Emergency Optic Canal Decompression for Vision Salvage in Fibrous Dysplasia

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Key words
- Decompression
- Optic nerve
- Optic nerve canal
- Visual loss

Abbreviations and Acronyms
FD: Fibrous dysplasia
MRI: Magnetic resonance imaging
OD: Right eye
OS: Left eye

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BACKGROUND: **The most common neurologic manifestations of fibrous dysplasia (FD) are vision and hearing loss. Optic decompression for progressive vision loss has been shown to yield positive results in terms of visual outcomes; however, emergency optic decompression surgery for sudden loss of vision in FD has not to date been reported in the pediatric population. We report the first case of FD presenting with sudden vision loss and successfully managed with emergency optic decompression.**

CASE DESCRIPTION: **A 10-year-old male patient known to have FD with a 2-month history of progressive vision loss in his left eye presents with sudden blindness. Emergency decompression surgery restored vision in his left eye to 20/40, which remained stable on long-term follow-up.**

CONCLUSION: **There is no consensus on the benefit and safety of prophylactic optic decompression, yet therapeutic decompression has been shown to prevent vision deterioration. Our findings suggest that therapeutic decompression even when done in the emergency setting yields positive results, while prophylactic decompression carries an inherent risk for loss of vision in a seeing eye.**

BACKGROUND

Fibrous dysplasia (FD) usually presents with facial deformity or as an incidental radiologic finding; however, the most commonly encountered neurologic manifestations of FD are vision and hearing loss.[1] Mucocele, bony dysplasia, and hemorrhagic complications have all been reported as causes of optic nerve compression in FD.[2,3] However, optic nerve compression is not associated with, or necessarily progresses to, optic neuropathy in all cases, but elevated growth hormone levels and development of aneurysmal bone cysts could indicate an increased risk for developing optic neuropathy.[4]

Optic canal decompression has been reported to improve vision in pediatric cases with gradual vision loss.[5,6] There has also been a previous report of a case of acute vision loss in an adult patient with FD in whom decompression surgery together with mucocele drainage restored vision.[7] However, to our knowledge, no reports on urgent decompression surgery for sudden vision loss in a child patient exist in the literature. Also, our patient did not present with a cystic lesion, as is commonly the case.[5]

CASE REPORT

Clinical Presentation

The patient first presented at age 10 with a 2-month history of progressive vision loss in his right eye (OD). On examination he was neurologically intact, his visual acuity was 20/20 on the left (OS) with normal color vision and no visual field deficits, and he had no perception of light OD. His fundus examination showed moderate to severe disc pallor OD and moderate (grade 2) disc edema with obscuration of small vessels and 1–2 large vessels OS. Otherwise, his ophthalmologic examination was unremarkable. His past medical history was significant for asthma and attention deficit hyperactivity disorder. He was initially thought to have Foster Kennedy syndrome, but head computed tomography scan and magnetic resonance imaging (MRI) showed no intracranial mass lesions. Instead, the tests showed bilateral optic and internal auditory canal stenosis with right optic nerve edema and diffuse bony anomaly of skull and facial bones with severe foramen magnum stenosis (Figure 1). Spine MRI showed severe midcervical canal narrowing with cord compression. Skeletal survey findings suggested skeletal dysplasia. Right frontal lesional biopsy was performed for diagnostic purposes and showed some degree of osteosclerosis. Genetic evaluation afterwards suggested skeletal dysplasia, but whole exome sequencing was eventually not diagnostic of FD. Vision remained stable over the course of the following month, when vision OS suddenly decreased to perception of light with fundus examination showing progression to grade 3–4 disc edema, which prompted urgent optic canal decompression to save vision.

Surgery and Follow-Up

Microscopic left optic canal decompression through a left pterional extradural

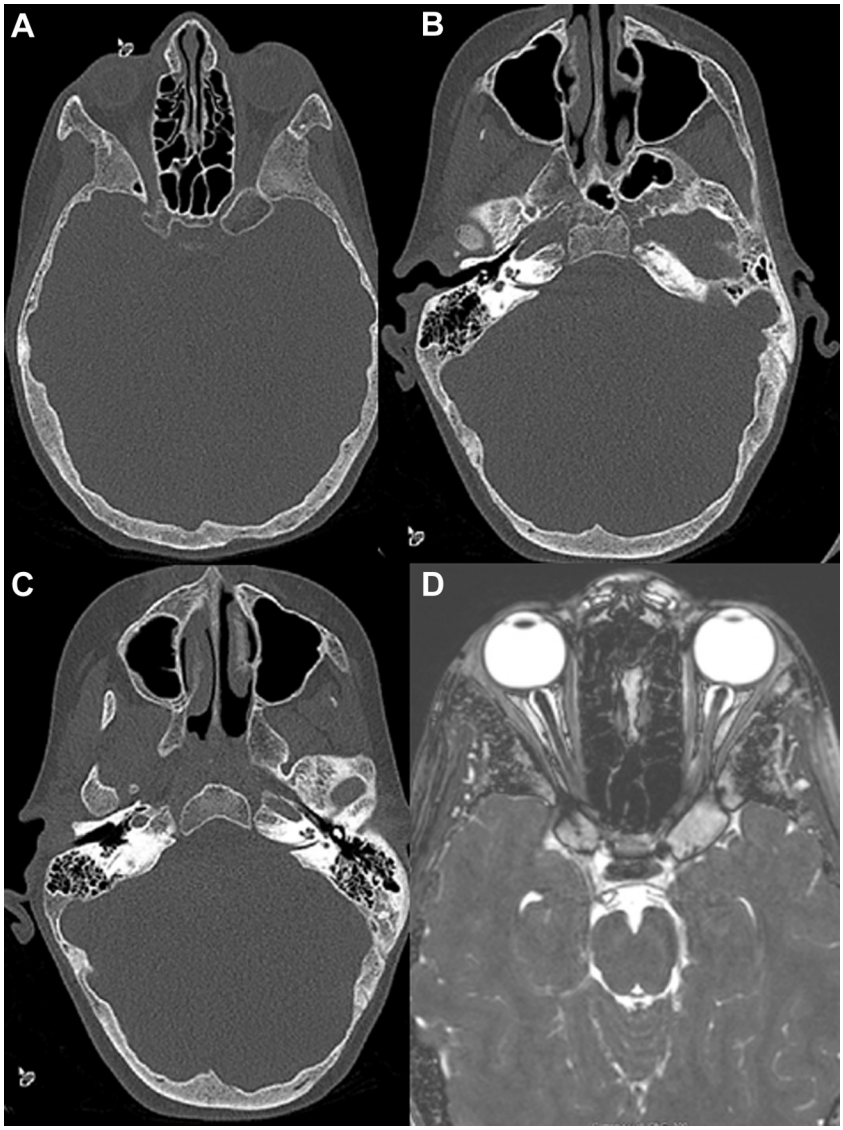

Figure 1. (**A**) Head computed tomography showing diffuse thickening of the calvaria, facial bones, and spinous processes of the upper cervical vertebrae, abnormal vertical orientation of the clivus, foramen magnum stenosis, severe narrowing of the bilateral optic canals with thickening of the anterior clinoid process, and narrowing of the right (**B**) and left (**C**) internal auditory canals (**D**). Brain magnetic resonance imaging T2-weighted imaging bilateral optic canal stenosis shows a smaller edematous right optic nerve compared with the left.

approach using image guidance (StealthStation surgical navigation system; Medtronic Sofamor Danek USA, Inc. Memphis, Tennessee) was done. Overgrowing bone tissue next to the left optic canal demonstrating thin cortical bone and a beehive-appearing center was removed in piecemeal fashion. Further pathologic characterization of the lesion was not possible due to the gross nature of the surgical specimen; however, given radiologic and genetic findings, FD was the most likely diagnosis. Postoperatively the patient's visual acuity OS improved to 20/300 and disc edema to grade 2. Postoperative imaging demonstrated decompressed left optic canal and persistent stenosis and crowding of the left orbital cavity (Figure 2). We extended the decompression into the posterior orbital cavity through the same approach 10 days later as our ophthalmologist suggested (Figure 3). Over the course of the following 4 weeks, the patient underwent further bilateral optic nerve sheath fenestration. Fundus examination showed improving OD and almost resolved OS (grade 0–1) disc edema. Visual acuity OS was 20/40 1 month postoperatively.

Over the following 2 years, the postoperative course was uneventful apart from surgical evacuation of epidural fluid collection secondary to cerebrospinal fluid leak from a dural tear that occurred during the apex of orbital cavity decompression surgery and occasional headaches managed conservatively. Two-year follow-up imaging showed some restenosis of the left optic canal (Figure 4), but vision remained stable at 20/40 OS.

DISCUSSION

Therapeutic decompression has been shown to prevent vision deterioration in approximately 50% of the cases.[8] In a previous report of a pediatric patient undergoing optic canal decompression for deteriorating visual acuity, vision improved postoperatively,[9] as was our case suggesting potential benefit for therapeutic surgical decompression of the optic nerve in reverting vision loss. In our patient, emergency optic decompression successfully reverted acute vision loss from perception of light back to a visual acuity of 20/40 on long-term follow-up, further suggesting that decompression surgery is as effective in reverting acute vision loss as it is in managing more insidious vision deterioration in FD patients.

On the other hand, decompression procedures have their own inherent risks. In a report by Edelstein et al,[10] a patient with 20/20 visual acuity suffered complete loss of vision