient with otocephaly was born without a mandible and tongue and showed at birth a complete separation between the oral and pharyngeal cavities. There was fusion of the masticatory muscles. A tracheostomy was performed and the child survived. The patient had been fed by gastrostomy for the first 16 months exclusively.

Then an operation was performed to connect the oral cavity with the oral pharynx by separating the masseter muscles. The macrostoma was also corrected.

Case 4 (Amniotic bands within bilateral oblique facial clefts)

Here we report a case of bilateral oblique facial clefting with amniotic tissue attached to the entire bottom of both clefts. Fetal ultrasound revealed an amniotic cavity with an amniotic septum and later a hydramnion. At birth amniotic bands inside the mouth and in both facial clefts were found. Careful clinical examination and computed tomography including 3D CT failed to classify the deformity into known classification systems (Tessier, AACPR, Boo-Chai, Karfik, Van der Meulen). Thus the bilateral clefts were clinically and radiologically "atypical".

Histology from the entire length of both oblique clefts (10 cm on the right side, 9 cm on the left side) revealed amniotic tissue which was attached partially to the periosteum and bone. Therefore we consider this patient so as to prove the hypothesis that amniotic bands can cause oblique facial clefts.

The patient is now six years old and underwent so far several procedures: removal of all amniotic bands, reconstruction of the cheeks, the lips, and the hard and soft palate. The next step is elongation of the short mandible by distraction osteogenesis.

Case 5 (Teleorbitism with encephalocele intraorally, causing a cleft palate 50 mm in width)

This patient had a teleorbitism of 45 mm, duplication of the nose and an encephalocele which had developed into the oral cavity, containing the pituitary gland. This encephalocele caused a cleft palate 50 mm in width.

In multiple reconstructive procedures the teleorbitism was corrected and the hard and soft palate reconstructed.

Conclusion

Today, craniofacial deformities of even extreme severity can be treated successfully.