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Case Report

Sphenoid Bone Dysplasia: A Rare Cause of Compressive Optic Neuropathy Mimicking Glaucoma

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Abstract

Fibrous dysplasia is a benign, rare bone disease in which bone is replaced by fibro-osseous tissue to varying degrees. It can present differently depending on the amount of compression caused by the fibro-osseous tissue. Patients are usually asymptomatic, but symptoms related to cranial nerve compression may occur. In this case report, we describe a 45-year-old woman with sphenoid bone dysplasia which compressed the optic nerve and caused unilateral optic disc cupping that mimicked glaucoma. Our case highlights the importance of including compressive etiologies associated with optic disc cupping in the differential diagnosis of glaucoma.

Keywords: Compressive optic neuropathy, glaucoma, sphenoid bone dysplasia

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Introduction

Fibrous dysplasia is a benign, rare bone disease of unknown etiology in which dysregulated osteoblast differentiation results in bone being replaced by fibro-osseous tissue to varying degrees.[1] The disease usually affects the craniofacial bones, and the amount of compression caused by the fibro-osseous tissue can influence how it presents.[2] Patients are usually asymptomatic, although headaches or other consequences of cranial nerve compression can occur.[3,4]

In this report, we describe a woman with sphenoid bone dysplasia which compressed the optic nerve and produced optic disc cupping that mimicked glaucoma. Our aim was to demonstrate the importance of including compressive etiologies associated with optic disc cupping in the differential diagnosis of glaucoma.

Case Report

A 45-year-old woman was referred to our clinic for detailed investigation of optic disc pallor in her right eye. She had visited another clinic complaining of pain in and around the right eye. She had no history of systemic or ophthalmologic disease. In the examination at the other center, disc pallor was observed in the right eye and intraocular pressure (IOP) was measured as 24 and 25 mmHg in the right and left eye, respectively. Suspecting glaucoma, the patient was prescribed a topical anti-glaucoma agent (brimonidine tartrate; Alphagan P 0.15% Allergan, Inc., Irvine, CA, USA) and referred to our clinic for further evaluation of the optic disc pallor.

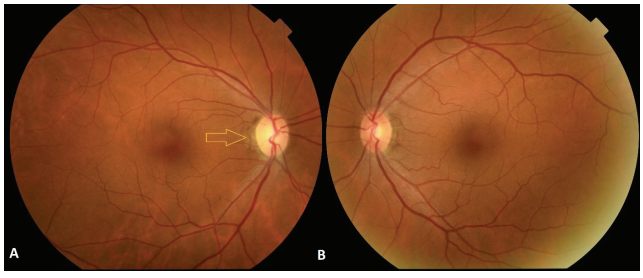

Figure 1. Color fundus photographs of patient's right (A) and left (B) eyes. In the right eye, optic disc pallor (yellow arrow) was observed on the temporal side of the optic disc

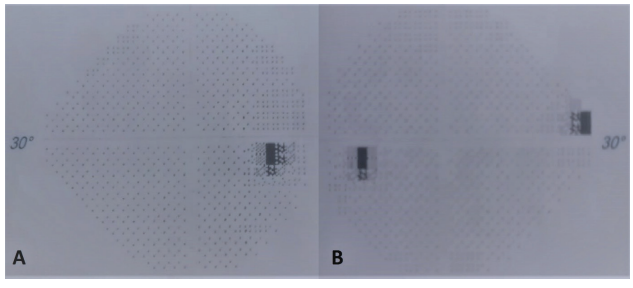

Figure 2. Visual field tests demonstrating normal appearance in the right (A) and left (B) eye

In the comprehensive examination performed in our clinic, the patient's best corrected visual acuity was found to be 10/10 on Snellen chart and her color vision was 15/15 with Ishihara plates in both eyes. Direct and indirect pupil reflexes were normal bilaterally and no relative afferent pupil defect was observed. Slit-lamp biomicroscopy of the anterior segment showed no abnormalities, and IOP measured with Goldmann applanation tonometer was 19 and 18 mmHg in the right and left eye, respectively, with 0.15% brimonidine tartrate. Gonioscopic examination showed open angles (grade 3 to 4) bilaterally. Corneal thickness measured by pachymetry was 580 µm in both eyes. Fundus examination revealed temporal rim pallor of the optic disc in the right eye (Figure 1A) with a cup/disc ratio of 0.4. The optic disc was normal in the left eye (Figure 1B).

In the 30-2 SITA Standard visual field test (Humphrey field analyzer, Carl-Zeiss Meditec, model 745i, Dublin, CA, USA), both right and left eye results were within normal limits (Figure 2). Optic nerve head optical coherence tomography (OCT) (Triton DRI OCT, Topcon, Tokyo, Japan) showed retinal nerve fiber layer (RNFL) thinning in all quadrants, and ganglion cell complex analysis revealed diffuse loss of the ganglion cell layer in the right eye (Figure 3A). Optic disc parameters were normal in the left eye (Figure 3B).

A compressive etiology in the right eye was suspected because of the asymmetric optic disc pathology. Computed tomography (CT) of the paranasal sinuses revealed a sphenoid bone dysplasia (Figure 4A). Magnetic resonance imaging (MRI) of the orbits was requested and the imaging showed that the sphenoid bone dysplasia was compressing the right optic nerve (Figure 4B). After diagnosis, the topical brimonidine therapy was discontinued. Neurosurgical and otolaryngology consultations were requested to plan surgery to address the compression. Informed consent was obtained from the patient.

Discussion

Glaucoma is the leading cause of irreversible vision loss in adults due to the progressive thinning of optic nerve fiber layer.[5,6] Distinguishing glaucoma from glaucoma-mimicking diseases such as compressive optic neuropathies is challenging. The assessment of factors including age (less than 50 years old), complaints such as headache or pain around the eye, visual field defects involving the vertical midline, rapid decline in visual acuity, disproportionate neuroretinal rim pallor relative to disc cupping, asymmetric visual acuity and/or visual field loss, and asymmetric cupping can be useful in the differential diagnosis.[7] These factors may increase the likelihood of detecting an intracranial compressive lesion.

In our case, at the initial examination in another clinic, IOP was measured as 24 and 25 mmHg in the right and left eye, respectively, and an anti-glaucoma drop was prescribed. In addition to this history, in our clinic we observed a 0.4 cup/disc ratio in the right eye. These findings could have contributed to a misdiagnosis of glaucoma. The IOP elevation could be

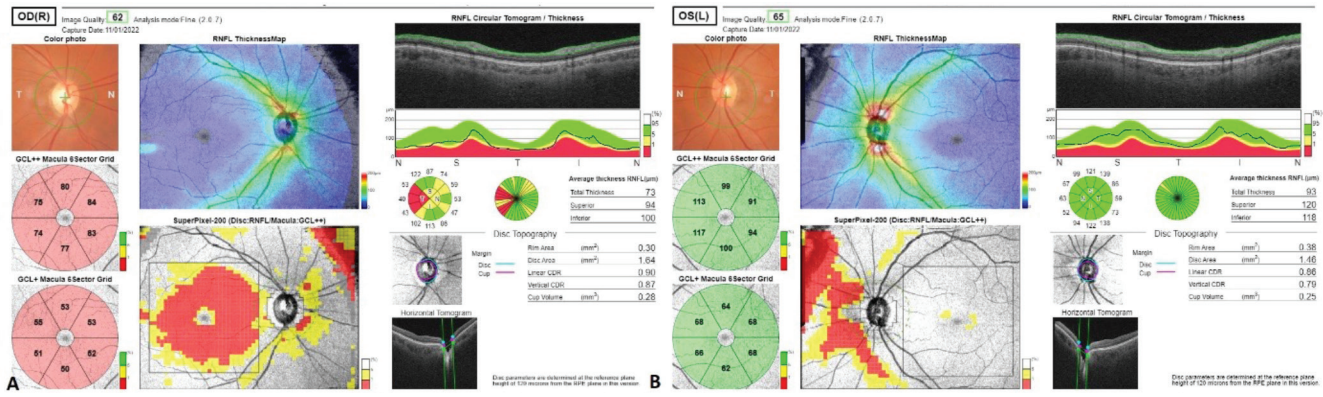

Figure 3. Optical coherence tomography images of both optic discs. The right optic disc (A) showed diffuse ganglion cell layer loss and peripapillary retinal nerve fiber layer thinning in all quadrants. The left optic disc (B) had normal optic disc parameters

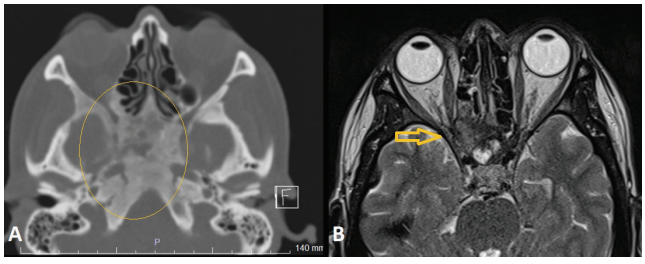

Figure 4. Radiologic images of the patient. Computed tomography of the paranasal sinuses (A) revealed sphenoid bone expansion and ground-glass appearance related to the fibrous dysplasia (yellow circle). On axial T2-weighted magnetic resonance imaging of the orbits (B), the sphenoid bone appeared hypointense and right optic nerve compression caused by obliteration secondary to bony expansion was observed at the level of the optic canal (yellow arrow)

explained by the corneal thickness (580 µm bilaterally) detected by pachymetry. However, the patient's younger age (45 years), pain around the eye, and most importantly, optic disc pallor in the temporal neuroretinal rim were warning signs suggesting that further evaluation for non-glaucomatous etiology would be necessary. As our case shows, optic nerve compression can easily be confused with glaucoma due to the optic disc cupping, with the addition of a thick cornea detected with pachymetry.

In non-glaucomatous optic neuropathy associated with compressive lesions, optic atrophy usually develops, but glaucomatous-type cupping can occur.[6] Bianchi-Marzoli et al.[8] reported that patients with compressive lesions demonstrated an increased cup/disc ratio. In addition, a study by Ahmed et al.[9] showed that 6.5% of patients initially diagnosed as having normal-tension glaucoma had clinically relevant intracranial compressive lesions.

Sphenoid dysplasia is a rare condition with non-specific symptoms, which makes the diagnosis especially challenging. On radiologic evaluation, fibrous dysplasia has characteristic appearances on CT, including ground-glass pattern (50%), a homogeneously dense pattern (25%), and cystic pattern (20%).[10] On MRI, the amount of bone trabeculation can affect signal intensity on T1- and T2-weighted images.[11] However, fibrous dysplasia characteristically shows low signal intensity in addition to well-demarcated borders.

Typical visual field defects in glaucoma include arcuate, paracentral, and biarcuate defects and eventually tunnel vision. These defects are manifestations of ganglion cell and RNFL damage and can therefore be seen in many non-glaucomatous conditions and neuroophthalmological diseases. As examples, arcuate defects can be seen in ischemic optic neuropathy, optic disc coloboma, optic disc drusen, or optic disc pits. Biarcuate defects or tunnel vision can be seen in retinitis pigmentosa, panretinal photocoagulation, and carcinoma-associated retinopathy, while paracentral defects can be seen in hereditary or nutritional/toxic optic neuropathy.[12] In the differential diagnosis of glaucoma-mimicking visual field defects, the rule that glaucoma respects the horizontal raphe, the existence of arcuate defects, and the preservation of a temporal island in advanced cases are usually useful. On the other hand, compressive optic neuropathies respect the vertical meridian. A detailed ophthalmological examination can also reveal serious decline in visual acuity and disproportionate neuroretinal rim pallor relative to the disc cupping.[12]

RNFL measurement with OCT is useful to diagnose and monitor progression in glaucoma. However, many non-glaucomatous optic neuropathies and central nervous system diseases can also cause RNFL thinning that mimics glaucoma. These pathologies include optic neuritis, compressive etiologies of the optic nerve, hereditary or traumatic optic neuropathies, multiple sclerosis, and degenerative diseases such as Alzheimer's and Parkinson's disease.[13]

Bock et al.[14] used OCT to analyze the RNFL in patients with multiple sclerosis and in patients with glaucomatous optic nerve changes and found that the temporal quadrant was more affected in patients with optic neuritis. In another study, Kim et al.[15] evaluated toxic and nutritional optic neuropathy and determined that RNFL thinning occurs as in glaucoma, but that the temporal fibers were affected first. Gupta et al.[16] determined that among patients with similar average RNFL thickness, those with non-glaucomatous optic nerve cupping had lower macular thickness and macular volume compared to those with glaucomatous optic nerve cupping. They also reported that nasal and temporal RNFL thickness were lower in patients with

non-glaucomatous optic nerve cupping compared to those with glaucomatous cupping.

RNFL thinning on OCT should be evaluated and analyzed with other additional examinations including the evaluation of non-glaucomatous cupping, central and color vision, and the existence of disproportionate optic nerve pallor. In RNFL measurements, wedge defects are more common along the superior and inferior vascular arcades in glaucoma. However, in non-glaucomatous etiologies, RNFL thinning could be sectoral, temporal, or diffuse, depending on the etiology, and nerve fiber loss is more common in the papillomacular bundle.[12]

There is controversy regarding the management of sphenoid bone dysplasia. Rates of optic nerve involvement range between 50% and 90% in various studies.[17] It is generally accepted that asymptomatic sphenoid bone dysplasia should be closely followed both clinically and radiologically, but does not require biopsy or surgery. Surgical decompression can be performed on patients with nerve compression-related symptoms. Surgical decompression of asymptomatic patients with signs of optic nerve compression on radiologic imaging remains debatable.[18] In our case, the existence of optic nerve compression required neurosurgical consultation, which resulted in the decision to perform neurosurgery in order to prevent further compression and damage.

Our case emphasizes the importance of diagnosing glaucoma with extreme care. Non-glaucomatous diseases such as compressive etiologies can mimic glaucoma by producing glaucoma-like optic disc cupping, like in our case. Misdiagnosis and delay in starting appropriate treatment in such cases can have detrimental effects on long-term visual outcomes. Ophthalmologists should be aware of the differentiating features of non-glaucomatous optic neuropathy and consider neuroimaging to exclude any compressive lesions that mimic glaucoma.

Ethics

Informed Consent: Obtained.

Peer-review: Externally peer reviewed.

Authorship Contributions

Surgical and Medical Practices: P.K., Ö.K., Ö.E., Concept: P.K., Ö.K., D.T.K., Design: Ö.K., D.T.K., Data Collection or Processing: P.K., D.T.K., Analysis or Interpretation: P.K., Ö.K., D.T.K., Ö.E., Literature Search: P.K., Writing: P.K., Ö.K.

Conflict of Interest: No conflict of interest was declared by the authors.

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