J Neurosurg 92:350–354, 2000

Visual loss associated with fibrous dysplasia of the anterior skull base

Case report and review of the literature

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The authors present a case of visual loss associated with fibrous dysplasia of the anterior skull base and the surgical management of this case. Preoperative computerized tomography scanning in this patient demonstrated a patent optic foramen and a rapidly growing cystic mass within the orbit, which was responsible for the patient's visual loss. A literature review revealed that this case is typical, in that cystic mass lesions of various types are frequently responsible for visual loss associated with fibrous dysplasia. The authors did not find significant evidence in the literature to support the notion that visual loss associated with fibrous dysplasia is the result of progressive optic canal stenosis, thus raising questions about the value of prophylactic optic canal decompression. Instead, as demonstrated by this case and those uncovered in the literature review, most instances of visual loss result from the rapid growth of mass lesions of cystic fibrous dysplasia, mucoceles, or hemorrhage. Findings of the literature review and the present case of fibrous dysplasia of the anterior skull base support a role for extensive surgical resection in these cases and indicate a need for additional prospective analysis of a larger number of patients with this disease.

KEY WORDS • fibrous dysplasia • visual loss • skull base

FIBROUS dysplasia is a bone disease of unknown origin that can occur in monostotic or polyostotic form. In either form the disease may involve the skull, with a preference for the frontal, ethmoid, sphenoid, and maxillary bones. The presenting symptoms are most commonly facial deformities, proptosis, or headaches.[16] Malignant or sarcomatous degeneration can occur in fibrous dysplasia and has been reported to occur in 0.5% of cases.[7] Visual disturbance occurs in a significant number of cases and, along with hearing impairment, is the most common neurological deficit caused by fibrous dysplasia.[16] Surgery is regarded as the treatment of choice for preventing or arresting visual loss in cases of fibrous dysplasia. Specifically, surgery to decompress the optic nerve within the optic canal has been recommended, both for the treatment of visual loss once it has occurred and prophylactically to prevent visual loss.[4,5,7,14,15] Our case and a review of the literature indicate that the usual cause of visual loss in fibrous dysplasia may not be optic canal stenosis.

Case Report

History. This 15-year-old girl first presented to us with a diagnosis of craniofacial fibrous dysplasia at 9 years of age after a 3-year history of progressive left nasal obstruction. At that time, findings on CT scanning were reported to be compatible with fibrous dysplasia involving the left supraorbital region and left orbit circumferentially, and also involving the nasoethmoid complex and left nasomaxillary region, causing an almost complete block of the left nasal passage. When she was examined at 9 years of age, she was reported to have left proptosis and left frontal bossing with no evidence of neurological deficit. She underwent partial resection of the lesion and a reconstructive procedure at another institution when she was 10 years old. Subsequent operations included a left-sided dacrocystorhinostomy when she was 11 years old and another procedure when she was 12 years old for left nasal obstruction and removal of reconstructive plates placed at the first operation. At the time of her tear duct surgery, the patient's visual acuity was noted to be 20/20 in both eyes. At the age of 13 years, the girl began to notice progressively decreased vision in her right eye. Three months before the surgery reported here, she had a best-corrected visual acuity of 20/70 in the right eye and 20/15 in the left eye. On Jaeger's test types, she read at a level of 12 points for near vision in the right eye and a level of 1 point in the left eye. She correctly identified six of eight Ishiara color plates by using the right eye and eight of eight by using the left. In each eye the pupil measured 7 mm with brisk light reaction; on the right side she exhibited a trace of afferent pupillary defect. At a base of 96 mm, the patient's Hertel exophthalmometry measurements were 16 mm in the right eye and 17 mm in the left eye, and there was left-sided hypoglobus. A motility examination revealed a mild under-

Abbreviations used in this paper: CT = computerized tomography; MR = magnetic resonance.

action of elevation in the left eye, but the patient's vision was orthotropic for both near and far distances. The results of the slit-lamp and intraocular pressure examinations were normal. Ophthalmoscopy revealed a normal macula, vessels, and peripheral retina in each eye. The cup-to-disk ratio was 0.1 in each eye, and there was no disk edema or optic atrophy. Goldmann visual field testing revealed a dense central scotoma extending outward to the superior periphery in the right eye and an intact field in the left eye. By the time the patient presented for surgery, she had experienced an additional, subjective decrease in visual acuity in her already severely compromised right eye. During preoperative bedside testing, the patient's vision was assessed at the level of finger counting in the right eye. Additional, formal ophthalmological examination was not performed before surgery.

Preoperative CT scans of the brain and orbits demonstrated changes in bone that were consistent with fibrous dysplasia involving the sphenoid bone bilaterally, with greater involvement on the left side (Fig. 1). The lesion involved the posterior ethmoid bones and ethmoid sinuses bilaterally. Also, a large expansive, cystic soft-tissue component of fibrous dysplasia located posteriorly along the medial aspect of the right orbit appeared to compress the right optic nerve inferiorly and medially (Fig. 2). Although the optic canal was involved, the optic foramen appeared patent on the right and left sides (Fig. 2). A surgical resection was planned with several goals in mind: preservation of vision in the left eye, correction of the cosmetic deformity of the left orbit, and resection of the rapidly growing mass on the right side accompanied by histological examination to rule out sarcomatous degeneration.

Operation. In March 1997, a bifrontal craniotomy with extradural exposure of the anterior skull base was performed as previously described.[2] The anterior frontal floor and orbital roof were resected using a high-speed drill, curettes, and punches. This dissection was continued posteriorly and medially to the cribriform plate on the right side, and the ethmoid sinus was entered on the right side. The resection extended into the area of exuberant soft-tissue growth in the right medial orbit. This mass was removed to the level of the periorbitum on the right side and sent to the laboratory for frozen-section examination, which revealed benign fibrous dysplasia. The left frontal resection was continued until the left orbital roof and medial orbital wall were removed, exposing the periorbitum and leaving the orbital floor intact. This resection was extended posteriorly several millimeters beyond the posterior ethmoidal artery. In addition, remnants of the sphenoid sinus were opened on the right side, and the block of dysplastic bone composing the sphenoid sinus on the left side was resected. This dissection was continued inferiorly to include the anterior face of the sphenoid region and all ethmoid sinuses down through the perpendicular plate to the nasal septum. Next, a left-sided vertical orbital translocation was performed through an inferior orbitotomy by using autogenous bone grafts harvested from a location posterior to the craniotomy flap. Simultaneously, reconstruction of the left supraorbital rim was performed, followed by remodeling of the cranial bone flap. Closure was performed using a previously harvested pedicled, pericranial flap covering the frontal floor and sutured posteriorly to holes drilled in the anterior cut edge of the planesphenoid.[2]

FIG. 1. Preoperative coronal CT scan revealing fibrous dysplasia of the sphenoid and ethmoid bones and a cystic lesion of the right ethmoid bone.

Postoperative Course. The patient tolerated the surgical procedure well and her vision was stable postoperatively. Her postoperative hospital course was unremarkable. The girl did not experience any subjective change in vision. She exhibited marked cosmetic improvement and experienced no complications of surgery. Postoperative visual field testing revealed a central scotoma in the right eye, as noted preoperatively, but there was improvement in the right temporal visual field. The visual field in the left eye remained intact. On formal ophthalmological examination performed approximately 4 months postoperatively, the patient's vision was 20/20 in the left eye and 4/200E in the right eye, which was consistent with the subjective visual decline she observed prior to surgery. Initially the right optic disk appeared normal. The clinical features were consistent with those of a retrobulbar optic neuropathy. At sequential postoperative visits, mild optic atrophy was observed to develop in the patient. At the last follow-up examination conducted 17 months postoperatively, the patient reported no subjective change in vision compared with her preoperative vision. Visual acuity was stable at 20/20 in the left eye and 3/200E in the right, and her visual fields were stable as well.

Discussion

Fibrous dysplasia of the anterior skull base often results in proptosis and hypoglobus, as demonstrated in this case. Although visual compromise is less common,[3] both acute and chronic visual loss are well documented. Acute visual loss has been reported in association with mucoceles,[7,10,12,18,19] hemorrhage,[10,11,17] and hemorrhagic cysts,[5,10] as well as with fibrous dysplasia alone when it involves the region of the anterior skull base and optic canal.[1,3–5,7,9,15] Chronic, gradual visual decline also occurs with craniofacial fi-

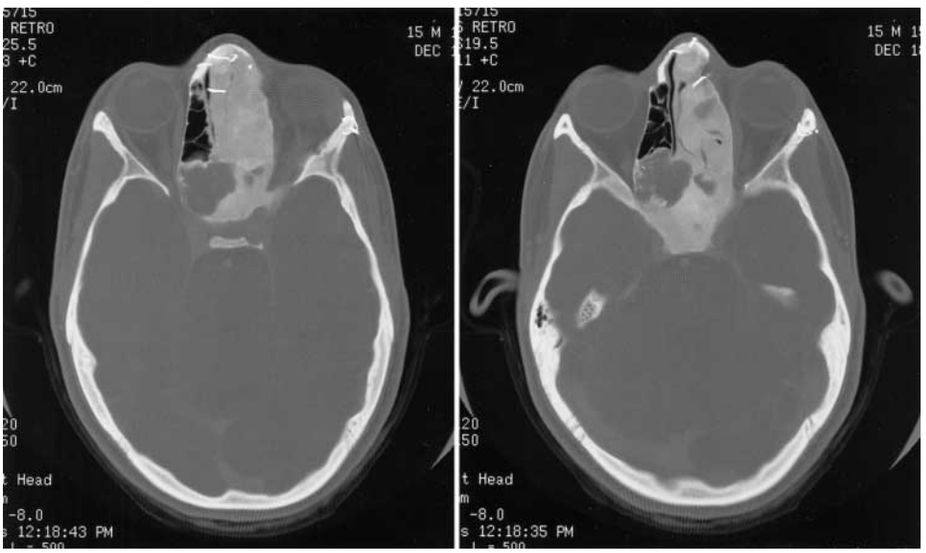

FIG. 2. Preoperative axial CT scans demonstrating a cystic lesion of the right ethmoid bone and patency of the optic foramen bilaterally.

brous dysplasia.[4,5,7,15] Several mechanisms have been proposed to cause for visual loss in patients with fibrous dysplasia. Most commonly, this phenomenon is thought to occur secondary to compression of the optic nerve within a stenotic optic foramen.[4,5,7,10,13,14] Another mechanism that has been suggested is displacement or distortion of the globe with traction on the optic nerve.[3,5,13] External compression by a cystic lesion, separate from stenosis of the optic canal, has also been recognized.[5,10] Finally, rare vascular events have also been reported to result in blindness.[5,11]

Reports of two series have recently been published in which prophylactic intracranial decompression of the optic canal is strongly advocated for treating fibrous dysplasia.[4,14] The recommendation that prophylactic decompression of the optic canal would benefit the patient by preventing visual loss is based on the theory that optic canal stenosis is most often the mechanism of such a loss. Several authors cite radiographs and tomograms obtained in cases treated before the era of CT scanning, which demonstrate fibrous dysplasia involving the bones of the optic canal, as evidence that this stenosis is the cause of chronic visual loss.[6,8,16] The diameter of the optic canal may appear narrowed, as evidenced by these radiographs and tomograms, whenever the sphenoid and ethmoid bones are involved by fibrous dysplasia. However, these studies may be misleading. Two points should be considered. First, before the common use of CT and MR imaging, plain x-ray films would have failed to identify the hemorrhagic lesions, cystic expansion, and mucoceles associated with fibrous dysplasia, which often cause visual loss. Second, the relationship of canal stenosis to visual loss is not well established. Although we recognize that fibrous dysplasia may involve the bones of the optic canal, producing optic canal stenosis, the relationship of this stenosis to visual loss has not been proven. Currently, high-resolution CT scanning allows more precise evaluation of the anatomy of the optic canal and the cause of visual loss in cases of fibrous dysplasia.

In the present case, 6 years after the patient underwent partial resection of the fibrous dysplasia involving the anterior skull base, she began to experience visual compromise and by the time she presented to our institution, she had lost functional vision in her right eye. The CT scans obtained in this patient did not reveal a compromised optic foramen, but instead demonstrated optic nerve compression within the right orbit, resulting from a mass of exuberant, cystic-appearing fibrous dysplasia that compressed the optic nerve from the medial side. This distinction in the mechanism by which fibrous dysplasia causes visual loss has important implications in the surgical treatment of craniofacial fibrous dysplasia. The present case shows a striking juxtaposition, in which a patient with a patent optic foramen has a cystic area of fibrous dysplasia that compresses the optic nerve within the orbit, causing visual loss on the right side. This patient's right orbit had no cosmetic or structural deformities except for a focal area of cystic fibrous dysplasia in the ethmoid region, which was enough to compress the optic nerve from the medial side and cause her visual loss. This case and others in the literature support the view that optic canal stenosis may not be the only element contributing to visual loss due to fibrous dysplasia. In addition, if canal stenosis is not responsible in such cases of visual loss, prophylactic canal decompression may be unwarranted.

To examine the actual cause of visual loss (mass lesion as opposed to stenosis) and the possible benefit of prophylactic optic canal decompression, we reviewed cases of visual loss from fibrous dysplasia reported in the literature during the period in which CT or MR imaging has been available.[1,3–5,7,9–11,13,15,17–19] All known cases of fibrous dysplasia accompanied by visual loss in which the results of CT or MR imaging were reported were reviewed. The review included 13 individual papers that were either case reports[1,3,9,11,13,15,17–19] or accounts of operative series.[4,5,7,10] From these 13 papers, 20 cases contained sufficient CT or MR imaging data for review. The review included four papers describing operative series,[4,5,7,10] in which only those

TABLE 1

*Summary of cases reported in the literature in which fibrous dysplasia was associated with visual loss**

Authors & Year	Patient Age (yrs), Sex	Presentation of Visual Loss	Location of FD†	Findings‡	Prior Op
Sevel, et al., 1984	9, F	acute, unilat	rt superior & lat orbit, sphenoid wing	hemorrhage	none
Edgerton, et al., 1985					
Case 3	18, F	acute, unilat	rt frontal, ethmoid, parietal, orbital	mucocele	none
Case 4	15, F	chronic, unilat	lt orbit, anterior cranial fossa, & maxilla	cystic FD	6 attempted resections
Derome, et al., 1986					
Case 48	21, F	acute, bilat	bilat sphenoid, clivus, lesser sphenoid wings	hemorrhagic cyst	none
Case 34	13, F	acute, unilat	bilat frontal floor, sphenoid, lesser sphenoid wings, anterior clinoid	cystic FD	none
Case 32	5, M	chronic, bilat	bilat anterior skull base & medial facial bones	cystic FD	none
Osguthorpe & Gudeman, 1986	21, F	acute, unilat	lt frontal, sphenoid, ethmoid	"mixed," fibroosseous lesion	biopsy
Kurokawa, et al., 1989	11, M	acute, unilat	lt ethmoid, sphenoid	hematoma/hemorrhagic cyst; lt ophthalmic artery occlusion	none
Saito, et al., 1990	4, F	chronic, unilat	lt frontal, ethmoid, sphenoid	"typical" FD only	none
Weisman, et al., 1990	29, F	acute, unilat	rt maxillary, ethmoid, sphenoid	mucocele	orbital floor resection
Arroyo, et al., 1991	22, F	acute, unilat	rt frontal, sphenoid, ethmoid	"typical" FD only	none
Bland, et al., 1992	25, F	acute, unilat	rt frontal, sphenoid, ethmoid	cystic FD	craniotomy, frontoparietal resection
Joseph, 1995	24, F	acute, unilat (rt) & chronic (lt)	bilat ethmoid, sphenoid	cystic FD	craniotomy, bilat OCD
Steel & Potts, 1995	20, F	acute, unilat	rt sphenoid, ethmoid	mucocele	none
Chen, et al., 1997					
Case 1	28, M	chronic, bilat	not specified	hypervascular, aggressive FD (no focal hemorrhage)	radical resection, rt OCD
Case 12	13, F	acute, unilat	rt sphenoid, posterior ethmoid	cystic FD	none
Katz & Nerad, 1998					
Case 4	32, M	acute, unilat	rt orbital roof, sphenoid	aneurysmal bone cyst	none
Case 5	25, F	acute, unilat	rt ethmoid, optic canal	mucocele	several ops for debulking & reconstruction
Case 6	19, F	acute, unilat	rt skull base	aneurysmal bone cyst	none
Case 7	16, F	acute, unilat	bilat orbital roofs, sphenoid	hemorrhage	none

* FD = fibrous dysplasia; OCD = optic canal decompression.
† Sphenoid = related to sphenoid sinus, plane, or body of sphenoid; sphenoid wing involvement is specified as such.
‡ Only cases in which CT or MR imaging were available are included.

cases in which CT or MR imaging data were actually reported or shown were considered. Patient presentation, location and type of lesion, and the type of operation, if any, that had been performed before the patient suffered visual loss are contained in Table 1. Clinical and operative findings included in these reports were also considered in characterizing the lesions because descriptions of CT and MR studies were often limited.

Of the 20 cases reviewed, 16 patients had visual loss that was clearly caused by lesions other than progressive optic canal stenosis.[3–5,7,9–11,17–19] These included six patients who had cystic fibrous dysplasia,[3–5,7,9] four who had mucoceles,[7,10,18,19] four with hemorrhagic lesions,[5,10,11,17] and two described as having "aneurysmal bone cysts."[10] The authors believed that, in cases such as these 16, prophylactic optic canal decompression was not likely to have prevented the development of such mass lesions and subsequent visual loss. Of the remaining four patients described in the literature review in whom there were no discrete mass lesions, two achieved good outcomes in response to treatment following episodes of acute blindness.[1,13] The remaining two patients without mass lesions probably would not have benefited from prophylactic decompression of the optic canal. One had what was described as a very aggressive vascular lesion that initially improved with optic canal decompression, but eventually deteriorated into a poor result.[4] The other patient began to experience visual loss at 3 years of age and, after undergoing optic canal decompression at the age of 4 years, continued to experience blindness on that side.[15] Prophylactic decompression probably would not have been a practical consideration in a 2- or 3-year-old child. Thus, prophylactic optic canal decompression may not have been of significant benefit in any of the 20 reported cases of fibrous dysplasia and visual loss.

Because we do not have sufficient data on this problem to advocate one type of treatment in all cases, patients with fibrous dysplasia should be carefully analyzed individually before recommending surgery. Ideally, surgical treatment would reduce the risk of visual compromise due to mucoceles, hemorrhage, or rapid growth of cystic fibrous dysplasia. Rather than treating these patients with optic canal decompression, resection of areas of rapid or exuberant growth before visual symptoms occur may be appropriate in many patients. Extensive resection does not appear to have been performed in any of the 16 patients suffering visual loss due to cysts, mucoceles, or hemorrhage (Table 1). In 11 of these cases there is no mention of prior surgery.[4,5,7,10,11,17,18] One patient only underwent a cosmetic procedure.[19] Three underwent what were described

as partial resections or debulking procedures.[3,7,10] In the final case,[9] the patient underwent repeated optic canal decompressions, both intra- and extracranial for recurrent episodes of visual loss. In this case, CT scans revealed a mass of cystic fibrous dysplasia in the medial orbital wall, but no mention is made of an operation to resect that area. We propose that an extensive resection including cystic masses of fibrous dysplasia may treat the problem of visual loss more directly than optic canal decompression alone.

In summary, a skull base resection, as performed in the present case, may be appropriate in the management of fibrous dysplasia involving the anterior skull base, orbit, and paranasal sinuses. Although limited in scope, this case and the review of the literature indicate that visual loss associated with fibrous dysplasia is more likely due to mass lesions than progressive encroachment of the optic canal. These mass lesions that cause optic nerve compression can be a result of hemorrhage into fibrous dysplasia, mucocele, or rapid cystic growth of fibrous dysplasia alone. We advocate careful analysis of the anatomy in cases of visual loss, because the cause may be attributable to a primary or secondary mass lesion rather than stenosis of the optic canal. Surgery should be directed accordingly. The causes of visual loss in craniofacial fibrous dysplasia certainly deserve more thorough, prospective analysis because prophylactic optic canal surgery may not be warranted in these cases.

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Manuscript received September 21, 1998.
Accepted in final form October 5, 1999.
This work was supported in part by an unrestricted grant to Dr. Lee from Research to Prevent Blindness, Inc., New York, New York.

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