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"Well Digging" Subcraniotomy Strategy with Navigation for Optic Nerve Decompression in Frontoorbital Fibrous Dysplasia: Preliminary Experience

Yunhe Lu, MD, PhD
Junyi Yang, MMed
Yingzhi Wu, MD
Sida Pan, MD
Jiawen Lu
Xiongzheng Mu, MD, PhD

Background: During the past decades, surgical intervention has been the primary treatment modality for frontoorbital fibrous dysplasia involving optic nerve. However, controversy has surrounded the role of optic nerve decompression in a number of ways. Herein, we describe 3 patients with frontoorbital fibrous dysplasia involving optic nerve, who underwent a "well digging" subcraniotomy strategy with navigation for intraorbital unit optic nerve decompression.
Methods: From 2013 to 2015, 3 patients with frontoorbital fibrous dysplasia were investigated in a retrospective manner. They underwent unilateral intraorbital optic nerve decompression with the help of "well digging" strategy and navigation. The key procedures comprise preoperative software simulation, frontoorbital subcraniotomy (like digging a well), expanding cone-shaped surgical field, intraorbital unit optic nerve decompression with navigation, correcting frontal-orbital dystopias, and deformities.
Results: Both at the immediate postoperative period and during the 3–12 months follow-up, 2 cases showed improvement of visual acuity in the affected eye and 1 case showed no deterioration. Other ocular examinations including eye movement were stable. Subsequent reconstruction yielded a satisfactory cosmetic result. No postoperative complications happened.
Conclusions: In our philosophy, surgical management should be tailored to each patient, which is based on the most possible potential etiology. We consider that the intraorbital optic nerve decompression may be more feasible and safer with the help of "well digging" strategy and navigation, especially for those with exophthalmos, orbital volume decreasing, and nonacute visual loss. *(Plast Reconstr Surg Glob Open 2016;4:e1080; doi: 10.1097/GOX.0000000000001080; Published online 8 November 2016.)*

Fibrous dysplasia (FD) is a "benign" overgrowth of bone, first reported by Von Recklinghausen in 1891,[1] which is a congenital skeletal disorder characterized by thinning of the cortex and replacement of the marrow with fibrous tissue. FD has been reported to account for 2.5% to 7.0% of all benign bone tumors, with an equal predilection for both sexes.[2] Craniofacial bones are involved in approximately 20% of FD.[3] Clinically, the signs and symptoms of craniofacial FD vary depending on its type and location.

Among craniofacial FD, fronto-orbital FD may originate within bone adjacent to the optic canal, grow gradually, and compress the optic nerve leading to the most potentially devastating complications. During the past decades, surgical intervention has been the primary treatment modality for frontoorbital FD involving optic nerve.[4] How-

From the Division of Plastic and Reconstructive Surgery, Huashan Hospital, Fudan University, Shanghai, People's Republic of China. Drs. Lu and Yang contributed equally to this work.

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ever, controversy has surrounded the role of optic nerve decompression in a number of ways: some authors support prophylactic optic nerve unroofing,[5,6] whereas others think that unroofing should only be used when visual symptoms have developed,[7] although the timing of therapeutic decompression is not standardized. Some think that optic canal stenosis is the most common cause of visual loss,[8,9] whereas others begin to have doubts about whether surgery has any positive effect on disease progression and whether it is necessary to be overly aggressive in decompression. Despite the aforementioned controversy, there are 2 noteworthy issues. One is that it is the intracanal unit which is focused on and controversial in literature regarding the optic nerve decompression.[10] The other is that there is a concern that prophylactic or therapeutic decompression may lead to unnecessary injury to the optic nerve due to direct iatrogenic trauma or loss of blood supply.[11]

Herein, we describe 3 patients with fronto-orbital FD involving optic nerve who underwent a "well digging" subcraniotomy strategy with navigation for intraorbital unit optic nerve decompression. Knowledge and experience gleaned from the treatment of these patients over the past 3 years has allowed us to develop and refine appropriate treatment paradigms in tandem with our evolving understanding of this disease.

PATIENTS AND METHODS

From 2013 to 2015, 3 patients with fronto-orbital FD were investigated in a retrospective manner. The attention was on those with radiologic or clinical optic nerve involvement; meanwhile, ophthalmology and neurosurgery services were co-consulted. Fronto-orbital osteotomy and reconstruction were simulated for each patient using "mirror technique" by Mimics 15.0 (Materialise Co., Leuven, Belgium). Subsequently, intraorbital unit optic nerve decompression was performed during surgical correction of orbital dystopias and craniofacial deformities. We noted the preoperative and postoperative vision as assessed by the same ophthalmologist, both at the immediate postoperative period and during the 3 to 12 months follow-up. The progression of the disease was also noted.

Surgical Protocol

Navigational Procedure

The routine 3-dimensional (3D) navigational craniofacial CT data set was acquired with a 64-multislice CT (General Electric Medical Systems). Section thickness was 5 mm. In all cases, the Stealthstation Treon neuronavigator (Medtronic Sofamor Danek Co., Minneapolis, Minn.) was used. The 3D craniofacial CT data set was transferred into the workstation through a magnetic optical disk or CD by using the Digital Imaging and Communications in Medicine protocol. Registration of CT Image and patient was achieved with StealthMerge software (Medtronic Sofamor Danek Co.) for intraoperative surgical guidance purpose. A combined strategy for registration was adopted, more specifically, by means of anatomical point mapping with surface mapping. The routine anatomical point included nasal tip, glabellum, and lateral canthus. After registration, the real-time position of the probe tip in the surgical space was displayed on the workstation's monitor with the corresponding location in the image space. Consequently, the real-time anatomical relationship between the FD lesion and the optic nerve was assessed intraoperatively whenever needed.

"Well Digging" Strategy for Intraorbital Unit Optic Nerve Decompression

A coronal modified wave-shaped incision was outlined 2–3 cm behind the hairline. A subgaleal scalp flap was lifted anteroinferiorly to expose the superior orbital rim with the underlying pericranium intact. The pericranial dissection was extended into the orbit. The supraorbital neurovascular bundle was released by osteotomizing the inferior portion of the supraorbital foramen. The periorbital tissue was released from its superolateral extension. To allow access to the deep recesses of the orbital cone, the affected side of the supraorbital rim was first removed temporarily. Then according to preoperative software simulation, fronto-orbital subcraniotomy was performed similarly to digging a well with the aim of expanding surgical field (Fig. 1). Subsequently, the optic foramen and neurovascular bundle was localized, in the vicinity of which decompression was performed to remove diseased bone involving these regions within orbit. Finally, orbital dystopias and frontal deformities were corrected by means of contouring, osteotomy, and reconstruction.

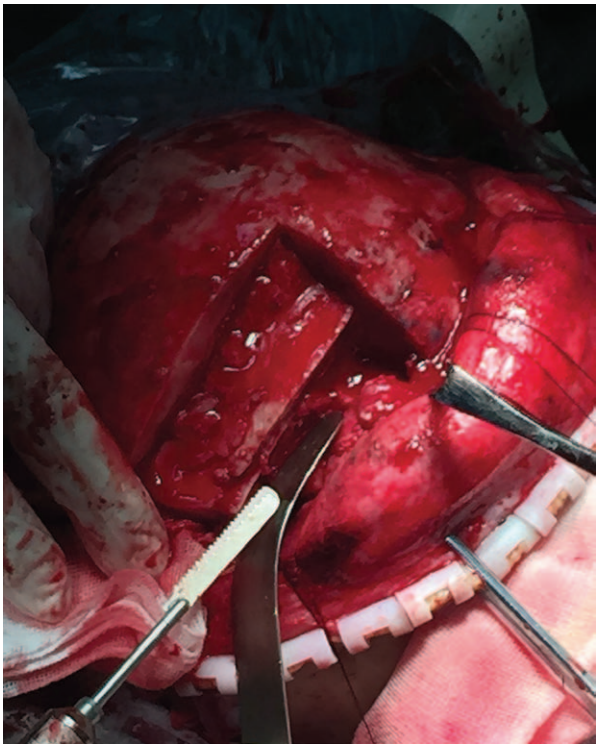

Fig. 1. "Well digging" strategy followed by intraorbital unit optic nerve decompression.

RESULTS

Case 1

A 15-year-old girl presented with an asymmetrical face and right visual blurring. Visual acuity was counted with finger at a distance of 1 m in the right side and 0.2 m in the left side. She also had visual field defect in the right side. The funduscopy confirmed papilloedema of right optic nerve (Fig. 2A). Right exophthalmos measured 3 mm by Hertel exophthalmometry. CT showed FD of the right anterior cranial base, ethmoid bone, sphenoid bone, superolateral orbit, and surrounding the optic canal. Ipsilateral orbital volume was also decreased. The patient underwent resection of the FD of the right frontoorbital bones with ipsilateral intraorbital optic nerve decompression and split cranial bone-graft reconstruction. After therapeutic decompression, her visual acuity in the affected eye improved markedly to 0.2 (Fig. 2B). Postoperative CT during 3 to 12 months showed an enlarged intraorbital volume without optic foramen involvement.

Case 2

A 16-year-old boy complained of progressive protrusion of right forehead and supraorbital ridge with mild dystopia in the right eye. His visual acuity was 0.6 in the right eye and 1.0 in the left eye. He had neither visual field defect nor atrophy of optic nerve. Right exophthalmos measured 3 mm by Hertel exophthalmometry. CT examination exhibited the ground-glass appearance of FD lesion involvement in the superolateral orbit, frontal, and sphenoid bones. Optic canal was surrounded and ipsilateral orbital volume was decreased. Right intraorbital optic nerve decompression was done and subsequent orbital reconstruction yielded a satisfactory cosmetic result. His postoperative CT, ocular examination, and orbital positioning were stable at 3 to 12 months of follow-up.

Case 3

A 20-year-old man was admitted to our hospital with severe left orbital dystopia, exophthalmos, and visual impairment. Visual acuity was 0.2 in left side and 0.8 in the right side. Ophthalmologic examination was normal with no evidence of visual field loss and optic nerve compromise. Four millimeters of proptosis was documented by Hertel exophthalmometry. His CT scans revealed extensive frontoorbital FD surrounding and narrowing the left optic canal (Figs. 3A, B). Ipsilateral orbital volume was also decreased. Left intraorbital optic nerve decompression was done, which was followed by frontoorbital contouring and reconstruction (Figs. 3C, D). Postoperatively, his visual acuity in the affected eye improved slightly to 0.3. His appearance, especially the orbital dystopia, improved markedly during 3 to 12 months of follow-up. CT scans and ocular examination also remained stable.

DISCUSSION

Of all the sequelae of FD, visual impairment is the most feared and potentially debilitating. Visual loss also represents the most common neurological complication of FD affecting the skull.[1,11] It is noteworthy that the optic nerve comprises 5 major units: chiasmatic, intracranial, intracanal, intraorbital, and intraocular parts.[12] It is the intracanal unit optic nerve decompression that has received considerable attention. However, both intracanal prophylactic decompression and therapeutic decompression have been thought to play a limited role.[8] This is because pathoetiological basis of visual impairment is still controversial. Consequently, indications for optimal treatment paradigms remain unclear.

In our series, all the cases had exophthalmos to different degrees and preoperative CT scans revealed 2 common characteristics. First, by careful scrutiny, there was still visible space between the optic neurovascular bundle and the canal wall although the latter was surrounded by FD lesion. Second, ipsilateral orbital volume was also decreased significantly. Three patients underwent unilateral intraorbital optic nerve decompression. During 3 to 12 months of follow-up, 2 cases showed improvement of visual acuity in the affected eye and 1 case showed no deterioration.

Although many think that the etiology of visual impairment may be multifactorial, we do consider that elucidation of the most possible underlying causes has important implications for the planning of therapeutic paradigms. Two main theories have emerged and endeavored to preserve or improve visual function.[6,13] The first theory attempts to prevent the occurrence of visual dysfunction through prophylactic intracranial decompression of the optic canal. The second aims to stabilize or restore visual function in patients with acute or progressive visual loss. In our hands, we chose neither of them although it

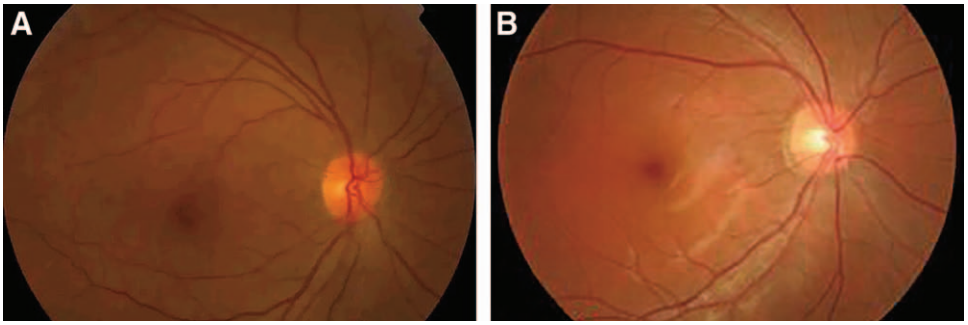

Fig. 2. A, Preoperative examination of retina (case 1). B, Postoperative examination of retina (case 1).

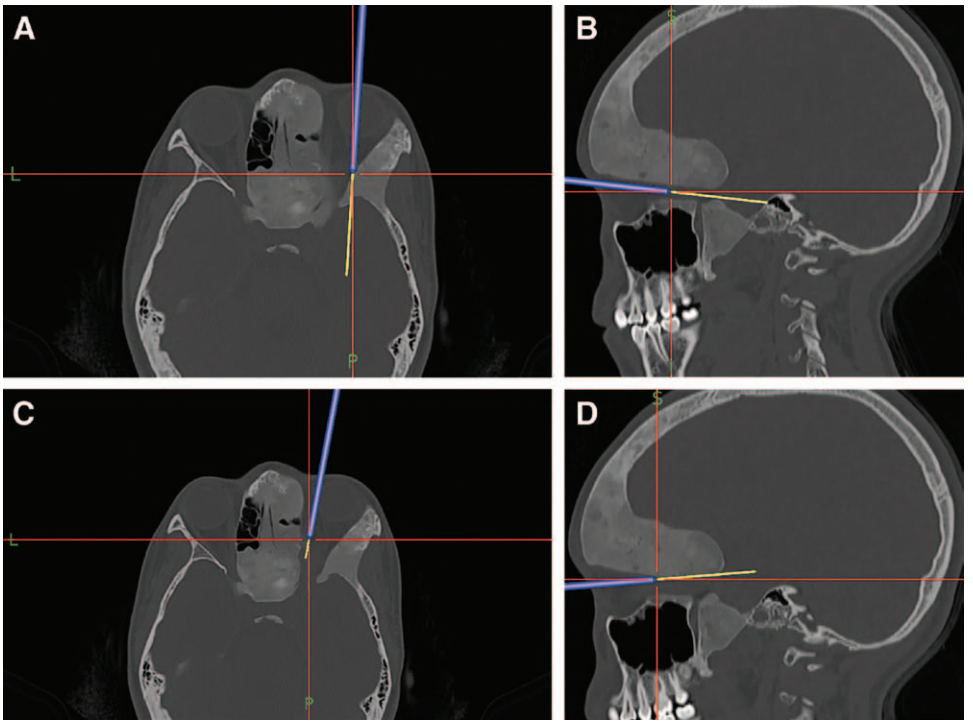

Fig. 3. A, Predecompression of left intraorbital optic nerve (axial, case 3). B, Predecompression of left intraorbital optic nerve (sagittal, case 3). C, Postdecompression of left intraorbital optic nerve (axial, case 3). D, Postdecompression of left intraorbital optic nerve (sagittal, case 3).

seemed we had indications for both. For the prophylactic philosophy, it has been assumed that FD is a progressive disease. There is a concern that progressive compression of the optic nerve will lead to blindness ultimately. Therefore, some authors believe early unroofing will prevent such disastrous consequences.[8,14,15] Our team and others have questioned whether the most common cause of visual loss in FD is indeed bone overgrowth-induced optic canal stenosis. Chen et al[8] demonstrated that prophylactic decompression of clinically or radiographically confirmed optic canal stenosis preserved vision in 67% of patients in whom visual symptoms were present. In a more extensive review, Lee et al[7] showed through radiographic analysis in a cross-section of patients that there was no correlation between optic canal diameter and the development of visual compromise. For the therapeutic philosophy, it certainly has a questionable value in cases of established blindness.[12] Although there has been evidence for restoration of useful vision after early intervention in cases of rapid loss of vision,[12,14] it has been found that visual loss persisting over a 1-month period was not improved by decompression of the optical canal.[11,16] Moreover, we believe both theories are associated with visual loss as a complication in and of itself.

Our philosophy for visual impairment in fronto-orbital FD is that treatment paradigms should be tailored to the individual depending on several circumstances. In a word, treatment should be targeted against appropriate pathological substrates and not on coincident and unrelated phenomena. According to the characteristics of our patients, the most possible underlying causes here may be traction and ischemia of the optic nerve. As the orbital volume gradually decreases by FD involvement, rising intraorbital pressure would cause an increase in intraluminal pressure of the retinal veins, which in turn leads to a drop in perfusion pressure by means of the capillary bed of the retinal artery, with eventual cessation of retinal perfusion. In addition, exophthalmos usually indicates traction, which makes the optic nerve more susceptible. Furthermore, the risks of intracanal optic nerve decompression are not trivial, which may have more probability of unnecessary optic nerve injury due to direct trauma, burring, thermal damage, traction, loss of blood supply, and vascular thrombosis.[12,17] Finally, we consider that the intraorbital optic nerve decompression may be more feasible and safer with the help of preoperative simulation, "well digging" strategy, and navigation. Preoperative software simulation could help us deal with the important decompression site more precisely. During the course of our procedure, fronto-orbital subcraniotomy had the advantage of fewer traumas, which was performed similarly to digging a well to safely expand the surgical field step by step. We also applied navigation to optic nerve decompression, thereby localizing the optic neurovascular bundle in real-time and avoiding iatrogenic injury. Last but not the least, fronto-orbital reconstruction is easier to achieve by our strategy.

Our preliminary results seem favorable. However, shortcomings are obvious because we have fewer patients and no results of long-term follow-up. We will increase sample size and update the data in future work. If possible, with the approval of ethics committee, we will attempt

to compare the follow-up results between intracanal and intraorbital optic nerve decompression in patients of fronto-orbital FD without preoperative visual impairment.

CONCLUSIONS

Visual loss is the most feared complication of frontoorbital FD involving the orbit. Although traditionally attributed to optic canal stenosis, other causes such as ischemia, lesion degeneration, and globe displacement-induced optic nerve traction seem to be more common. In our practice, surgical management is tailored to the individual patient and is founded on intimate clinical, radiological, and pathoanatomical data. We consider that the intraorbital optic nerve decompression may be more feasible and safer with the help of "well digging" strategy and navigation, especially for those with exophthalmos, orbital volume decreasing, and nonacute visual loss.

Xiongzheng Mu, MD, PhD
Division of Plastic and Reconstructive Surgery
Huashan Hospital
Fudan University
No.12 Wulumuqi(M) Road
200040 Shanghai
People's Republic of China
E-mail: cranio@vip.163.com

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