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Bilateral zonular dehiscence during cataract surgery in a patient with systemic sclerosis

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ABSTRACT

Purpose: Systemic sclerosis, also known as scleroderma, is a rare and chronic autoimmune connective disorder that affects most organs. While clinical findings of scleroderma patients in the context of the eye have been described to include lid fibrosis and glaucoma, almost nothing has been reported regarding ophthalmologic surgical complications in scleroderma patients.

Observations: Here, we report bilateral zonular dehiscence and iris prolapse during two independent cataract extractions performed by separate experienced anterior segment surgeons in a patient with known systemic sclerosis. The patient did not have any other known risk factors for these complications to occur.

Conclusions: and Importance: In our patient, bilateral zonular dehiscence raised the possibility of poor connective tissue support secondary to scleroderma. We recommend that clinicians are aware of potential complications in performing anterior segment surgery in patients with known or suspected scleroderma.

1. Introduction

Systemic sclerosis is a chronic multisystem connective tissue disease with well-described dermatologic and visceral manifestations.[1] The underlying pathophysiology is not completely understood, but it is hypothesized to result from overactivation of fibroblasts leading to unregulated production of extracellular matrix and collagen, which ultimately deposit in affected tissues. Most patients develop the primary feature of fibrotic skin thickening. Additional systemic features which occur with varying prevalence may include digital ischemia, cardiac conduction defects, interstitial lung disease, renal dysfunction, dysphagia, and muscular weakness, among others. Diagnosis is made based on clinical features and may be supported by the presence of specific serum tests including anti-topoisomerase I, anticentromere antibody, and anti-RNA polymerase III among others. Skin biopsy may provide additional support, though it is not considered necessary for diagnosis. Different classification systems have been developed to measure disease severity, with the Medsger scale being commonly utilized to determine organ involvement and prognosis. Here, 9 organ specific severity scales are given for each affected tissue and a score is given of 0, no involvement; 1, mild involvement; 2, moderate involvement; 3, severe involvement; and 4, end stage failure.[2]

Excluding the more common findings of lid fibrosis and dry eye, ocular manifestations of scleroderma are infrequently reported. Like the rest of the skin, the eyelids are at risk of fibrosis, pigmentary changes, and microvascular abnormalities. Decreased tear production and nasolacrimal duct obstruction lead to keratoconjunctivitis sicca and chronic dacryocystitis.[3] Multiple reports have found an increased prevalence of glaucomatous optic neuropathy among scleroderma patients,[4–6] which may be attributable to inflammation or microvascular disease.[6] Microvascular abnormalities within the retina are also described, though it is unknown whether these result directly from the scleroderma disease given the increased prevalence of hypertension within this population.[4] Ocular inflammation has also been described, including episcleritis, scleritis, and uveitis.[7] The incidence of cataracts in scleroderma patients is comparable to that of age-matched controls,[8] though the use of systemic steroids for scleroderma may be a confounding factor. Intriguingly, almost nothing has been reported regarding ophthalmologic surgical complications in scleroderma patients.

Here, we present a patient with systemic sclerosis who developed

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bilateral zonular dehiscence during each of his independent cataract surgeries by different surgeons. This possible association is important for ophthalmologists to be aware of prior to attempting anterior segment surgery in patients with known or suspected systemic sclerosis.

2. Case report

A 55-year-old African American male with a past ocular history of bilateral open angle glaucoma presented with gradual blurring of vision, glare, and halos. His medical history included scleroderma, diagnosed 10 years prior with manifestations including Raynaud's phenomenon with digital ulcerations, digital skin tightness, pulmonary hypertension, and gastric reflux, with affected organ systems falling in the mild to moderate Medsger severity scales. Additional medical history included discoid lupus erythematosus, hypertension, and atrial fibrillation. Prior hematologic testing had revealed a positive anti-nuclear antibody with titer 1:640 in a nucleolar pattern. Notably, he was of normal body habitus with a height of 6 feet and weight of 165 pounds, and with no known diagnosis of homocystinuria or Marfan syndrome. His medications included Levothyroxine, Bosentan, Omeprazole, Amlodipine, and specifically with no current or prior alpha-blocker usage. Ocular medications included Latanoprost, Timolol, and Brimonidine, and he had no prior surgical or laser procedures on either eye. Social history included a 35-pack year smoking history with moderate to heavy alcohol use.

On presentation, his best-corrected vision was 20/40+ in the right eye (OD) and 20/70- on the left (OS). Intraocular pressures were 15 and 17 mmHg, respectively. His external exam showed chronic lid fibrosis with inelasticity of the bilateral upper and lower eyelids. His anterior segment was notable for combined cataracts. His right eye had a trace central posterior subcapsular opacity with 2+ nuclear cataract, while his left eye had a denser 1–2+ posterior subcapsular component with 2+ nuclear cataract. There was no noted iridodonesis, phacodonesis, iris sphincter tears or pigment on the lens capsule to indicate evidence of previous trauma. His pupils dilated to 6 mm bilaterally, and there was no evidence of pseudoexfoliative material on the anterior lens capsule after dilation. His posterior exam was unremarkable other than his cupped nerves consistent with his glaucoma history. On biometry, axial lengths were 23.77 mm OD and 23.92 mm OS, with anterior chamber depths of 3.56 mm OD and 2.59 mm OS. Keratometry readings OD were 46.50 diopters at 128° and 44.25 diopters on the flat axis, and OS 44.74 diopters at 163° and 44.54 diopters at 73°. After extensive discussion, the patient elected to proceed with cataract extraction first in the left eye.

During the initial steps of the cataract surgery the anterior capsule was noted to be abnormally thickened. Inferonasal zonular instability and iris prolapse through the main wound were apparent during capsulorrhexis formation. Even with careful manipulation, zonular dehiscence extended 180° inferiorly, leading to posterior subluxation of the lens and anterior prolapse of vitreous. Iris hooks were placed to control the iris and the decision was made to convert to an intracapsular cataract extraction. The cataract was successfully delivered through a scleral tunnel using an irrigating vectis. A limited anterior vitrectomy was performed, and the patient was left aphakic. The patient had the expected post-operative course and at month four, a 25-gauge pars plana vitrectomy with scleral sutured intraocular lens was completed by the retina specialist without complication. After resolution of post-operative inflammation and corneal edema, his best-corrected vision in the left eye stabilized at 20/40.

Almost one year later, the patient returned with subjective worsening vision and glare in his right eye. Best-corrected visual acuity at that visit was stable at 20/40- in the left eye, and 20/40- in the right eye. Slit lamp re-examination revealed a stable and centered IOL in the left eye, and progressing 2+ nuclear sclerotic cataract with 1+ posterior subcapsular component on the right eye, with now appreciable phacodonesis and inferior lens displacement. The patient was scheduled for a planned intracapsular cataract extraction due to the anticipated complexity of the case and lack of zonular support. Again, extensive zonulopathy was noted early in the case with lens hypermobility and inferonasal displacement during viscoelastic fill. There was again iris prolapse mid-case. During delivery of the intact lens with an irrigating vectis through the scleral tunnel, there was rupture of the fragile capsule, allowing for some small lens fragments to the posterior vitreous. The patient was again left aphakic with a planned secondary lens placement. At post-operative month one, the patient underwent a 25-gauge pars plana vitrectomy with Yamane scleral fixated intraocular lens without intraoperative complications, though his post-operative course and vision have been complicated by corneal decompensation, likely from the sequential surgeries. He is now under the care of a corneal specialist, post-op month 10, with the latest vision is vision in the right eye at count fingers at 10 feet, limited by corneal edema.

3. Discussion

In reviewing the literature, we found a paucity of knowledge regarding cataract surgery outcomes in patients with scleroderma. Scleroderma is known to cause extensive multi-organ dysfunction, but its effects are not well-described in the anterior segment of the eye. Unless a careful medical history is taken, ophthalmologists may not even be aware of this diagnosis in their patients. Cataract surgery is generally highly successful, with complication rates of zonular dehiscence at 1.1%, and lens subluxation at 0.3%.[9] Despite these favorable statistics, our patient developed nearly identical intraoperative zonular dehiscence during cataract surgery in each of his two eyes, on separate occasions and with different surgical teams. We hypothesize that this is suggestive of underlying zonular instability secondary to scleroderma. Additionally, common findings encountered in both eyes included steep corneas, iris prolapse, and inferonasal zonulopathy.

This clinical presentation may stem from the microanatomy of the Zonules of Zinn. The lens zonules, composed of bundles of glycoprotein microfibrils, originate at the nonpigmented epithelium of the ciliary body and insert circumferentially on the equatorial lens capsule to suspend the lens. One of the major known components is fibrillin, a large glycoprotein found in connective tissues and vasculature. The lens capsule, composed of glycosaminoglycans and type IV collagen,[10] is the avascular basement membrane of the lens epithelial cells into which the zonules anchor. For normal zonule integrity and function, the proximal and distal anchoring points, as well as the body of the zonules, must be intact.

Zonular dehiscence and lens subluxation may occur in the setting of inherent zonule instability, secondary to another process, traumatic, or iatrogenic. In this case, neither eye had a known history of secondary processes. Spontaneous dehiscence is associated with several systemic diseases, which provide insight into causes of innate zonular weakness. Marfan syndrome, caused by an autosomal dominant mutation in the fibrillin 1 protein, is a direct cause of zonule weakness from which 60% of patients develop spontaneous ectopia lentis.[11] In Marfan syndrome, however, the lens is typically subluxated in the superotemporal direction, whereas in this patient the left lens was displaced inferiorly, similar to the displacement typically encountered in homocystinuria. Homocystinuria, an autosomal recessive deficiency in an enzyme responsible for homocysteine metabolism, weakens the zonules by accumulation of abnormal glycoproteins. In this case, our patient had no history or physical characteristics common to these diseases.

A well-known example of zonular weakness due to a systemic condition can be seen in pseudoexfoliation syndrome. In this disease of abnormal extracellular matrix turnover, deposits of fibrillin and elastin fibers, as well as laminin and glycosaminoglycans, accumulate in ocular tissues including the zonules and ciliary body. These patients are at greater risk of cataract surgery complications than the general population, with overall complication rates of 21.5%, zonular dehiscence 6.7%, and lens subluxation 2%.[9] Our patient had no characteristic findings of PXE material on the anterior lens capsule. Furthermore, our patient was of African American descent, in which pseudoexoliation syndrome is

relatively rare.[12] We hypothesize that that the zonules and their attachments to the ciliary body were inherently weak and unable to withstand the mechanical stress of surgery. There are some noted findings on biometry for this patient, including a deeper anterior chamber depth in the right eye during initial pre-operative measurements. Though there was no noted phacodonesis in the right eye initially, the deeper anterior chamber depth implies possible zonular instability at that time. His subsequent evaluations later revealed phacodonesis and lens displacement, confirming the asymmetric zonular states in either eye.

What does this mean for the comprehensive ophthalmologist? A thorough medical history is critical for pre-operative planning and individualized patient counselling. Pre-operative examination should always include careful assessment for adnexal skin changes and phacodonesis, which may indicate increased risk of intraoperative complications. If zonular dehiscence occurs in one eye, an underlying etiology should be investigated prior to operating on the second eye. Patients with known scleroderma should be counseled regarding the potential for zonular dehiscence and lens subluxation with a possible later need for additional surgery. In some cases, a planned intracapsular lens extraction may be preferable, and surgical equipment for any of the above scenarios should be available in the operating room on the day of surgery. We submit this case to alert clinicians to this potential risk factor for intraoperative complication in order to provide the best possible care for these patients.

4. Conclusions

Ophthalmologists performing cataract surgery on patients with systemic sclerosis should be prepared for zonular instability and should counsel the patient pre-operatively of this possibility.

Patient consent

Informed consent for the above case information to be published was provided by the patient.

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