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CASE REPORT

Pediatric/Craniofacial

Orbitocranial Fibrous Dysplasia: Immediate Reconstruction with Titanium 3-dimensional Printing

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Summary: We present the case of an 18-year-old man who was referred to our department with a suspected tumor in the right orbital region, which caused exophthalmos due to its pressure effect. Preoperative CT imaging revealed an expansile ground-glass appearance of bone mass, with sclerotic bone on the right frontal bone (right superior orbital wall). There was no orbital mass and no enlargement of the extraocular muscles and tendons; the retrobulbar fat and optic nerve appeared unremarkable. A preoperative tissue biopsy confirmed the FD. Complete tumor removal was performed via the right hemicoronal approach. The orbital roof and temporal bone defect were immediately reconstructed using the 3-dimensional titanium printing plate. *(Plast Reconstr Surg Glob Open 2020;8:e3114; doi: 10.1097/GOX.0000000000003114; Published online 21 September 2020.)*

INTRODUCTION

Fibrous dysplasia (FD)[1] of bones was first described by Von Recklinghausen in 1891. Lichtenstein coined the term in 1938.[2] FD is a benign fibro-osseous developmental disorder of the growing bone in which fibrous and osseous tissue replace the normal medullary bone. It represents approximately 2.5% of all bone tumors and causes 5%–7% of benign bone tumors. FD is mostly observed between years 3 and 15, with the majority of patients diagnosed before the age of 30 years,[3,4] and it is equally distributed in both sexes.[5] Craniofacial FD describes lesions confined to the bones of the craniofacial skeleton; it occurs in 25% of all patients with FD. The most affected bones in craniofacial FD are the maxilla, mandible, sphenoid, ethmoidal, and frontal bones, while occipital bones are less frequently affected.

No medical treatment is available to cure or prevent FD. There have been some medical trials to stop the progression with bisphosphonates, calcitonin, and mithramycin, but they showed little success.[6,7] Systemic steroids are used for acute visual loss due to optic canal compression as temporary therapy until the patient can undergo optic nerve decompression surgery.[8] Radiotherapy plays no role and carries a high risk of malignant change.

Conservative surgical therapy such as curettage, contouring, or remodeling are always associated with sub optimal results, and surgical revision may be needed if the disease progresses, while radical resection is potentially curative.[9] However, craniofacial FD, especially in the orbital region, makes it difficult to reach optimal cosmetic outcome after immediate reconstruction due to the anatomic variation of each patient.[10] The aim of immediate reconstruction was to restore contour, symmetry, orbital volume, intact cranial base, and barrier. Different materials have been used to reconstruct the cranio-orbital defects, such as autogenous bone grafts, chondrocostal grafts, titanium meshes, and other alloplastic material. Nowadays, synthetic materials such as polyethelene (Medpor) and titanium mesh are increasingly used globally due to the advantage in donor site morbidity when compared with autogenous tissue.[11,12]

Three-dimensional (3D)-printing titanium alloys (Ti alloys) technique "additive manufacturing" (AM) is a technology that adds material layer-by-layer to fabricate a 3D object. The metal AM was developed in 1920 by Baker (US patent, 1,533,300), who put forth the concept of layer manufacturing to create walled structures and decorative articles using an electric arc and metal electrode. It was used to develop directed energy deposition techniques that integrate the idea to fabricate a part directly from computer-aided design (CAD) data.[13] From the 1960s to the 1990s, there were several AM technology and 3D-printing technology released to the global industry. Until in 1997, the laser additive manufacturing (LAM) was developed early for use in high energy powder to produce titanium parts from metal powder by the AeroMet company.[14]

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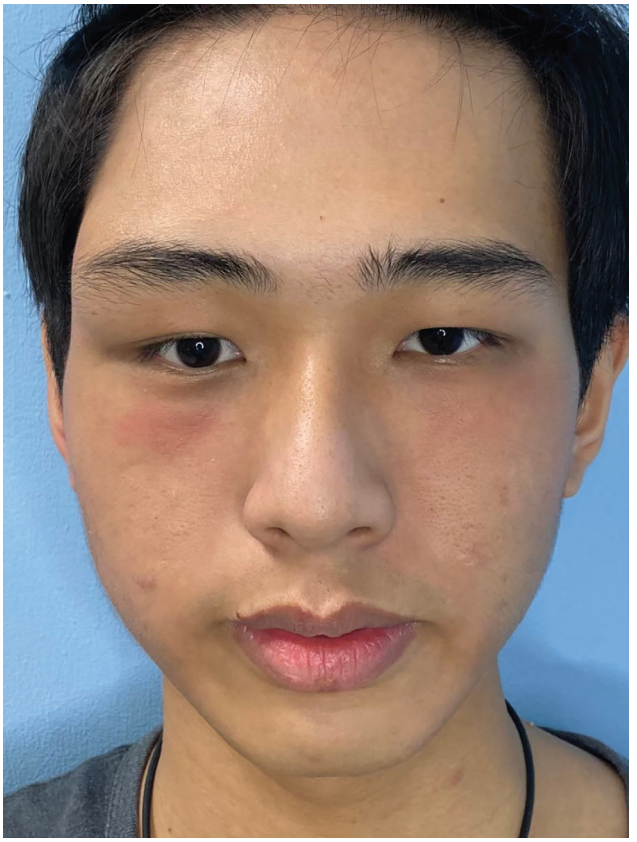

Fig. 1. Clinical presentation, showing a right orbital mass caused exophthalmos.

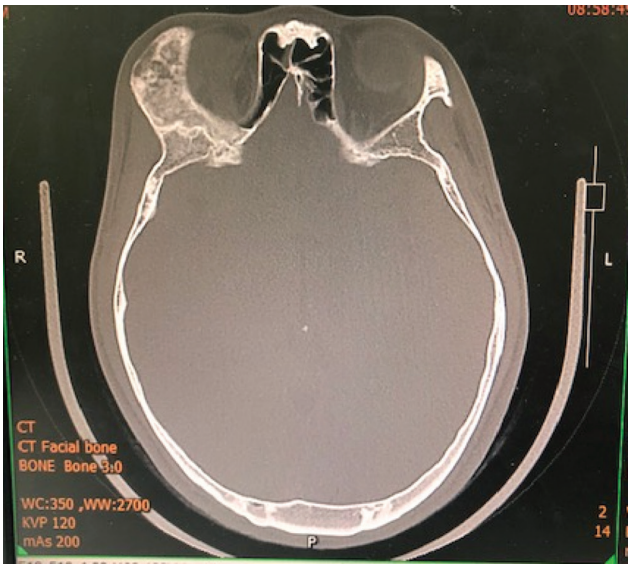

Fig. 2. A CT scan image showing an expansile ground-glass appearance of bone mass with the sclerotic bone on the right frontal bone.

CASE REPORT

An 18-year-old man was referred to our department with a suspected tumor in the right orbital region, which caused exophthalmos due to its pressure effect (Fig. 1). He observed that the lesion developed 3–4 years ago, which slowly progressed and stopped growing for a year. Physical examination of the lesion showed a nontender swelling on the right supraorbital region responsible for proptosis of the right eye. His visual acuity and visual fields were normal. He had no significant pain and discomfort. Preoperative computed tomography (CT) revealed an expansile ground-glass appearance bone mass with sclerotic bone at the right frontal bone (right superior orbital wall), which measured about $4 \times 3 \times 5\,cm^3$. There was no orbital mass or enlargement of extraocular muscles and tendons; the retrobulbar fat and optic nerve appeared unremarkable (Fig. 2). Preoperative tissue biopsy confirmed FD. The resection line was discussed and planned by mapping on a 3D CT of the facial bone after which a customized titanium 3D printing was prepared. The neurosurgery team was consulted for the craniotomy and intracranial resection.

To fabricate the patient-specific implant, a CT scan image of the cranium and maxillofacial bone was first acquired according to the imaging protocol. Data were stored or were kept as Digital Imaging and Communications in Medicine (DICOM), where the DICOM files must have a layer thickness within the acceptable range (≤1 mm). These DICOM files consisted of a 2-dimensional slice from the anterior to posterior side of the patient's skull. The 3D reconstruction was then carried out to segment the anatomical bone, as well as to identify the morphological landmarks via the Avizo software (Thermo Fisher Scientific, Ma.) (Fig. 3).

First, the tumor region was marked according to the resection plan. Subsequently, 3D mirror reconstruction technique was used to create a contour surface for restoring the bone defect. The mirroring was based on the mid-sagittal plane and assumed that the anatomy was symmetrical. A pre-contoured mesh of the implant with screw hole fixation was then designed based on anatomical bone characteristics and the surgeon's specific requirements using the Solidworks software (Dassault Systèmes, France).

Upon the surgeon's approval of the design, the patient-specific implant was 3D-printed by selective laser melting using medical grade Ti-6Al-4V alloy (Meticuly Co., Ltd., Thailand). Subsequent postprocessing techniques, including surface polishing and sterilization, were performed according to routine standards. The entire fabrication process was certified by ISO13485 standard for design, manufacturing, and sterilization of medical device.

The right hemicoronal approach was used to identify the lesion and completely remove the tumor. A 3D-printed titanium plate was used for immediate reconstruction, and the plate was fixed with a screw of 1.5 mm (Fig. 4). The patient postoperatively received antibiotics and analgesics. A pressure dressing was given and suction drains were removed after 48 hours postoperatively when the contents were <15 mL. Gross examination showed that the affected bone was expanded. The lesion was well-demarcated and intramedullary. The lesional tissue was dense, fibrous, gritty, and abnormally tan-gray in color. Microscopic findings revealed branching and anastomosing irregular trabeculae of woven bone ("C" and "S" shapes) with no conspicuous osteoblastic rimming. Intervening fibrous

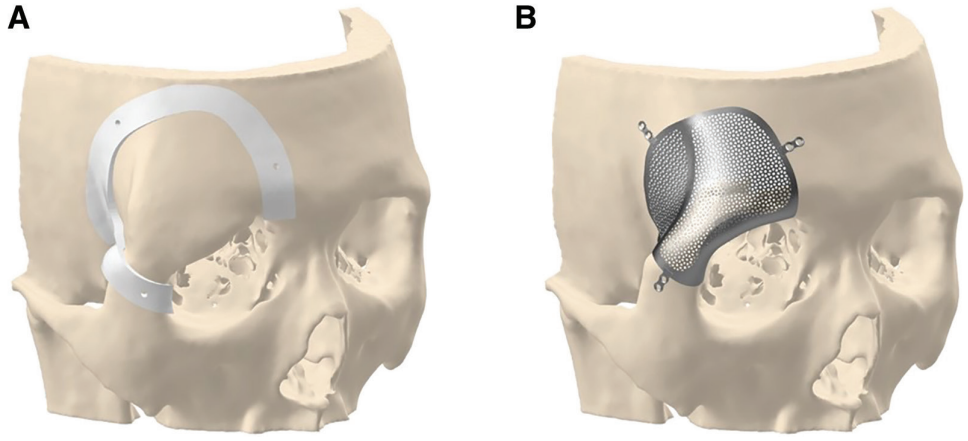

Fig. 3. 3D titanium plate simulation. A, Surgical cutting guide. B, 3D printing titanium plate.

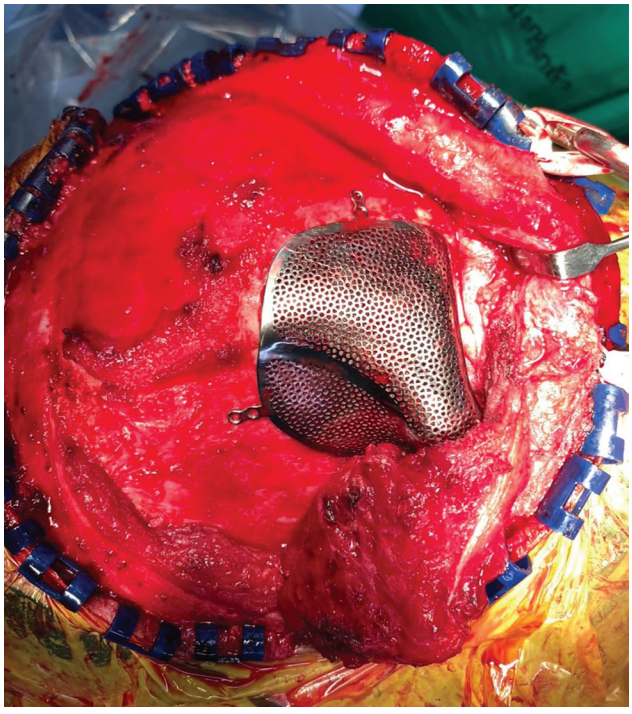

Fig. 4. Intraoperative field after reconstruction with the 3D titanium plate.

stroma containing cytologically bland spindle cells were present without prominent cytologic atypia. No mitotic figures were seen.

The patient's facial contour symmetry was observed over a 1-year follow-up. No abnormal feeling and pain were noted by the patient. No recurrence of the tumor was identified (Fig. 4). Our patient provided consent for the use of the case and images for publication.

DISCUSSION

FD is a non-neoplastic developmental hamartomatous disease of the bone, characterized by a blend of fibrous and osseous elements in the region. With an incidence of 1:4000–1:10,000, it is a rare disease.[10] Initially described as "osteitis fibrosa generalisata" by von Recklinghausen in 1891 in a patient with skeletal deformities due to fibrotic bone changes, the disorder became known as FD in 1938 when Lichtenstein introduced the term.[2,15] FD is associated with a gene mutation in the α subunit of the stimulatory G protein encoded by the gene on *GNAS1* gene[16,17] that affect the cells that produce bone. The germ-line mutation occurs in the early stages of fetal development. The gene mutation causes an abnormal proliferation of undifferentiated mesenchymal bone-forming cells, which fail to mature into lamellar bone (arrest of bone development in the woven phase) and lead to disorganized, poorly calcified fibrous bone trabeculae; so gradually the normal bone is replaced with fibrous bone tissue.[18]

This condition represents approximately 2.5% of all bone lesions and about 5%–7% of all benign bone tumors.[19] The 3 subtypes of FD are monostotic, polyostotic, and craniofacial. The term craniofacial FD is used to describe FD where the lesions are confined to contiguous bones of the craniofacial skeleton. Most cases of craniofacial FD cannot be truly categorized as monostotic because of the involvement of multiple adjacent bones of the craniofacial skeleton. They are also not truly polyostotic because bones outside the craniofacial complex are spared. They are seen in the first three decades of life and usually stabilize when the patient reaches skeletal maturity. The lesions show a rapid growth with cortical bone expansion with displacement of adjacent structures such as the eye and teeth in young children and prepubertal adolescents. The symptoms include facial asymmetry, visual changes, nasal congestion, pain, paresthesia, hearing impairment, and malocclusion.[20] FD involves the maxilla almost twice as often as the mandible, frequenting the posterior region and is usually unilateral in nature.

Many patients present with external visible aesthetic deformity. The most frequent finding of FD around the eye are dystopia hypertelorism due to the involvement of frontal, sphenoid, and ethmoid bones, and proptosis. Other findings are difficulty in lid closure, strabismus, optic neuropathy, nasolacrimal duct obstruction, tearing, and

trigeminal neuralgia due to skull base involvement.[21–23] In our case, the main clinical presentations were facial asymmetry and proptosis.

The management of cranio-orbital FD represents a dilemma for the surgeons because of the anatomic confines of the orbit and cranial base. The aim of the treatment of FD is to correct or prevent functional problems and to achieve an aesthetic improvement. The decision is between performing a total excision (sometimes very deforming) and just aesthetic recontouring of the deformed craniofacial skeleton with observation. A surgical success may turn out to be a social disaster if the resultant disfigurement is worse than the one produced by the disease.

If complete resection was considered good, reconstruction will play a main role for postoperative outcome after cure from the disease. A previous study showed good outcome by preoperatively using a standard skull model to shape and cut the mesh, a technique described by Andrades et al. The 3D-printing technique is more customizable and more fitting to each patient's defects and shows more cosmetic outcomes with no serious complications, including infection and extrusion in a 6-month follow-up.

Titanium and its alloys were originally used for producing aircraft components because of its high strength alloys and light weight.[24] From 2006–2008, the Arcam company released the patent electron beam melting machine with its titanium grade 2 material and worked with Adler Ortho Group to produce a medical device that resulted in European Conformity (CE-certification) for hip implants made from the Arcam machine. It makes an impact to use titanium alloy 3D printing for medical application.[14,25]

The Ti6Al4V(Ti64) alloy (one of the titanium alloys' composition) is widely used in medical application because of its low density, high strength-to-weight ratio, and superior biocompatibility.[26] Moreover, titanium alloy implants have excellent osseointegration properties, and its elastic modulus is only half that of 316L stainless steel.[27]Therefore, Ti64 alloys with the 3D-printing technique was commonly used to produce patient-specific dental or medical implants.[28–30]

In our case, it only took us 7 days to prepare the 3D-printed titanium. Hence, this short period of time can prove to be beneficial and be applied to tumor and malignancy cases in routine practice. Moreover, we can reduce intraoperative reconstructive time by 2–3 hours compared with using an autologous bone graft. Our material costs around 1600 USD. However, like that of other nonautologous prosthesis, the limitation of the 3D-printing technique is that it cannot be used while a child is growing; the operation should be performed once the development is stable.

CONCLUSIONS

Complete resection of cranio-orbital FD was potentially curative with no extra morbidity. It can achieve good aesthetic and functional results by using a 3D-printed plate. The process of preparing the material did not delay operative planning.

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PATIENT CONSENT

The patient provided written consent for the use of his image.

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