Posterior Scleritis: A Unique Sequela of Cogan Syndrome

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CASE REPORT

A 42-year-old female with a history of Cogan syndrome, sensorineural hearing loss, and coronary vasculitis presented with three days of deep, boring, right-eye pain and photophobia. Visual acuity was 20/200 on the right and 20/20 on the left. Pupils were without an afferent pupillary defect. Extraocular movements were limited in all positions of gaze. Examination revealed scleral injection (**Fig.1**), pain with extraocular movements, and 3 mm of proptosis of the right eye. Dilated fundus exam showed chorioretinal folds (**Fig. 2**). Ophthalmic B-scan ultrasound of the right eye showed thickening of the sclera with fluid in tenon's space (**Fig. 3**). Left eye appeared normal. A diagnosis of posterior scleritis was made and the patient was treated with intravenous steroids. She was transitioned to oral prednisone, infliximab, and methotrexate with resolution of her ocular symptoms and improvement of visual acuity to 20/40 of the right eye.

Figure 1. External photograph of the right eye with scleral injection and deep corkscrew vessels. Mid-peripheral cornea with interstitial corneal deposits without an associated epithelial defect.

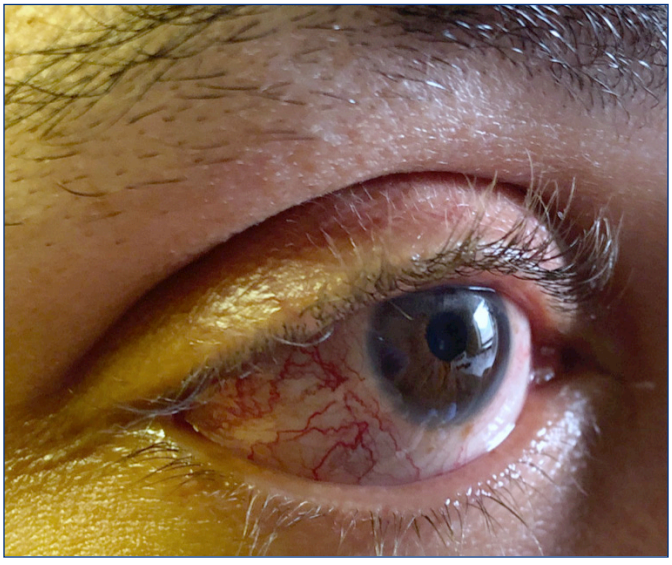


Figure 2. Fundus photograph of the right eye demonstrates chorioretinal folds emanating from the optic nerve head (green arrow).

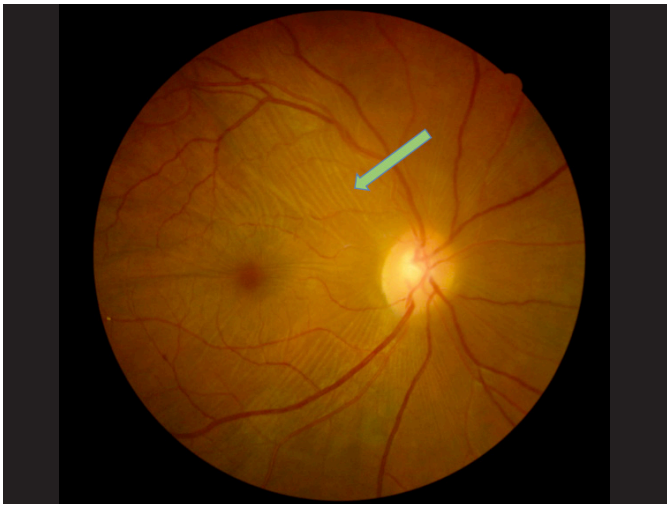
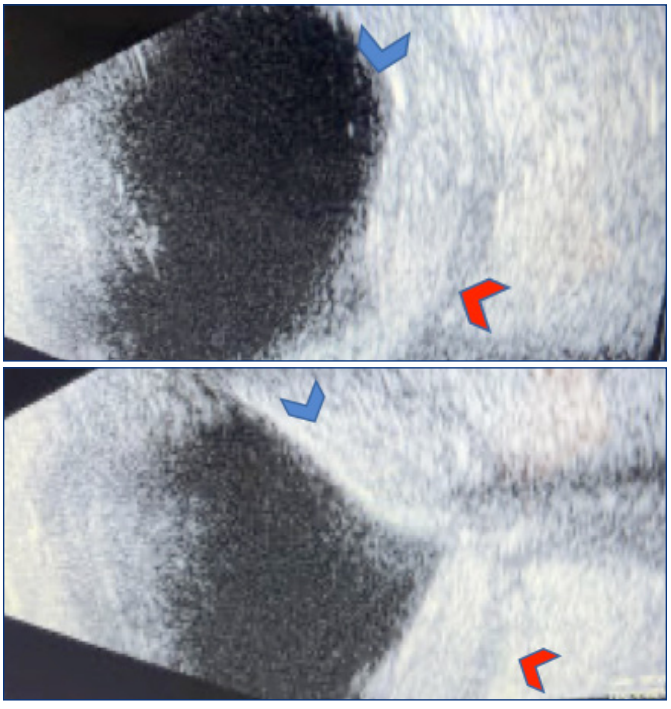


Figure 3. Ophthalmic B-scan ultrasound of the right eye shows thickening of the sclera (blue arrowheads) and fluid in tenon's space (red arrowheads).



DISCUSSION

Cogan syndrome is a chronic inflammatory disease typically seen in young adults that often leads to sensorineural deafness and ocular inflammation by a presumed autoimmune etiology.[1] Ocular manifestations may vary but typically involve interstitial keratitis.[1]

In addition to vision and hearing deficits, systemic involvement is present in up to 80% of Cogan syndrome patients including coronary vasculitis, headache, fever, arthritis, and myalgias.[1] Atypical ocular presentations of Cogan syndrome include episcleritis, choroiditis, uveitis, optic disc edema, angle closure glaucoma, and central vein occlusion.[1] There are few reported cases of posterior scleritis related to Cogan syndrome.[1,2,3] This rare finding presented in our patient with ocular pain, vision loss, proptosis, choroidal folds, and scleral thickening noted on B-scan.

CONCLUSION

Cogan syndrome should be considered in those with ocular inflammation and hearing loss. Diagnosis is often delayed given the lack of definitive testing and the fact that auditory symptoms and keratitis commonly do not present simultaneously, often occurring several months to years apart.[1] Given the vision and life-threatening sequelae of Cogan syndrome, it is critical for providers to be aware of the various ocular and systemic manifestations.

References

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