Highly Asymmetric Keratoconus and Optic Neuropathy in Monostotic Fibrous Dysplasia: A Case Report

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Background

- Fibrous dysplasia (FD) is a primary orbital bone tumor in which proliferation of osteoid and fibrous tissue distorts and replaces medullary bone
- Orbital FD typically presents with periorbital facial asymmetry and proptosis with apical involvement of the orbit
- To our knowledge, there is only one reported case of unilateral isolated cranial FD with ipsilateral keratoconus

Case History

Chief Complaint: 22-year-old Caucasian male was referred to the clinic for a medically-necessary contact lens fit OS

History of Present Illness: wears glasses most of the time and depends heavily on OD, had previously been unsuccessfully fit in a corneal gas permeable lens OS, optic neuropathy OS being managed by OMD but difficulty testing due to keratoconus

Ocular History: severe keratoconus, orbital proptosis, and optic neuropathy OS, subclinical keratoconus OD

Medical History: monostotic craniofacial fibrous dysplasia with involvement of the left frontal bones, extending into the ethmoid and sphenoid bones, involving the superior and lateral walls of the left orbit

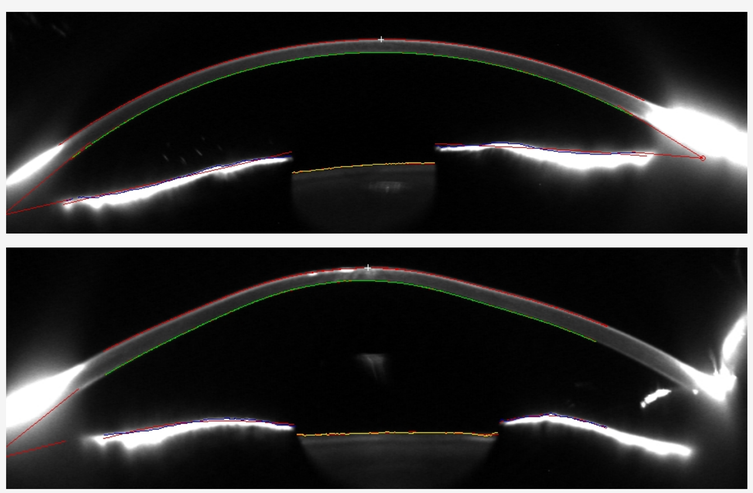

FIGURE 1: Scheimpflug image OD (top) and OS (bottom)

Case Details

Best-corrected spectacle VA	OD: -1.50-1.00x120 20/20 OS: +2.25-1.25x114 20/30
External examination	macrocephaly with frontal bossing and left proptosis
Corneal findings	OD: clear, no apparent stromal thinning OS: Vogt striae, stromal thinning, positive Munson sign
Keratometry readings	OD: 41.5D/43.0D@068 OS: 59.4D/67.4D@052
Pachymetry	OD: 510 microns OS: 405 microns
Optic Nerve Head	OD: pink, flat, sharp margins, 0.2/0.2 C/D OS: diffuse pallor, flat, sharp margins, 0.2/0.2 C/D

Assessment

1. Severe keratoconus OS, subclinical keratoconus OD
2. Fibrous dysplasia with optic neuropathy OS (managed by OMD). Historic testing with HVF revealed cecocentral scotoma OS with temporal depression, no defects OD. Unreliable ganglion cell and retinal nerve fiber layer OCT scans OS, no defects OD. Normal color vision OD, OS. Positive APD OS.

Plan

1. Fit with Synergeyes VS scleral lens OS and after modifications, achieved 20/20 OS. Continue spectacle wear, replace OS lens on current specs with plano polycarbonate lens. Refer for corneal cross-linking OS.
2. Refer to OMD for repeat testing to monitor optic neuropathy. Increased reliability and scan quality expected with scleral lens wear OS.

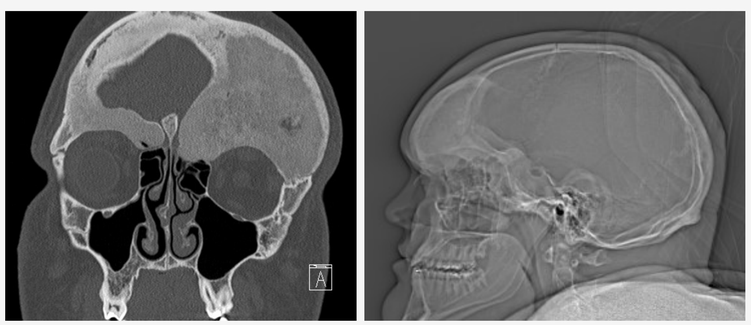

FIGURE 2: CT of orbits, maxillofacial bones, and paranasal sinuses without contrast (left) and CT topogram (right)

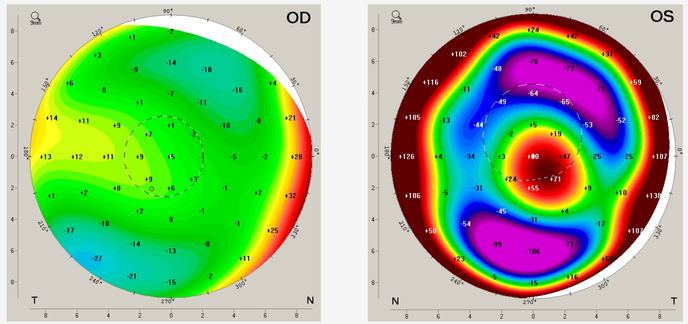

FIGURE 3: Corneal front elevation map OD and OS

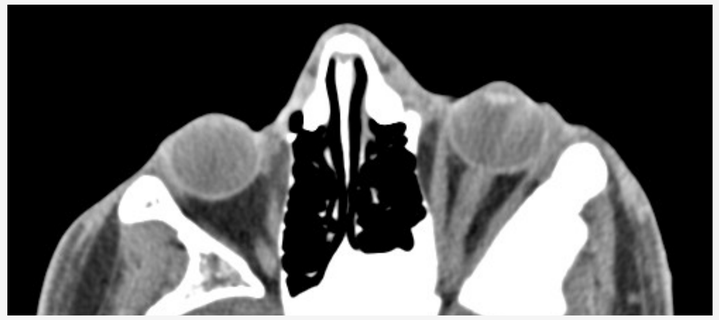

FIGURE 4: CT of orbits without contrast

Conclusions

- While the development of keratoconus in FD is suspected to result from external globe pressure and displacement, the exact etiology is unknown. Mechanical disturbance of corneal structures has been proposed, yet still not proven.
- Due to the progressive nature of the disease, patients with FD and keratoconus need to be more closely monitored with corneal topography and pachymetry, and more immediate referral for corneal crosslinking may be indicated.

References

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3. Cruz AA, Constanzi M, de Castro FA, dos Santos AC. Apical involvement with fibrous dysplasia: implications for vision. Ophthal Plast Reconstr Surg 2007;23(6):450–454.