Visual Field Improvement Achieved by Decompression Surgery for Craniomaxillofacial Fibrous Dysplasia: A Case Report

Nana Li
Peking University Third Hospital
Xuhao Chen
Peking University Third Hospital
Yanjie Tian (fygeneral@sina.com)
Peking University Third Hospital https://orcid.org/0000-0003-4455-1376
Chun Zhang
Peking University Third Hospital

Case report

Keywords: Fibrous Dysplasia, Visual Fields, Decompression, Optic Nerve, Case Report

Posted Date: September 3rd, 2020

DOI: https://doi.org/10.21203/rs.3.rs-68496/v1

Abstract

Background

Craniomaxillofacial fibrous dysplasia (CFD) is a type of congenital benign bone disorder that progressively replaces the healthy bone marrow with fibrous tissue. When the optic nerve is influenced, the efficacy of therapeutic decompression is doubtful, as vision loss is irreversible with the thinning nerve fiber layer.

Case presentation

A 22-year-old female presented to our institution with left painless, slowly-progressing orbito-fronto-temporal swelling. She had undergone several plastic surgeries before. Enlargement in her left frontal area involving the orbital region was seen, with increased frontal contour and vertical dystopia and minimal inferior displacement of the globe. We observed significant visual field defects in her left eye, and her retinal fiber layer was thin on examination. She was therefore diagnosed as FD. Decompression operation was performed, and her visual acuity and central visual field improved in the 1-month follow-up.

Conclusions

Our case indicated that visual defect caused by prolonged time course of optic nerve compression by craniomaxillofacial fibrous dysplasia can be relieved partially by decompression surgery.

Introduction

Fibrous dysplasia (FD) is a non-neoplastic, non-hereditary, and congenital benign bone disorder that progressively replaces the healthy bone marrow with fibrous tissue, accompanied by thinning of cortical bone [1, 2]. Typically, FD was classified by the sites of pathological changes. Among them, the craniofacial region deformation is the most frequent [2]. Clinically, progressive swelling in affected area without pain was the most common chief complaint [3]. Other symptoms may include facial asymmetry, pathological fractures, and nerve compression [4, 5]. With deformation of the ocular bones, the optic canal becomes fragile due to its location in the center area. The efficacy of therapeutic decompression is doubtful as the previous study showed its failure to reverse the vision loss if the nerve fiber layer has displayed signs of damage in relevant ophthalmic examinations [2]. Regarding this, we reported a case of craniomaxillofacial fibrous dysplasia (CFD) with optic canal involved. Contracted visual field caused by compression to the optic nerve significantly improved after decompression surgery.

Case Presentation

A 22-year-old female presented to our institution with left painless, slowly-progressing orbito-fronto-temporal swelling in May 2018. At the age of five, her family noticed her forehead protuberance in the left

face for the first time and referred to the local hospital. She was then diagnosed as FD and underwent surgeries to improve her appearance. However, as residual diseases continued to enlarge, optic nerve was compressed and progressive contraction of visual field was found in her left eye afterward. She suffered from diplopia and exotropia caused by the exophthalmos and mandibular protrusion. Disorder of the upper and mandibular occlusion worsened as her facial asymmetry grew. The progression finally ceased at the age of 12, and over the past ten years, her symptoms remained stable. During the process, there was no evidence of endocrinological disorders or an apparent decline in her visual acuity. She began to seek further treatment for cosmetic purposes two years ago. Her maxillofacial malformation improved after three more surgical remodeling procedures on the left maxillary bone and mandibular zygomatic region. As her vision-threatening orbital deformities remained, she was referred to us for further ophthalmic treatments.

Her visual acuity was 20/25 in the left eye with normal color vision. Her physical examination showed severe enlargement in her left frontal area involving the orbital region, with increased frontal contour and vertical dystopia and minimal inferior displacement of the globe (Figure. 1). The patient had no café-au-lait macules. The fundus examination showed moderate disc pallor in her left eye, and the cup-to-disc ratio(C/D)was 0.7. The ocular movement in all directions was intact.

Visual evoked potential (VEP) tests demonstrated that both in the 15 and 60-second grid test, the amplitude of the a and b waves in the left eye were all moderately lower than those in the right. A dynamic visual field test revealed an inferior visual field defect (Figure. 3C). Optical coherence tomography (OCT) showed that the thickness of the nerve fiber layer above the left eye significantly decreased. Computed tomography (CT) scan revealed a left predominant expansion of the frontal, ethmoid, and sphenoid bones as well as of the temporal bone and the upper mandible. Additionally, the entire left maxillary sinus, ethmoid sinus, and bilateral frontal sinus cavities were obliterated. The lesions showed loss of normal corticomedullary differentiation, replaced by a homogeneous ground-glass appearance. There was also ocular proptosis in the left eye (Figure. 2).

Therefore, CFD was further confirmed. Reconstruction of the left eye socket was performed to improve her deformity. Under general anesthesia, bone tissue of the inner orbital wall was found proliferated, and the ethmoid sinus had disappeared. We therefore removed the bone tissue from the outer orbital wall to the apex of the orbit. (Figure. 3A). The CT image right after the operation showed the removal of bones, as well as the correction of the left eye exophthalmos (Figure. 3B). One month after the operation, her visual acuity was 20/20 on the left, and exotropia was significantly improved. There was a limitation of intraocular rotation but alleviated with time, and the central visual field was improved after the operation (Figure. 3C). After three months, her eye movement function had fully recovered, but its position remained oblique. There was no more significant improvement in the visual field.

Discussion And Conclusions

FD is a benign lesion with a low rate of malignancy. Typical classification of FD includes three variants: monostotic, polyostotic, and McCune-Albright syndrome (associated with endocrinopathies and skin hyperpigmentation) [2, 6]. CFD usually starts in infancy. After a slow growth, typical symptoms will appear in childhood or adolescence. It will then enter a stable stage and finally stop processing during adulthood. However, in a small proportion of cases, the course of the disease can reach 70 years [7]. In this case, the 22-year-old patient suffered significant facial deformities. She had a long history of progression, but facial deformity, visual acuity, and visual field remained stable about ten years ago. Her disease course was typical.

For the diagnosis of FD, clinical, radiographic, and histopathological features are usually necessary. Besides, examinations, including visual acuity, visual field, color vision, and VEP tests are crucial for ophthalmologists. Pathologically, spindle cells surrounded by a fibrous matrix will replace the normal bone [8]. CT representation of FD consists of three varieties: pagetoid, or ground-glass pattern (56%), sclerotic lesions (23%) and cystic variety (21%) [9]. The characteristics of FD in CT include the expansion of the involved bone with a heterogeneous pattern of CT densities associated with scattered or confluent islands of bone formation. Specifically, almost all CT findings of CFD show the glass-like changes [10]. Hence, it is usually the first choice to choose CT for the examination of CFD. As for our patient, our CT results showed a homogenous, “ground-glass” lesion, with a left predominant expansion of the ethmoid and the temporal bone. CT images after the surgery presented our removal of the mass. Differential diagnosis should include nonossifying fibroma, fibrous dysplasia of bone, aneurysmal bone cysts, osteoma, giant cell tumor of bone, low osteosarcoma, low degree chondrosarcoma, and fracture healing. History, location, and biopsy are essential for the identification of certain diseases [1].

There are no standard treatments for FD currently, but it is recognized that tailored approaches according to the characteristics of the patient will be more appropriate. Surgical treatments will be required when symptoms occur and progress, especially when bothering one’s daily activities. If the optic nerve is involved, either therapeutic or prophylactic decompression should start as soon as possible. Prophylactic decompression is performed to avoid further progression of visual loss caused by compression of optic canal stenosis, but the loss of vision is still difficult to control. While the effect of therapeutic decompression remains controversial, some experts believe that if the thickness of the retinal nerve fiber layer is in normal status, visual function can recover after decompression. On the contrary, if the nerve fiber layer has thinned, decompression cannot reverse the process, and the decline in visual acuity will progress [2]. The aims for surgical treatment are not only for cosmetic purposes but also to restore the physiological function of the affected organ. Although FD is usually of the multi-bone type involves a wide range of bones without clear boundaries, the recurrence rate is low in adult patients. Orbital bones will be removed to expose the surgical area, but extensive resection can lead to severe secondary deformities and damage to facial function. Therefore, sufficient hemostasis and precise operation are necessary to prevent accidental injury to the optic nerve. Otherwise, visual acuity may be irreversibly damaged.

During our examination, VEP displayed a functional loss of visual pathway, while OCT proved the structural damage to the optic nerve. After the removal of an appropriate amount of orbital bone during surgery, her appearance considerably improved. Surprisingly, visual acuity was preserved, and we observed an overall improvement in her visual function. Besides, the central visual field area enlarged at one month after surgery. Other than emergent decompression to salvage one's vision [11], our case indicated that even when the compression has damaged the optic nerve for a prolonged disease course, relieving the compression can still improve visual function partially. Possible explanation was that the central visual field might be recently affected, which was therefore reversible. However, overall or quadrant thickness of retinal nerve fiber, which was most commonly used to assess the structure of optic nerve, could not distinguish that. Such phenomenon further displayed the importance of early decompression surgery in order to salvage the potential function of vision.

The patient's eye position was still oblique after surgery, and the left eye was not able to turn inward, which was one of the limitations of our treatments. We believed that the extraocular muscle was damaged but functional exercise can accelerate her recovery. There is no definite information in the literature currently on how long it might take to restore the extraocular muscle movement to preoperative levels. This case is still being followed-up and we wish to determine whether the oblique position can recover and provide reference data for the treatment of future patients.

Conclusions

CFD is a type of disease that may significantly influence one's visual function. Our case indicated that decompression may improve the visual function even the compression of optic nerve exists for a prolonged disease course.

Abbreviations

CFD: Craniomaxillofacial fibrous dysplasia, FD: Fibrous dysplasia, VEP: Visual evoked potential, OCT: Optical coherence tomography, CT: Computed tomography.

Declarations

Authors' contributions

Collection of data (NL, XC, YT), preparation of the manuscript (NL, XC), and supervision (YT, CZ). All the authors read and approved the final manuscript.

Acknowledgements

Not applicable.

Funding

No funding was received by any of the authors for writing this manuscript.

Availability of data and materials

All data supporting these findings are contained within this manuscript.

Ethics approval and consent to participate

This study complied with the tenets of the Declaration of Helsinki. IRB approval was exempted because this was a single case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Competing interests

The authors declare no competing interests.

Author details

[1]Department of Ophthalmology, Peking University Third Hospital, Beijing, China.

[2]Beijing Key Laboratory of Restoration of Damaged Ocular Nerve, Peking University Third Hospital, Beijing, China

References

1. Mierzwinski J, Kosowska J, Tyra J, et al. Different clinical presentation and management of temporal bone fibrous dysplasia in children. World J Surg Oncol. 2018;16(1):5.
2. Lee JS, FitzGibbon EJ, Chen YR, et al. Clinical guidelines for the management of craniofacial fibrous dysplasia. Orphanet J Rare Dis. 2012;7(Suppl 1):2.
3. MacDonald-Jankowski D. Fibrous dysplasia: a systematic review. Dentomaxillofac Radiol. 2009;38(4):196–215.
4. Tabibkhooei A, Taheri M, Rohani S, Azimi A. Polyostotic craniofacial fibrous dysplasia with bizarre radiologic finding: Mandible, anterior skull base, frontal, temporal, parietal, and occipital bones involvement. Surg Neurol Int. 2017;8:160.
5. Galie M, Carnevali G, Elia G, et al. Craniomaxillofacial Fibrous Dysplasia: Conservative Treatment and Maxillary Osteotomy Using the Schuchardt-Kufner Technique. Craniomaxillofac Trauma Reconstr. 2018;11(1):54–8.
6. DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. J Bone Joint Surg Am. 2005;87(8):1848–64.

7. Valentini V, Cassoni A, Marianetti TM, et al. Craniomaxillofacial fibrous dysplasia: conservative treatment or radical surgery? A retrospective study on 68 patients. Plast Reconstr Surg. 2009;123(2):653–60.
8. Kimitsuki T, Komune S. Asymptomatic fibrous dysplasia of the temporal bone. J Laryngol Otol. 2015;129(Suppl 2):42–5.
9. Brown EW, Megerian CA, McKenna MJ, Weber A. Fibrous dysplasia of the temporal bone: imaging findings. AJR Am J Roentgenol. 1995;164(3):679–82.
10. Lisle DA, Monsour PA, Maskiell CD. Imaging of craniofacial fibrous dysplasia. J Med Imaging Radiat Oncol. 2008;52(4):325–32.
11. Elwy R, Gokden M, Cai R. Emergency Optic Canal Decompression for Vision Salvage in Fibrous Dysplasia. World Neurosurg. 2018;109:134–7.

Figures

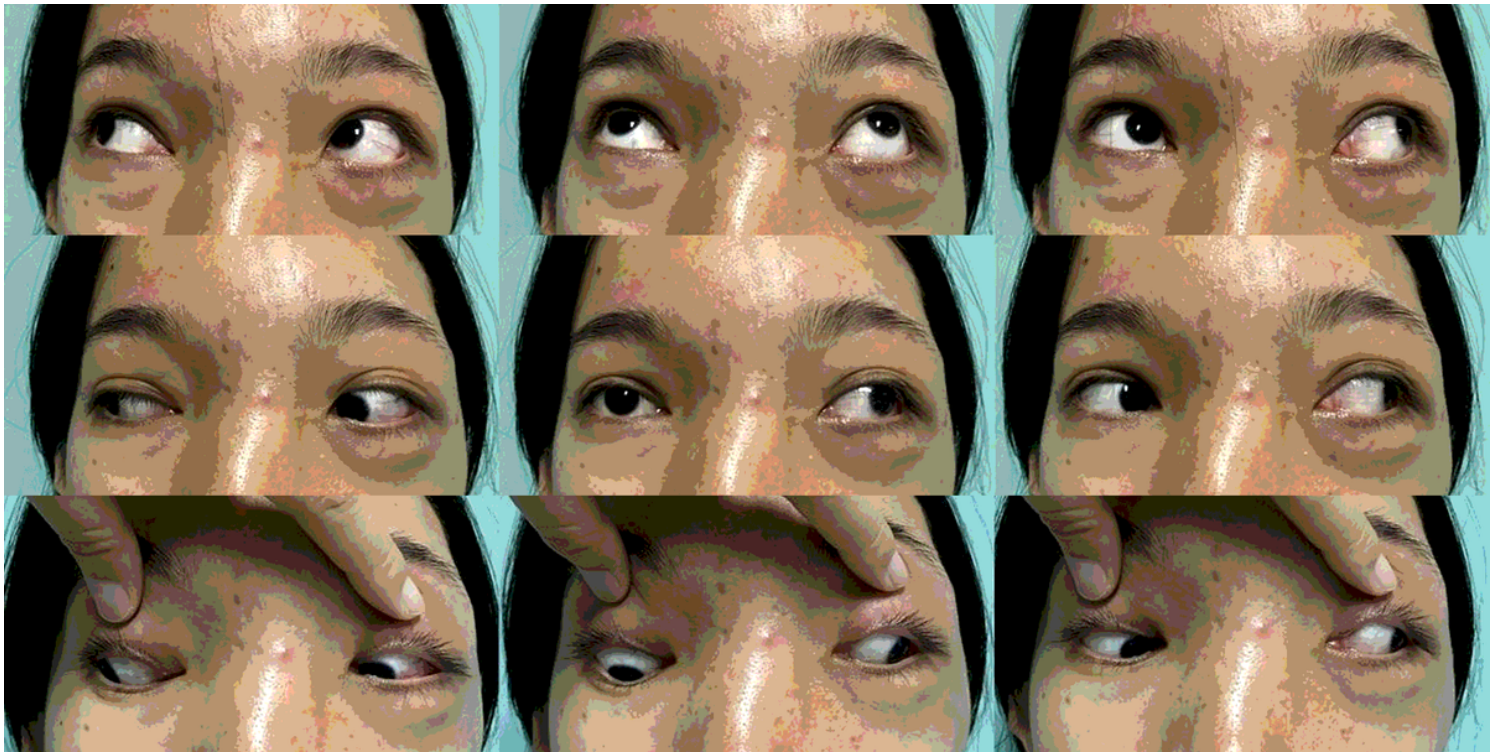

Figure 1

Preoperative documentation

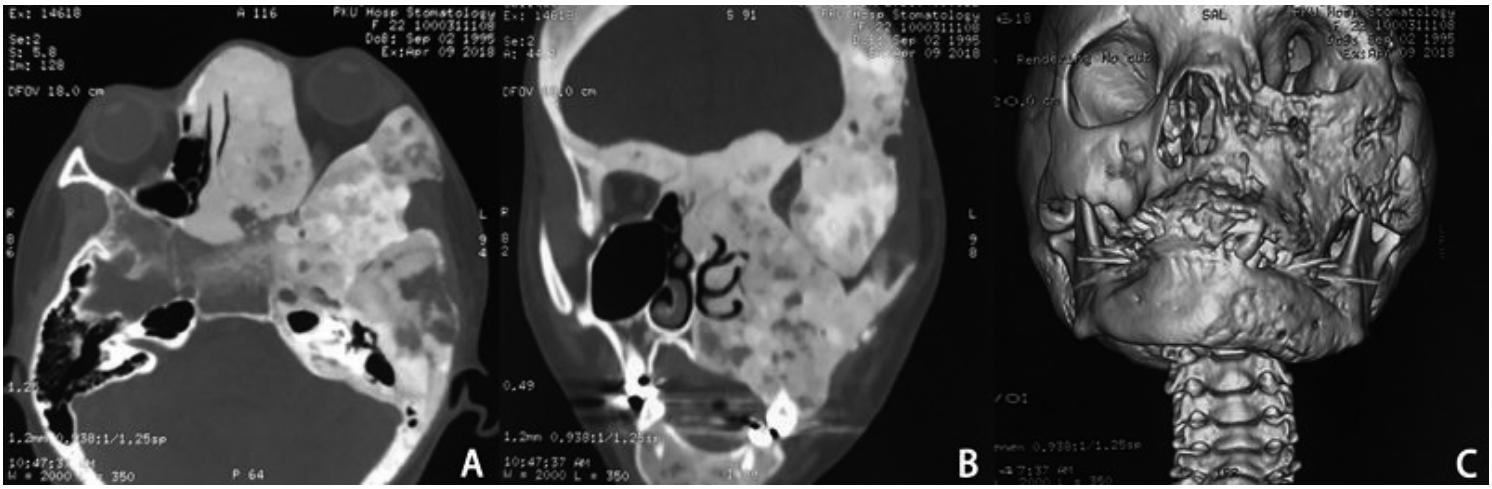

Figure 2

CT images demonstrated the pathognomonic findings A: appearance of FD, a homogenous, “ground-glass” lesion. Images revealed a left predominant expansion of the ethmoid and the temporal bone. B: The entire left maxillary sinus, ethmoid sinus, and bilateral frontal sinus cavities had been obliterated. C: The reconstructed CT image gave a sense of the three-dimensional shape of the lesion and showed the presence of a little stenosis of the left optic canal compared with the right.

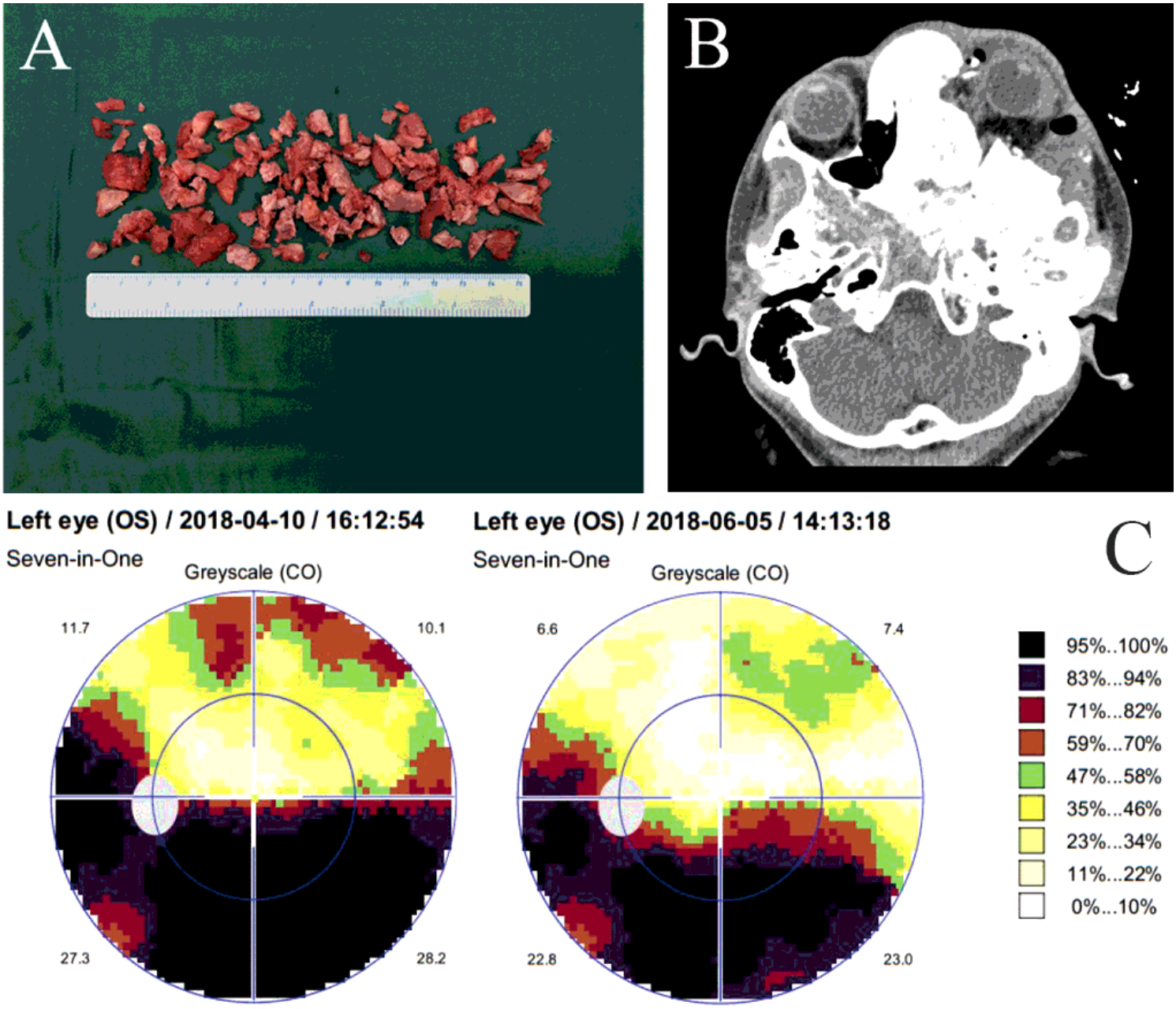

Figure 3

Surgery details. A: All the bones removed during surgery. B: The CT image after the operation showed the removal of bones, as well as correction of the left eye exophthalmos. C. The central visual field area enlarged obviously after surgery. Pre-operation visual field was presented on the left (mean sensitivity=10.1dB, mean defect=19.2dB). Post-operation visual field was on the right (mean sensitivity=14.5dB, mean defect=14.8dB).

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- CAREchecklistEnglish2013.docx