“Apartment” Decompression for Saving Optic Nerve in Fronto-Orbital Fibrous Dysplasia: Strategy and Advantage

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Abstract

Background: During the past decades, surgical intervention is the primary treatment modality for fronto-orbital fibrous dysplasia involving optic nerve. However, controversy has surrounded the role of intra-canal decompression in a number of ways.

Objective: Our philosophy of saving optic nerve is that treatment paradigms should be tailored to the individual. Herein, we describe three patients with fronto-orbital fibrous dysplasia involving optic nerve who underwent an “Apartment” sub-craniotomy strategy with navigation for intra-orbital unit optic nerve decompression.

Methods: From 2013 to 2015, three patients with fronto-orbital fibrous dysplasia were investigated in a retrospective fashion. They underwent unilateral intra-orbital optic nerve decompression with the help of “Apartment” strategy and navigation. The key procedures comprise preoperative simulation, fronto-orbital sub-craniotomy (like entering apartment), expanding cone-shaped surgical field, intra-orbital unit optic nerve decompression with navigation, correcting fronto-orbital dystopias and deformities.

Results: Both at the immediate postoperative period and the 3-12months follow-up, two cases showed improvement of visual acuity in the affected eye and one case showed no deterioration. Other ocular exams including eye movement were stable. Subsequent reconstruction yielded a satisfactory cosmetic result. No postoperative complications happened.

Conclusion: In our philosophy, surgical management should be tailored to each patient, which is based on the most possible potential etiology. We consider the intra-orbital optic nerve decompression may be more feasible and safer with the help of “Apartment” strategy and navigation, especially for those with exophthalmos, orbital volume decreasing, and non-acute visual loss.

Keywords: “Apartment” sub-craniotomy; Fronto-orbital fibrous dysplasia; Intra-orbital unit; Navigation; Optic nerve decompression

Introduction

During the past decades, surgical intervention is the primary treatment modality for fronto-orbital Fibrous dysplasia (FD) involving optic nerve [1]. However, controversy has surrounded the role of optic nerve decompression: prophylactic unroofing is supported by some [2], while others think unroofing should only be used when visual symptoms have developed [3]. Some think that optic canal stenosis is the most common cause of visual loss [4], whereas others have doubts about whether surgery has any positive effect on disease progression. Despite above controversy, there are two noteworthy issues. One is that it is the intra-canal unit which is focused on and controversial in literatures regarding the optic nerve decompression [5]. The other is that there is concern intra-canonical decompression may lead to optic nerve injury due to iatrogenic cause or loss of blood supply.

Here in, we consider the intra-orbital optic nerve decompression may be more feasible and safer with the help of “Apartment” strategy and navigation, especially for those with exophthalmos, orbital volume decreasing, and non-acute visual loss.

Methods

Statement

The experimental protocol and informed consent were approved by the Institutional Review Board of Huashan Hospital, and that all subjects gave informed consent.
Patients

From 2013 to 2015, three patients with fronto-orbital FD were investigated in a retrospective fashion. We focused our attention on those with radiologic or clinical optic nerve involvement, meanwhile ophthalmology and neurosurgery services were co-consulted. Fronto-orbital osteotomy and reconstruction was simulated for each patient using “Mirror Technique” by Mimics 15.0 (Materialise Co.). Subsequently, intra-orbital 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 3-12 months follow-up. The progression of the disease was also noted.

Surgical protocol

Navigation: The Stealthstation Treon neuro-navigator (Medtronic Sofamor Danek Co., Minneapolis, MN) was used. The data set was transferred into the workstation by using the Digital Imaging and Communications in Medicine protocol (DICOM). A combined strategy for registration was adopted, more specifically, by means of anatomical point with surface mapping. After that, the real-time position of the probe in the surgical space was displayed on the workstation with the corresponding location in the image. Consequently, the real-time anatomy between the lesion and the optic nerve was re-assessed whenever we needed intraoperatively.

“Apartment” strategy: A coronal wave-shaped incision was outlined 2 cm behind the hairline. A subgaleal scalp flap was lifted anteroinferiorly to expose the superior orbital rim with underlying pericranium intact. The pericranial dissection was extended into the orbit. The supraorbital neurovascular bundle was released. The periorbital tissue was released from its superolateral extension. In order to access to the deep recesses of the orbital cone, the affected side of the supraorbital rim was first removed temporarily. Then according to preoperative simulation, fronto-orbital subcraniotomy was performed like “entering apartment” with the aim of expanding surgical field (Figure 1). Subsequently, the optic foramen and neurovascular bundle was localized, in the vicinity of which decompression was performed within orbit. Finally, orbital dystopias and frontal deformities were corrected by means of contouring, osteotomy and reconstruction according to “Mirror Technique” (Figure 2).

Results

Case presentation 1

A 15-year-old girl presented with an asymmetrical face and right visual blurring. Visual acuity was count finger at the distance of 1 meter in right side and 0.2 in left side, respectively. She also had visual field defect in right side. The funduscope confirmed atrophy of right optic nerve. 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 also decreased. The patient underwent resection of the FD of the right fronto-orbital 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. Postoperative CT at 3-12 months showed an enlarged intraorbital volume without optic foramen involvement.

Case presentation 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 also 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 3-12 months follow-up.

Case presentation 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 right side, respectively. 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 fronto-orbital FD surrounding and narrowing the left optic canal. Ipsilateral orbital volume also decreased. Left intraorbital optic nerve decompression was done, which was followed by fronto-orbital contouring and reconstruction. Postoperatively, his visual acuity in the affected eye improved slightly to 0.3. His appearance, especially
the orbital dystopia, improved markedly at 3-12 months follow-up. CT scans and ocular examination also remained stable.

**Discussion**

It is noteworthy the optic nerve comprises five major units: chiasmatic, intracranial, intra-canal, intra-orbital, and intraocular part [6]. It is the intra-canal unit decompression that has received considerable attention. However, it has been thought to play a limited role [5,7]. This is because patho-etiological basis of visual impairment is controversial. Consequently, indications for optimal treatment paradigms remain unclear.

For the prophylactic philosophy, it has been assumed FD is a progressive disease, which will lead to blindness ultimately. Therefore, early unroofing may prevent such disastrous consequences. We and others have questioned whether the most common cause of visual loss is indeed bone overgrowth-induced optic canal stenosis [8]. For the therapeutic philosophy, it certainly has a questionable value in cases of established blindness [9,10]. Moreover, both may be associated with visual loss as a complication in and of itself.

Our philosophy is treatment paradigms should be tailored to the individual. According to characteristics of our practice (exophthalmos and orbital volume decreasing), the most possible underlying causes may be traction and ischemia of optic nerve. As orbital volume gradually decreases by FD involvement, rising intra-orbital pressure would cause an increase in intra-luminal pressure of the retinal veins, which in turn leads to a drop in perfusion of the retinal artery, with eventual cessation of retinal perfusion. Additionally, exophthalmos usually indicates traction, which makes the nerve more susceptible. Furthermore, the risks of intra-canal decompression are not trivial, which may have more probability of injury due to direct trauma, burring, thermal damage, traction, loss of blood supply, and vascular thrombosis.

**Advantage**

Finally, we consider the intra-orbital decompression may be more feasible and safer with the help of preoperative simulation, “Apartment” strategy and navigation. Preoperative simulation could help us dealing with important decompression site more precisely. Fronto-orbital sub-craniotomy has the advantage of less injury, which was performed like “entering apartment” in order to safely expand surgical field step by step. We also apply navigation, thereby we could localize the optic neurovascular bundle in real-time and avoid iatrogenic injury. Last but not least, fronto-orbital reconstruction could be easier to achieve by this strategy.

**Limitation**

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. And if possible, with the approval of ethic committee, we attempt to compare the follow-up results between intra-canal and intra-orbital optic nerve decompression in patients of fronto-orbital FD without preoperative visual impairment.

**Conclusion**

Visual loss is the most feared complication. Besides canal stenosis, other causes such as ischemia, lesion degeneration and optic nerve traction appear more common. We consider the intraorbital decompression may be more feasible and safer with the help of “Apartment” strategy, especially for those with exophthalmos, orbital volume decreasing, and non-acute visual loss.

**References**