

Reconstruction With Autoclavized Dysplastic Bone Graft in Craniofacial Fibrous Dysplasia: A Case Report

Kraniofasyal Fibröz Displazide Otoklavize Displastik Kemik İle Rekonstrüksiyon: Bir Olgu Sunumu

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Abstract: Fibrous dysplasia is a congenital, benign and progressive disease of bone. It is characterized by the replacement of normal bone by fibro-osseous tissue and when craniofacial region is involved enlargement of the affected bones, neurovascular compression and facial deformities can be seen. Complete resection of the affected bones and immediate reconstruction with unaffected cranial bone grafts is the best choice for the treatment of craniofacial fibrous dysplasia. When the dysplastic area involves a significant portion of the craniofacial skeleton as if in here presented case, autoclaved dysplastic bone can be also used for reconstruction.

Key Words: Autoclavized bone graft, fibrous dysplasia, reconstruction

Özet: Fibröz displazi konjenital, benign ve progresif seyirli bir kemik hastalığıdır. Normal kemiğin yerine fibro-osseoz bir dokunun geçtiği bu hastalıkta kraniofasyal bölgede etkilenen kemiklerde kalınlaşma nörovasküler yapılarda bası ve yüzde deformite meydana gelebilir. Etkilenen kemiklerin tam olarak çıkarılması ve aynı seansta sağlıklı kranial kemik greftleri ile onarımı en uygun tedavi seçeneğidir. Sunulan vakamızda olduğu gibi, kraniofasyal iskeletin büyük bir bölümü etkilenmişse displastik kemiğin otoklavize edilerek yerine iade edilmesi, diğer bir rekonstrüksiyon seçeneğidir.

Anahtar Kelimeler: Fibröz displazi, otoklavize kemik grefti, rekonstrüksiyon

INTRODUCTION

Fibrous dysplasia is a benign, metabolic, nonhereditary and congenital bone disease that is characterized by the replacement of normal bone by fibro-osseous tissue. It usually appears during the first two decades of life. Although it is a pathologically benign disease, it may show a malignant clinical progress. There are three variants

of the disease: monoostotic form, polyostotic form and McCune Albright syndrome. Contrary to first descriptions, the disease is monoostotic in 70% of the patients and lesions most commonly involve femur and ribs. Polyostotic form usually occurs in the craniofacial skeleton and sphenoid and frontal bones are the commonly affected bones (2,8). Skull base and orbital involvement may result in entrapment of the neurovascular structures and symptoms related to

compression can be encountered. Patients usually present with complaints such as deformity of face, atypical facial pain and headache, progressive visual loss, hearing loss and epiphora (8). In this article we present a case of polyostotic fibrous dysplasia with extensive craniofacial involvement that was treated with radical resection of the affected bones with intracranial optic nerve decompression and reconstructed with reshaped and autoclavized dysplastic bone grafts.

CASE REPORT

31-year-old-man was presented with a swelling on his glabellar region that had been slowly growing over the past two years. On examination a hard mass that extends to the frontal region was detected. Plain X-rays revealed ground glass appearance on the frontal bone and litic-sclerotic alterations on the periorbital bony structures. Computed tomography scanning showed extensive enlargement of the frontal, parietal, ethmoid and sphenoid bones (Fig. 1). Visual field examination and laboratory findings were all in normal limits. On the base of these findings surgery was performed under the

presumptive diagnosis of fibrous dysplasia. A bicoronal flap was elevated to expose the fronto-parietal, orbital and nasal regions. When the temporal fascia and orbital contents were dissected in the subperiosteal plane it was seen that the frontal, parietal, nasal and sphenoid bones were severely affected and significantly enlarged (Fig. 2). With the help of frontal and parietal craniotomies and fronto-

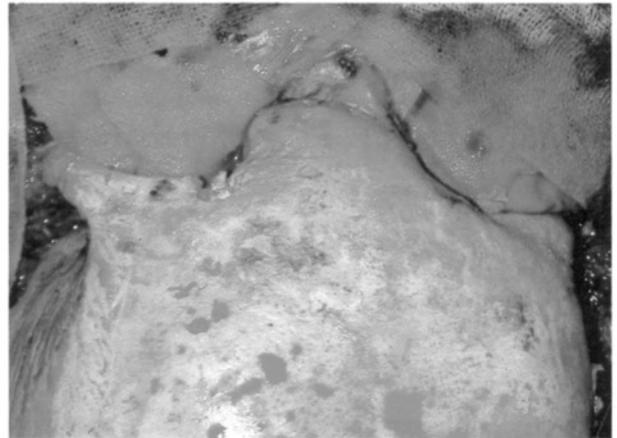


Figure 2: Intraoperative view of fronto-orbital region after coronal flap was elevated.

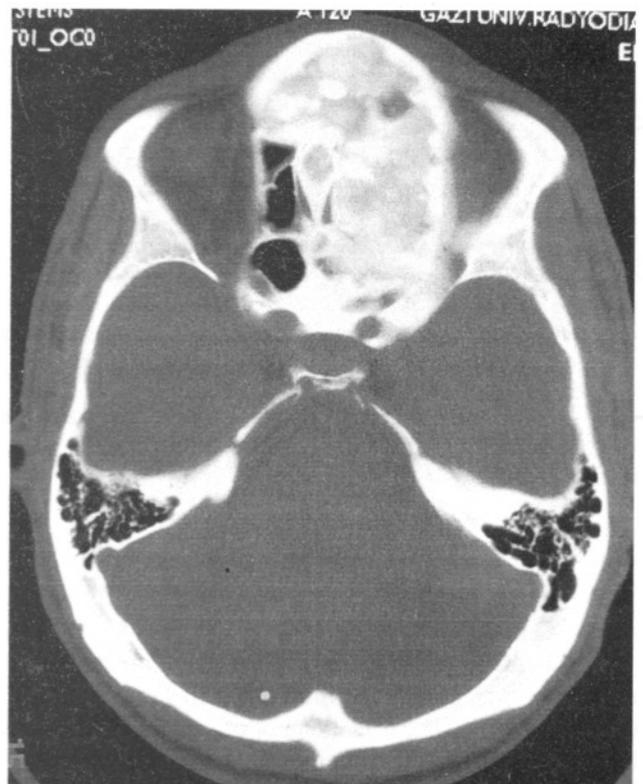
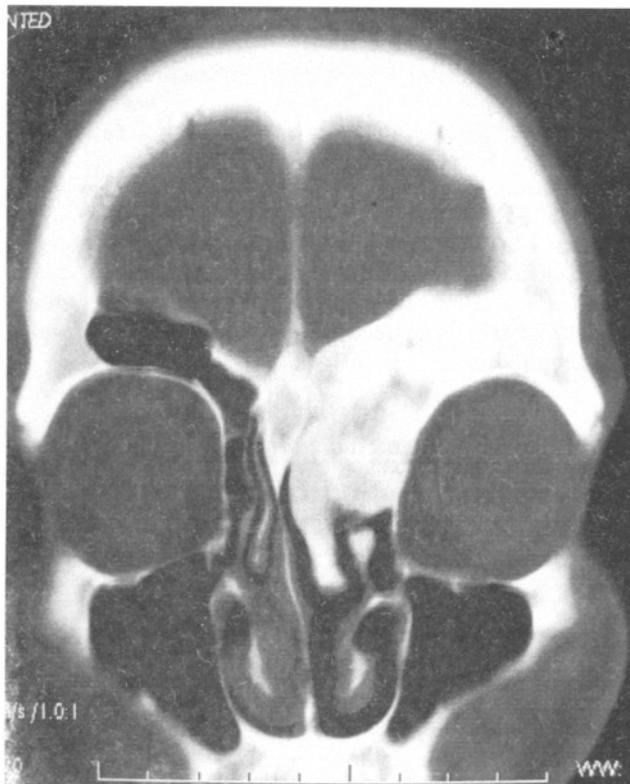


Figure 1: Coronal and axial sections of preoperative CT scans, demonstrating the involved parietal, frontal and sphenoid bones and narrowing of the left optic canal.

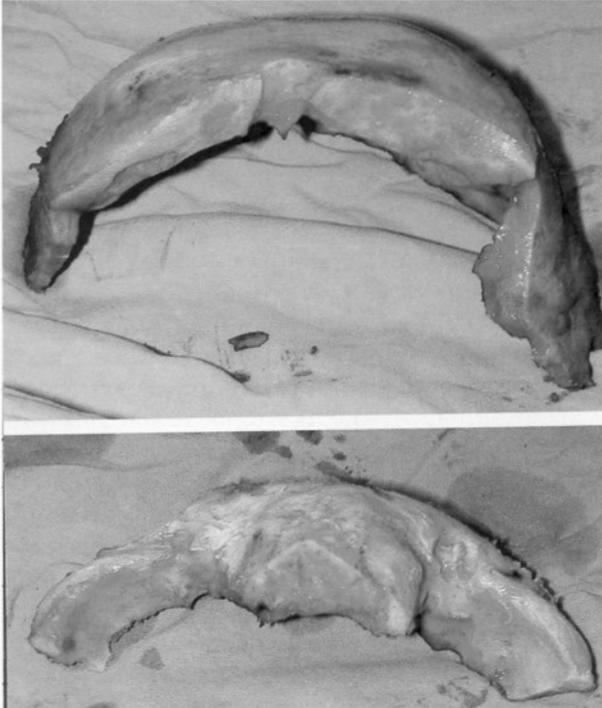


Figure 3: Fronto-parietal and fronto-orbital bones were removed with craniotomies. Dysplastic alterations were significantly enlarged the affected areas of the bones.

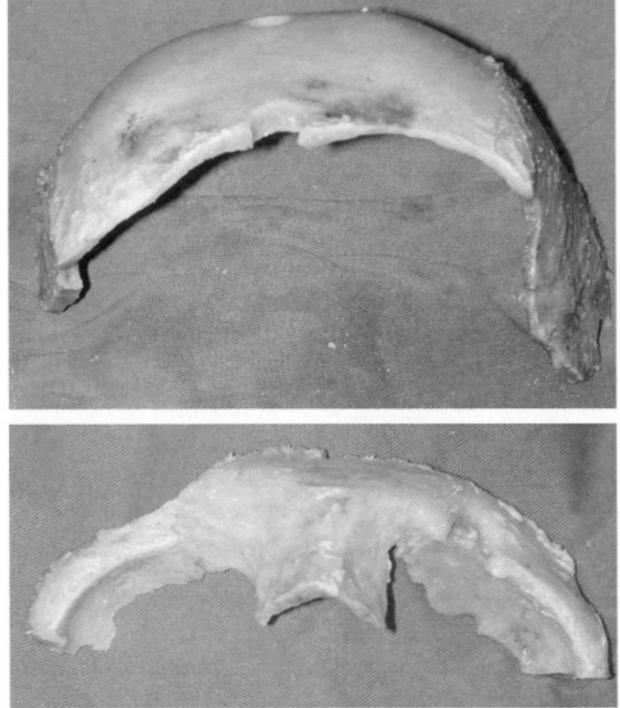


Figure 4: Appearance of the bones that were countered down with a high speed pneumatic drill and autoclaved for 30 min. at 120 °C.

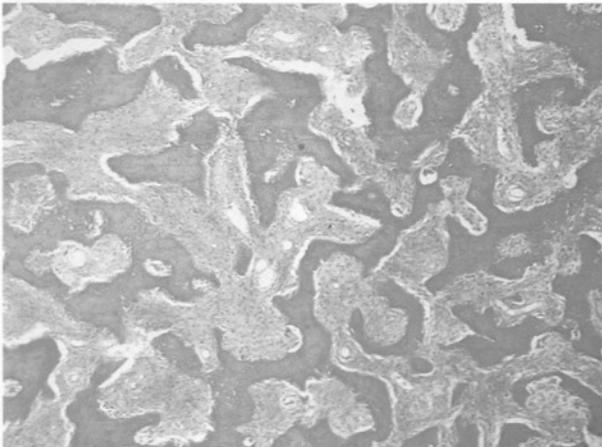
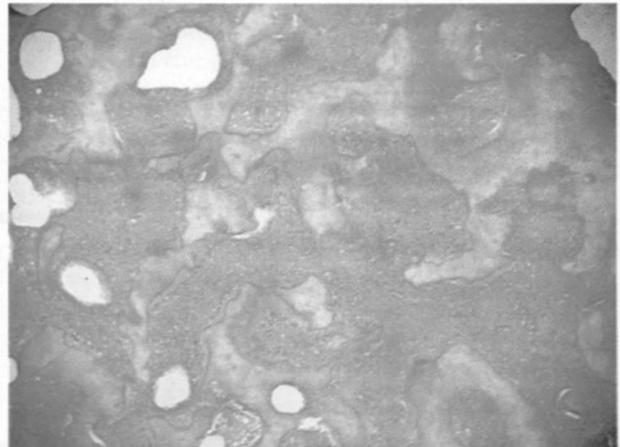


Figure 5 a: Histopathological appearance of the dysplastic bone, showing the fibro-osseous bone formation with in the fibrous stroma. (HEX40)



b: Histopathological appearance of the dysplastic bone after autoclaving, showing destructed cells.(Hex100)

zygomatic, fronto-maxillary, fronto-orbital osteotomies both the fronto-parietal area and then fronto-orbital bar were removed in two pieces (Fig. 3). Affected areas of the nasal and sphenoid bones were countered down until a margin of no affected bone was reached by using a high-speed drill.

Although, there was no visual field defect, prophylactic optic nerve decompression was performed because of the marked narrowing of the optic canal. Fronto-parietal and fronto-orbital bones, which had been removed, were countered down to their normal thickness and autoclaved for 30 min. at

120 °C. These autoclaved dysplastic bones were then replaced back to their anatomical places for reconstruction and secured by using titanium 2.0mm plate and screw system (Fig. 4). Then the coronal flap was closed over two 10-mm. silicon suction drains.

Histopathologic examination of the dysplastic bone demonstrated bony trabecules resembling Chinese letters in a rich fibroblastic stroma (Fig. 5a). This typical histopathologic appearance confirmed the diagnosis of fibrous dysplasia. The dysplastic bone was also examined following autoclavization, which revealed significant deformation of the dysplastic cells (Fig. 5b).

The postoperative period was uneventful and the patient was discharged from hospital in 8 days. The follow up period extending over two years showed neither recurrence nor severe bone graft resorption (Fig 6).

DISCUSSION

Although, fibrous dysplasia has been described, as a kind of proliferative fibro-osseous tissue secondary to bony metaplasia, its etiology was unclear. Recently it has been determined that the

disease is due to a functional disorder of the osteogenic cells. This functional disorder appears to be caused by a mutation of the a subunit of G signaling protein (Gs-a) in osteoblastic cells (1,11). As a result of this mutation, uncontrolled production of fibrotic bone matrix and overgrowth occurs at the affected bones.

Histologically irregularly spreaded bone trabecules (Chinese letters) and rarely cartilage islands have been detected in a fibrous matrix. The disease starts from the medullar cavity and expands the bone without disturbing its cortex.

The radiological appearance depends on the location of the lesion and the proportions of fibrous and osseous structures. They can appear as sclerotic, litic or mixed type. The bone cortex more and rapidly expands in thinner bones such as orbital plate of maxillary, ethmoid and frontal bones than in thicker cortical bones. On the other hand, expansion is slower and the lesions have sclerotic appearance in thicker bones. Computerized tomography is the most useful radiological study to determine the location and extend of the lesion. Where as MR can be especially helpful for the evaluation of neurovascular structure compressions (2,8). Bone scintigraphy is also useful

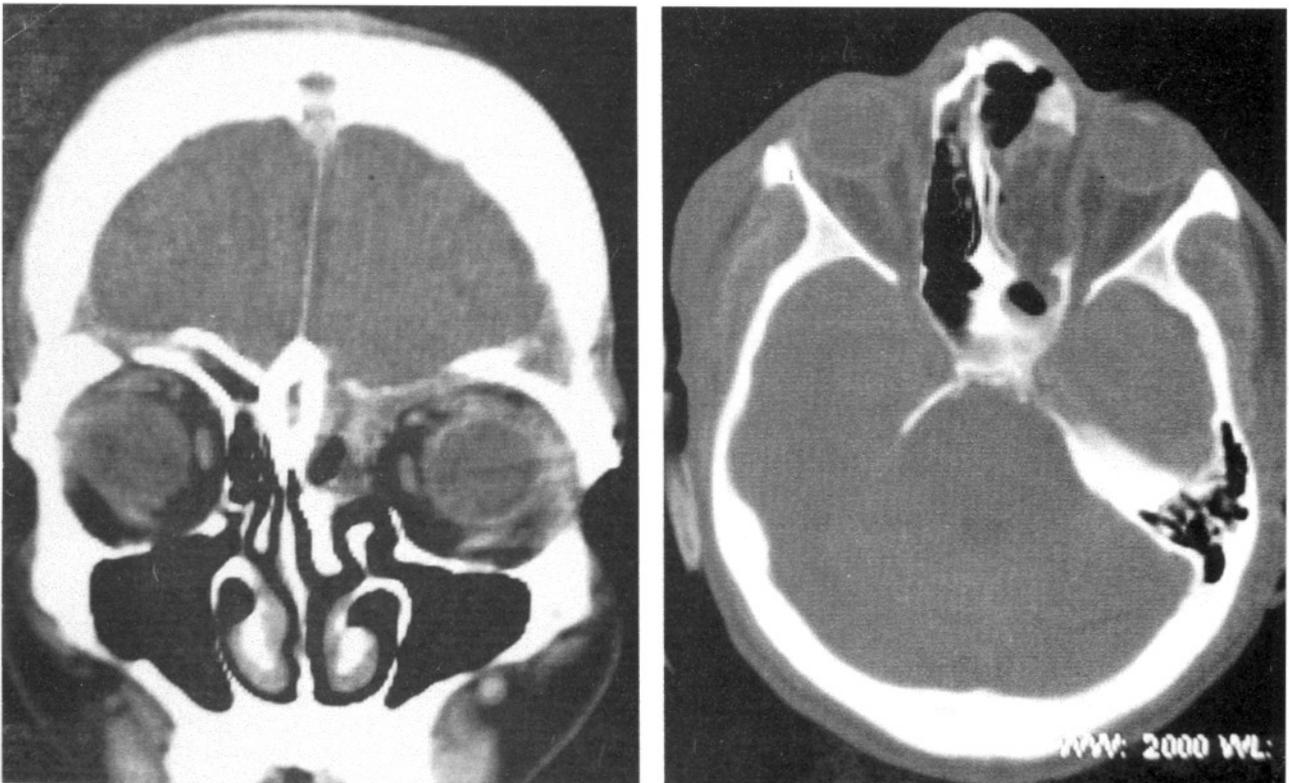


Figure 6: Two years after surgery: there is no recurrence or severe bone graft resorption in follow up cranial CT scans.

and sensitive tool in evaluating the full extent of polyostotic fibrous dysplasia (3).

There is no specific laboratory study to diagnose fibrous dysplasia other than histopathologic confirmation. However high serum alkaline phosphatase and urinary hydroxyproline levels may be detected in some of the patients (6,11).

Nonsurgical treatment of the fibrous dysplasia is controversial. Radiation therapy is not recommended due to both lack of response and potential of malignant transformation to osteosarcoma or chondrosarcoma. Although, medical agents such as corticosteroids, aluminum acetate, biphosphonates (pamidronate, etidronate etc.) and mithramycin have been used for the treatment of the disease, their benefits are minimal and temporary. Therefore, radical surgery and immediate reconstruction is advocated by many for the treatment of fibrous dysplasia. However, there is no consensus on the timing and extensiveness of surgery or on the methods of reconstruction. Occasionally, the disease can be self-limiting so some physicians recommend that the surgery must be delayed until the post puberty. On the other hand, others are in favor of early extensive surgery as incomplete resection is associated with 25% local recurrence (2,6,8,10,11).

There are three alternatives for reconstruction of defects. The first option is reconstruction with autogenous bone grafts. Iliac, costal and cranial bone auto grafts have been used for the purpose of reconstruction for many years. However their usage can be troublesome due to donor site morbidity and limited supplies. Therefore, when extensive resections are planned autogenous bone grafts fail to provide adequate reconstruction material. Another problem with this option is the difficulty of giving the desired shaped to the bone grafts, especially when complex anatomical places are needed to be reconstructed.

The second alternative is reconstruction with synthetic materials. Some of surgeons prefer large methyl methacrylate implants for the reconstruction of extensive cranial defects when autogenous bone grafts are insufficient. Recently hydroxyapatite implants are popularised, as unlike methyl methacrylate, they do not cause exothermic reaction and inflammatory response. Donor site morbidity is avoided with the use of these synthetic materials. On the other hand, to shape these materials so that they

can fit into the areas that are going to be reconstructed is much more difficult than autogenous bone grafts (2,10).

The last choice is the usage of autoclavized dysplastic bone grafts. The use of dysplastic bone after boiling or autoclaving for the reconstruction of the large bony defects has been performed in orthopedic surgery since 1937 (4). As for craniofacial surgery it was first used by Lauritzen et al. (7) in 1985. Reposition of the cranial bone that belongs to the patient reduces the operation time significantly and also provides the excellent cosmetic results. This also reduces the problem of bone availability, donor site morbidity and tissue reaction and provides a perfect fit. However, the delayed bone healing and bone graft resorption can be problematic in some cases making this option to be left only for very extensive involvements. When compared with nonautoclaved healthy bone grafts they poses a longer healing period during which they might show a much more pronounced resorption. Although, this may lead us to think that this option is much more secure in smaller defects, one should remember that there are better alternatives in terms of healing and final result for such reconstruction needs. However, for the larger defects as in here presented patient the reconstruction options are quite limited and the surgeon might prefer autoclaved bone only because the other options can not provide superior results which is not the case in small defects.

Histological studies of the autoclaved bone showed that autoclavization destroy the entire cellular structures in the bone. On the other hand, the noncellular frame is protected to some extent. Following implantation of the autoclaved bone revascularisation, migration of osteoblasts from adjacent healthy bone, resorption of dead structures and formation of new bone occur (4,7,9). Biomechanical properties of bone also get weak after autoclavization. Köhler et al. showed the decrease in strength and stiffness of autoclaved bone that is closely related to the autoclaving time and temperature. The loss of biomechanical properties is minimum for high temperature-short time applications. Although, this weakness can be a cumbersome for weight-bearing bones like femur and tibia, it is not a problem for skull (5). Several authors recorded their concern that devitalized tissue may increase the risk of infection. However, equal infection rates were found for autoclaved bone grafts when compared to the nonautoclaved ones in experimental studies (9).

CONCLUSION

Fibrous dysplasia is a local aggressive disease that is characterized by the replacement of normal bone by fibro-osseous tissue. Complete resection of the affected bones and immediate reconstruction with unaffected cranial bone grafts is the best choice for the treatment of craniofacial fibrous dysplasia. When the dysplastic area involves a significant portion of the craniofacial skeleton as if in here presented case, autoclaved dysplastic bone can be also used for reconstruction. This technique is not a first choice and other techniques like bone grafts or alloplastic materials must be preferred in reconstruction of small defects. However if the defect is large, and difficult to be reconstructed with synthetic materials or bone grafts, then the use of the autoclaved affected bone segments come forward.

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Computed tomography characteristics of non-syndromic craniofacial fibrous dysplasia.

Chen YR, Wong FH, Hsueh C, Lo LJ.

Fibrous dysplasia is a benign fibro-osseous tumor of bones commonly involving the craniofacial region. Computed tomography (CT) imaging study of the disease is useful for evaluation and treatment planning. The average number of bones involved was 3.2 bones per patient. Involvement of more than one craniofacial bone occurred in 70% of patients. The maxilla, orbital, and frontal bones were most commonly involved. CT images appeared sclerotic or homogenous in 34%, mixed white and dark or heterogenous in 55%, and cystic in 11%.