

An 18-year-old Male with Severe Bilateral Cataracts and Atopic Dermatitis—A Case Report and Review of the Literature

Lisa Mondzelewski, M.D.,* Catherine Hagan, M.D.,† and Andrew A. White, M.D.‡

**Department of Pediatrics, Naval Hospital Camp Pendleton, California, †Department of Ophthalmology, Naval Medical Center, San Diego, California, ‡Department of Allergy and Immunology, Naval Medical Center, San Diego, California*

The association of atopic dermatitis (AD) with cataract formation was first reported in 1914 (1). Despite this early recognition, a unifying explanation for cataract progression in patients with AD is lacking. A cataract is an opacity of the lens which can lead to loss of vision. The general classification of cataract includes nuclear, cortical and posterior subcapsular subtypes. The atopic cataract (AC) typically begins as a posterior and/or anterior subcapsular opacity with eventual development into a fully mature cataract (2,3) (Figs. 1 and 2). They are most often bilateral and are primarily seen in adolescents and young adults. Although cataracts are known to be associated with AD, this seems to be an underappreciated phenomenon.

CASE REPORT

An 18-year-old, Filipino American male, was followed for a longstanding history of asthma, allergic rhinitis, food allergy, and AD. His asthma was well controlled with inhaled corticosteroids (ICS). His AD had been present since childhood, and controlled with topical emollients, pimecrolimus 1% cream, and cetirizine. There was no other significant past medical history.

At age 15 he described decreasing visual acuity. At this time an optometrist diagnosed him with an early cataract in his left eye. Over the next 12 months, his vision worsened to the extent that it was performance limiting in

school and fell outside of the legal driving limits. Two years after his initial unilateral cataract diagnosis; he was diagnosed with visually significant bilateral cataracts. He denied eye pruritis, burning, discharge, redness or eyelid dermatitis. He denied any application of topical steroid medications to the face or around the eyes. He remained on a moderate dose of inhaled corticosteroid for asthma (fluticasone propionate/salmeterol diskus 250 µg/50 µg), and a nasal corticosteroid for allergic rhinitis. Physical exam revealed postinflammatory hyperpigmentation on his face and periorbital region. The patient had diffuse xerosis over his trunk and extremities. He admitted to frequent tapping or rubbing of his face and body because of pruritis.

After the development of the second cataract an extensive laboratory workup to rule out a systemic cause of progressive cataracts was unrevealing. Upon presentation to the ophthalmologist, the patient's visual acuity was 20/60 right eye and 20/100 left eye. The anterior ocular exam showed moderate hyperkeratosis of the eyelid skin, healthy bulbar conjunctiva, palpebral conjunctiva with fine papillae and corneal epithelium with only few punctuate epithelial erosions indicating mild ocular surface involvement. The changes in the left lens were denser than the right lens, both including a whitish nuclear sclerosis with posterior and anterior subcapsular changes in a rosette formation as well as wrinkling of the anterior lens capsule (Fig. 3). No lens/zonule instability

Address correspondence to Andrew A. White, M.D., Allergy and Immunology Clinic, 2051 Cushing Road, San Diego, CA 92106, or e-mail: drew_white@hotmail.com.

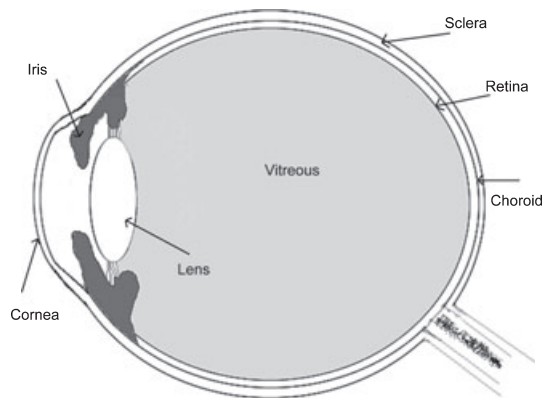


Figure 1. Ocular anatomy.

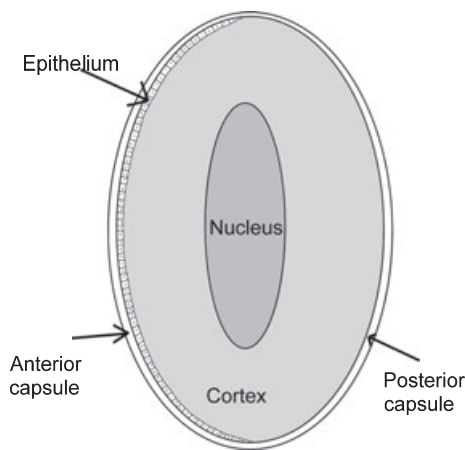


Figure 2. Anatomy of the lens.

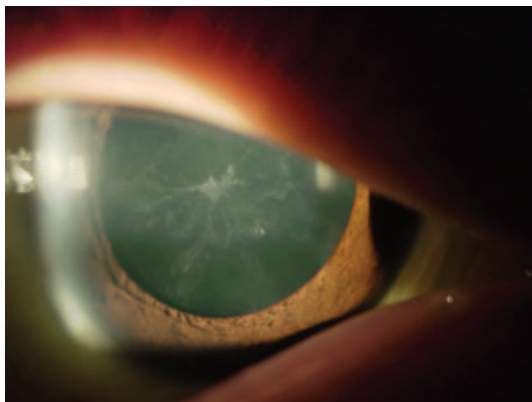


Figure 3. Right lens with whitish, hazy flower shaped cataractous changes.

was appreciated. The fundus exam was normal in each eye without retinal tears, holes, or detachments. There was no evidence of current or previous anterior or posterior intraocular inflammation. His ocular exam and

keratometry showed no evidence of keratoconus, in which the central or paracentral cornea undergoes progressive thinning and bulging, leading to a cone shaped cornea.

The patient underwent cataract extraction with intraocular lens implantation of both eyes within a 5-month period. The lens was removed using a standard clear corneal phacoemulsification technique with a continuous curvilinear capsulorhexis. This is a method by which a circular opening is created in the anterior portion of the capsule. The lens is removed in small fragments with ultrasound (phacoemulsification) and the remaining capsule can then act as a "sac" for the new intraocular lens. An acrylic, single-piece lens was then inserted within the capsular bag without complication. A thorough fundus exam using the slit lamp with 3-mirror lens and indirect methods showed no retinal defects in either eye. Since his surgery, he has returned to school and reported full activities of daily living. His final corrected visual acuity was 20/15-3 in the right eye, and 20/15 in the left eye.

DISCUSSION

The pathophysiology linking AD and cataract development is unclear, although several clinical features seem to play a role and may help predict patients who are at highest risk. Eye rubbing commonly occurs in AD patients as a means of reducing periorcular pruritis (4,5). Blunt trauma from frequent eye rubbing results in the breakdown of the blood-aqueous barrier, thereby releasing proteinaceous particles into the anterior chamber (6).

Cataract progression in AD has been associated with facial dermatitis severity and possibly with soft contact lens wear (7). A higher frequency of AC with the more severe forms of AD in general may represent susceptibility versus an absolute correlation (8,9). Some reports found a lack of relationship between the distribution of skin lesions, clinical course of the dermatitis, history of respiratory atopy, serum IgE level, duration of topical corticosteroid use of the face, or treatment with systemic corticosteroids with AC formation (3,8,9).

Various proteins have been studied and implicated as possible mediators of cataract formation in AD. Major basic protein (MBP) is derived from the granules of eosinophils and is known to alter cell membrane permeability resulting in cell death. Major basic protein has been linked to lung tissue damage in asthmatics, and has been found in tears, corneal ulcerative lesions, and the conjunctiva of patients with vernal keratoconjunctivitis, as well as in the conjunctiva of patients with atopic keratoconjunctivitis (10). Deposition of MBP has been

found within the lens epithelial cells (LEC) of AC (2,11). Protein gene product 9.5 (PGP 9.5) has been studied in AC. Of its many properties, PGP 9.5 serves a defense function preventing abnormal proteins generated by stress, such as MBP, from spending metabolic energy. Once this defense system is overwhelmed, accumulation of abnormal proteins ensues leading to LEC degeneration and transition into fibroblasts with resultant cataract formation (11). Cytotoxic anti-lens epithelium-derived growth factor (LEDGF) autoantibodies have been implicated as an etiology for AC. Without LEDGF, LEC were much more vulnerable to stress (12). High levels of serum lipids and decreased superoxide dismutase activity appear to be correlated with AD, and also independently related to human cataract formation, thus suggesting another possible link between these two diagnoses (13).

Not only has there been a strong association of AD with cataract formation but also with retinal detachment. As awareness and retinal evaluation techniques improve, these findings are more often discovered together and have been referred to as a triad (3,14). There is an overall increase in frequency of retinal detachments in patients with AD (5,8). Many propose chronic ocular trauma from eye rubbing or tapping results in tears or breaks of the retina or ciliary body epithelium, with some progressing to retinal detachment (7). Globe compression in the antero-posterior direction causes globe expansion at the corneoscleral ring producing tension between the zonular fibers and their attachment to the ciliary epithelium (5). It is therefore not surprising that the majority of breaks and detachments are found in this region at the vitreous base and pars plana/plicata (5). It has also been noted there are common features shared by retinal detachments associated with AD and those due purely from trauma (5).

As with AC, the use of topical steroids, systemic steroids, or IgE level have not correlated with presence of retinal detachment (7). However, retinal detachments are reported to occur most frequently in patients who have already developed AC, with the highest frequency found in those with mature cataracts (14).

Interestingly, an exhaustive review of world literature describing case series of cataracts in AD reveals a majority of patients from Asian populations. A total of 31 articles are published from the Japanese population, and one series from Taiwan and Singapore. This is contrasted with 21 total case series, case reports, and clinical studies in the rest of the world literature in non-Asian populations. Based on this, it would seem that there is either a greater interest in AC in the Asian population, or the prevalence and significance of AC is greater in this population. A report from a Danish

population of 51 patients with AD demonstrated no patients with cataracts (15). Yet, in a Japanese population, of 153 consecutive patients with AD, 19 (12.4%) were identified with cataracts (8). Another Japanese series identified a prevalence of 25.3% (7). While no large epidemiologic studies existed to confirm this finding, it is certainly plausible that cataract formation in other non-Asian ethnic groups with AD might be considerably less. In a large practice of both pediatric and adult allergy patients, this is the first child with AD and clinically significant cataracts that we are aware of. That this patient was Filipino, could be consistent with the above observation. A larger study of prevalence in other patient populations is necessary.

Finally, an evaluation of cataract formation in the atopic patient must include corticosteroid exposure. In particular, ICS are frequently used for coexistent asthma in individuals with AD. There does appear to be an increased risk of posterior subcapsular cataract formation in adults on higher doses of ICS (16). Well powered studies that evaluate this issue in the pediatric population do not exist. While quite rare, ICS use should still be considered as a confounding factor in any patient being evaluated for AC.

While there have been many clinical associations noted in patients with AD and cataract formation, there are still no definitive answers as to what exactly links these two disorders. It is likely multifactorial with direct mechanical trauma, inflammation from the inherent atopic disease process, and cytotoxic proteins and/or autoantibodies contributing. The natural response of the human lens to any form of injury is to form a cataract. Whether aggressive control of facial or periocular dermatitis may help reduce the chance of cataract development and progression remains to be seen, but should be attempted. It is clear that any patient with AD and visual changes warrants prompt referral for ophthalmologic exam. Results of cataract and retinal detachment surgery are generally very good and associated with restoration of visual acuity and low complication rates. Whether generalized ocular screening exams should be performed or utilized in specific ethnic populations will remain a topic of future study.

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