Optic Nerve Decompression for Orbitofrontal Fibrous Dysplasia

**Takumi Abe, M.D., Ph.D.,¹ Kaneshige Sato, M.D., Ph.D.,²
Takaharu Otsuka, M.D., Ph.D.,² Noriyoshi Kawamura, M.D., Ph.D.,¹
Motohiko Shimazu, M.D., Ph.D.,¹ Hitoshi Izumiyama, M.D., Ph.D.,¹ and
Kiyoshi Matsumoto, M.D., Ph.D.¹**

ABSTRACT

Orbitofrontal fibrous dysplasia often involves the bony orbit and the optic canal. Although fibrous dysplasia reportedly produces compression of the optic nerve leading to visual disturbances, optic nerve decompression in patients without clinical signs of optic neuropathy is still controversial. We describe two patients with orbitofrontal fibrous dysplasia without signs of visual disturbance and one patient with McCune-Albright syndrome and progressive visual impairment. Optic nerve decompression was performed prophylactically for two patients and therapeutically for one patient through the transcranial extradural route. Dystopias and craniofacial deformities induced by fibrous dysplasia also were corrected. The micropressure suction-irrigation system was especially effective for decreasing heat transfer and thereby preventing thermal injury of the optic nerve. The orbitofrontal area was reconstructed from cranial bone, iliac bone, and ribs. Postoperative follow-up revealed no disturbances in visual function and no evidence of cerebrospinal fluid leakage. These findings suggest that optic nerve decompression may be effective in preventing visual disturbances with minimal risk of other neurological sequelae. Subsequent orbital reconstruction yielded satisfactory cosmetic results.

KEYWORDS: Fibrous dysplasia, optic nerve decompression

Skull Base, volume 12, number 3, 2002. Address for correspondence and reprint requests: Takumi Abe, M.D., Department of Neurosurgery, Showa University School of Medicine, 5-8 Hatanodai 1, Shinagawa-ku, Tokyo 142-8666, Japan. E-mail: takumi@med.showa-u.ac.jp. Departments of ¹Neurosurgery and ²Plastic Surgery, Showa University School of Medicine, Tokyo, Japan. Copyright © 2002 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662. 1531-5010,p;2002,12,03,145,154,ftx,en;sbs00288x.

Fibrous dysplasia manifests clinically as bone masses that are abnormal in both structure and growth. Orbitofrontal fibrous dysplasia may originate within bone adjacent to the optic canal, grow gradually, and compress the optic nerve, leading to visual disturbances.¹⁻⁶ Visual impairment, once noted, tends to be progressive^{1-3,5} despite the common belief that fibrous dysplasia tends to cease after puberty.^{4,6-8} Malignant or sarcomatous degeneration reportedly occurs in 0.5% of the cases.⁹ Optic nerve decompression in patients without clinical signs of optic neuropathy is still controversial. The controversy focuses on whether surgical relief should be instituted before moderate to severe loss of vision occurs or delayed until visual loss must be followed by emergent decompression. We describe two patients who underwent early optic nerve decompression before vision was lost and one patient with McCune-Albright syndrome who underwent optic nerve decompression for progressive visual impairment.

SURGICAL TECHNIQUE

Surgical approaches to the optic canal must adequately expose the deep orbital apex and optic nerve to allow ablation and decompression. First, lumbar drainage is performed to prevent injury when the brain is later retracted. The patient is placed in a supine position with the head moderately flexed.

A coronal incision is outlined 3 to 5 cm behind the hairline. A subgaleal scalp flap is elevated with the underlying pericranium intact. The pericranial dissection is extended into the orbit. The periorbit is released from its superolateral extension. The supraorbital neurovascular bundle is released by osteotomizing the inferior portion of the supraorbital foramen. A frontoparietal craniotomy is performed 1.5 to 2.0 cm above the superior orbital rim. One side of the supraorbital rim is removed along with a frontoparietal cranial bone to allow access to

the deep recesses of the orbital cone and to remove affected bone involving these regions.

Next, the dura of the frontal brain lobes is retracted gently to expose the superior posterior orbital cone and optic canal completely (i.e., an extradural approach). Because the orbital roof is affected and thickened by fibrous dysplasia, the superior orbital roof can be drilled carefully with a diamond burr under a microscope. Drilling directly over the optic canal is avoided. Vibratory and thermal energy can be transferred through the bone, injuring the optic nerve. Continuous irrigation and suction with a micropressure suction-irrigation system¹⁰ at the site of drilling is mandatory to decrease heat transfer and to prevent thermal injury to the optic nerve. Once the affected bone has been thinned at the posterior orbital cone, a sphenoid punch is used to remove the optic canal roof in pieces. Extreme care is needed to avoid compression of the optic nerve. The entire roof of the optic canal is removed to decompress the nerve completely. The orbital roof can be reconstructed from split cranial bone or iliac bone with microplate or stainless wires to ensure its structural integrity. The supraorbital bar can be reconstructed from calvarial bone or ribs and secured with microplates.

ILLUSTRATIVE CASES

Patient 1

An 18-year-old man with persistent right exophthalmos was readmitted to our hospital. When he was 13 years old, his protruding orbitofrontal bone and zygoma were chiseled out to correct an asymmetrical face with a protuberant right periorbital region and frontal bossing. When he was 18 years old, his computed tomographic (CT) scans showed extensive orbitofrontal fibrous dysplasia surrounding and narrowing the right optic canal (Figs. 1A and 1B). His visual acuity was 1.2 bilaterally. De-

Figure 1 Patient 1. An 18-year-old man with right orbitofrontal fibrous dysplasia underwent extensive resection, prophylactic decompression of the right optic nerve, and bone-graft reconstruction. (A) Preoperative axial CT and (B) three-dimensional CT demonstrating tight compression of the right optic nerve by fibrous dysplasia. (C) Postoperative axial CT and (D) three-dimensional CT revealing extensive resection of fibrous dysplasia and decompression of the right optic nerve but the posterior aspect.

spite his normal vision, this patient was soon likely to experience symptoms of optic nerve compression. We therefore resected the fibrous dysplasia extensively, prophylactically decompressed the right optic nerve, and reconstructed with iliac bone and two ribs. We slightly resected the posterior aspect of the optic canal along the optic nerve. The patient's postoperative clinical course was uneventful. Follow-up CT showed an enlarged optic canal and the posterior aspect of the optic nerve (Figs. 1C and 1D). Subsequent orbital reconstruction yielded a satisfactory cosmetic result. His visual acuity and visual field remained unchanged 1 year after surgery (Fig. 1E).

Patient 2

A 16-year-old girl was referred to our hospital with a 4-year history of gradual enlargement of the left orbitofrontal region and left proptosis. Her visual acuity was 0.03, correctable to 1.0 with lenses bilaterally. CT showed fibrous dysplasia of the left anterior cranial base, ethmoid bone, superior orbit, and optic canal (Fig. 2A). Fibrous dysplasia extended medially into the ethmoids, completely obliterating the sinus. The patient underwent bifrontal craniotomy with resection of the left frontal bone and the superior, medial, and lateral orbits. Prophylactically, the left optic foramen was decom-

Figure 1 (Continued) (E) Visual field illustration of the patient's right eye 1 year after the decompressive surgery.

pressed superiorly. The left frontal orbital region was reconstructed with split cranial bone grafts. Postoperative CT scans showed an enlarged optic canal (Fig. 2B). The patient's postoperative follow-up revealed no evidence of cerebrospinal fluid leakage. Early in the postoperative period, the patient had a transient nasal hemianopsia that resolved spontaneously. At her 21-month follow-up examination, her ocular examination, or orbital positioning, was stable (Fig. 2C).

Patient 3

A 52-year-old woman was admitted to our hospital with an asymmetric face, left exophthalmos, and left visual impairment. At 22 years old, she was found to have fibrous dysplasia of the left orbitofrontal region (McCune-Albright syndrome).

She underwent several surgeries for resection and bone grafting of fibrous dysplasia of her left zygoma and mandibular bone. On her most recent admission, her visual acuity was 1.2 in the right eye and 0.5 in the left eye. Left exophthalmos measured 3 mm by Hertel exophthalmometry. CT showed fibrous dysplasia of the left frontal, temporal, ethmoid, maxilla, and sphenoid bones, and substantial involvement of the left optic canal (Fig. 3A). The patient underwent resection of the fibrous dysplasia of the left frontal and sphenoid bones with left optic canal decompression and split cranial bone-graft reconstruction. After therapeutic decompression, her visual acuity in the affected eye improved markedly from 0.5 to 1.5. Postoperative CT showed an enlarged optic canal (Fig. 3B). Her ophthalmologic examination remained normal at her 46-month follow-up examination (Fig. 3C).

Figure 2 Patient 2. A 16-year-old girl with left orbitofrontal fibrous dysplasia underwent extensive resection, prophylactic decompression of the left optic nerve, and bone-graft reconstruction. (A) Preoperative axial CT revealing stenosis of the left optic canal by fibrous dysplasia. (B) Postoperative axial CT 1 year after surgery, showing extensive resection of fibrous dysplasia and the enlarged left optic canal. (C) Visual field illustration of the patient's left eye 21 months after the decompressive surgery.

DISCUSSION

The need for optic nerve decompression in craniofacial fibrous dysplasia has been addressed as both a prophylactic and a therapeutic modality.^{1,4,11} Two

studies strongly advocate prophylactic intracranial decompression of the optic nerve for treating fibrous dysplasia.^{1,11} Chen and colleagues recommend prophylactic decompression in two circumstances.¹ First, radiological involvement of the

Figure 3 Patient 3. A 52-year-old woman with left orbitofrontal fibrous dysplasia underwent resection of the left frontal and sphenoid bones with left optic canal decompression and split cranial bone-graft reconstruction. (A) Preoperative axial CT revealing stenosis of the left optic canal by fibrous dysplasia. (B) Postoperative axial CT 3 years after surgery, showing extensive resection of fibrous dysplasia and the enlarged left optic canal. After optic nerve decompression, her visual acuity in the affected eye improved markedly. (C) Visual field illustration of the patient's left eye 46 months after the decompressive surgery.

optic canal indicates a reasonably high incidence of visual loss in the affected eye and a very high incidence of some visual disturbance can be expected.^{1,6} Second, the speed with which visual deterioration can occur and the brief amount of time before it

becomes permanent indicate the need for prophylactic decompression.

Acute visual loss has been reported in association with mucocoeles,^{2,4,5,12} hemorrhage,^{5,13} and hemorrhagic cysts,⁵ as well as with fibrous dysplasia alone

when the optic canal is involved.^{1,3,4,14-16} Chronic, gradual visual decline also is associated with craniofacial fibrous dysplasia,^{1,4} although this phenomenon is commonly thought to reflect optic nerve compression within a stenotic foramen.^{1,4,5,11,17}

A review of published cases of visual loss from fibrous dysplasia since the advent of CT or magnetic resonance imaging (MRI)¹⁶ indicated that visual loss associated with fibrous dysplasia is more likely related to mass lesions than to progressive encroachment of the optic canal. High-resolution and three-dimensional (3-D) CT enable precise evaluation of the anatomy of the optic canal and the cause of visual loss in cases of fibrous dysplasia. Our patients were evaluated by high-resolution CT, 3-D CT, and MRI.

The visual acuity of several patients has improved after optic nerve decompression.^{1,18,19} The visual acuity of our third patient improved markedly in the affected eye after optic nerve decompression. In patients with acute visual loss caused by fibrous dysplasia, early decompression of the optic nerve produces encouraging results. Postoperatively, some patients have regained normal vision.^{1,14} Decompression delayed by 1 week is less likely to improve vision.² Acute visual deterioration from fibrous dysplasia has been controlled short-term by steroids,^{12,14,17} but long-term improvement has not been maintained. Thus, patients with gradually deteriorating vision who are not yet totally blind and patients with a recent sudden onset of visual deterioration related to fibrous dysplasia may benefit from optic nerve decompression. The efficacy of prophylactic decompression of the optic nerve is difficult to assess because, by definition, patients have no preoperative visual loss. Because predicting the arrest of pericanal fibrous dysplasia is difficult, the growth of fibrous dysplastic bone should be assumed to continue and optic nerve compression should be assumed to be relatively imminent.¹⁸ Prophylactic decompression of the optic nerve is likely to be of benefit on the theoretical grounds that once vision has been impaired, the chances of being able to restore vision are greatly reduced.

Extensive resection of craniofacial fibrous dysplasia and subsequent reconstruction with autogenous bone graft can be accomplished safely with minimal rates of operative morbidity. Patients with craniofacial fibrous dysplasia involving the optic canal can safely undergo excision of the fibrous dysplasia and decompression of the optic nerve. Although several approaches have been reported for optic canal decompression, the unilateral frontotemporal extradural approach is superior for many reasons. This approach allows extensive resection of the dysplastic frontal, ethmoidal, and sphenoidal bones, including the orbital roof, lateral orbital wall, sphenoid ridge, anterior clinoid process, and sphenoidal plane without leakage of cerebrospinal fluid or damage to the brain. Lumbar drainage is especially effective for preventing brain injury during retraction, particularly when the deep orbital apex is exposed. Decompression should be performed by an experienced neurosurgeon to minimize the risk of iatrogenic change to the optic nerve. The use of a diamond burr that transmits vibratory or thermal injury to the optic nerve resulting in edema should be avoided.^{1,10} Because the orbital roof is affected and thickened by fibrous dysplasia, careful drilling of the superior orbital roof with a diamond burr under a microscope, while avoiding drilling directly over the optic canal, is effective. During drilling, continuous irrigation and suction with a micropressure suction-irrigation system¹⁰ at the site of drilling are mandatory to decrease heat transfer to prevent thermal injury to the optic nerve. The irrigation system, which can be manipulated with one hand, can clean the operating field effectively and decrease heat transfer with its rapid flow. Once the diseased bone has been thinned at the posterior orbital cone, a thin sphenoid punch should be used to remove the optic canal roof in pieces. Extensive resection of the fibrous dysplasia may treat the problems of visual disturbance more directly than optic canal decompression alone.

Recurrent fibrous dysplasia after optic nerve decompression is not widely reported.^{1,20} Our patients had no ophthalmic complications during the

short-term follow-up. The extent of decompression of the optic canal is likely to be a factor in recurrence because any residual bone is likely to contain fibrous dysplasia.¹ Thus, as more walls are decompressed, the likelihood decreases that continued growth of residual fibrous dysplasia will entrap the optic nerve. The extensive decompression required and the risk associated with intracranial facial surgery may necessitate the transcranial extradural approach.

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Commentary

This small series of patients with orbito-frontal fibrous dysplasia who underwent optic nerve decompression describes the well-respected senior surgeon's operative techniques and his argument for prophylactic optic nerve decompression in asymptomatic patients. The authors treated three patients, two of whom were asymptomatic with respect to visual symptoms. The authors illustrate their anatomical results with computed tomography scans.

The authors used an extradural transcranial approach. They rightly emphasize the use of lumbar drainage and continuous irrigation and suction with a micropressure suction-irrigation system previously described by Lüdecke and Triage.¹ The authors state that "because predicting the arrest of pericanal fibrous dysplasia is difficult, the growth of fibrous dysplastic bone should be assumed to continue and optic nerve compression should be assumed to be relatively imminent." Unfortunately, this assumption is not based on natural history data, and the opposite assumption could just as