KJO
pISSN: 1011-8942 eISSN: 2092-9382

Acute Angle Closure Glaucoma in a Patient with Pseudoxanthoma Elasticum

Korean J Ophthalmol 2017;31(5):462-463
https://doi.org/10.3341/kjo.2017.0042

Dear Editor,

Pseudoxanthoma elasticum (PXE), also known as Gröenblad-Strandberg syndrome, is an inherited multi-systemic disorder characterized by progressive mineralization and fragmentation of elastic fibers in various organs including the skin, eyes, and vascular system. The reported ocular manifestations of PXE include optic nerve drusen, angioid streaks, large regions of retinal pigment epithelium atrophy, and peripapillary atrophy with large dehiscence of Bruch's membrane [1].

Although the relationship between PXE and the development of acute angle closure is not yet fully understood, one recent study suggested that PXE may be associated with mild myopia due to the presence of elastic zonular fibers that could predispose patients to anterior displacement of the lens [2]. The authors report a case of acute angle closure glaucoma (AACG) that is presumed to have arisen from zonular changes secondary to PXE.

A 60-year-old Asian female with no ocular disease history visited Keimyung University Dongsan Medical Center with a chief complaint of right ocular pain and visual disturbance 24-hour in duration. The patient was diagnosed with PXE via skin biopsy of multiple, yellowish, flat papules on the neck 20 years prior (Fig. 1A-1C). On initial examination, best-corrected visual acuity was 20 / 160 (right eye [OD]) and 20 / 25 (left eye [OS]); intraocular pressure (IOP) as measured by Goldmann applanation tonometry was 72 mmHg (OD) and 16 mmHg (OS). Slit lamp examination revealed diffuse corneal epithelial edema and an extremely shallow anterior chamber (Schaffer grade 0 by gonioscopy). Under the diagnosis of AACG (OD), immediate laser iridotomy was performed, and the IOP decreased to 27 mmHg. The following day, visual acuity improved to 10 / 25 and IOP decreased to 10 mmHg. After relief of corneal edema, angioid streaks radiating from the papilla were identified on bilateral fundus examination (Fig. 1D-1G). However, because the anterior chamber was still shallow and IOP was unstable, cataract surgery was performed. During surgery, phacodonesis and overall zonular weakness were identified and a capsular tension ring was implanted with an intraocular lens to stabilize the capsular bag. After cataract surgery, the depth of the anterior chamber increased and IOP stabilized (Fig. 1H, 1I). Although there was no acute angle closure event, the anterior chamber was also shallow in the left eye and characteristic features of PXE were observed on fundus examination. Therefore, we performed prophylactic laser peripheral iridotomy during the follow-up period and anterior chamber depth and IOP remained stable.

PXE is a rare, multi-systemic disorder of autosomal recessive inheritance that has an estimated prevalence of 1 : 25,000 to 1 : 100,000; it is associated with mutations in the *ABCC6* gene and leads to calcification and fragmentation of connective tissues rich in elastic fibers [3]. The few histologic studies that have investigated the eyes of patients with PXE have suggested a characteristic loss of choriocapillaris underneath a thickened and calcified Bruch's membrane and significant thinning of the choroid and sclera [1,4].

In this case, we report a case of AACG with anterior shift of the lens-iris diaphragm, which was presumably secondary to pathologic zonular changes resulting from PXE. A recent study suggested that the elastic zonular fibers may be affected in PXE, as in Marfan syndrome, and that patients with PXE probably have a lens that is readily movable anteriorly because of the presence of pathologic zonular fibers [5]. Therefore, we hypothesized that patients with PXE may also be vulnerable to AACG for the same reason. The current case is thought to support this hypothesis.

However, the number of studies investigating PXE and acute angle narrowing remains inadequate. Additional studies including larger populations, histologic findings and genetic studies are needed to confirm these preliminary findings.

Jung Yeob Han, Chong Eun Lee, Yu Cheol Kim

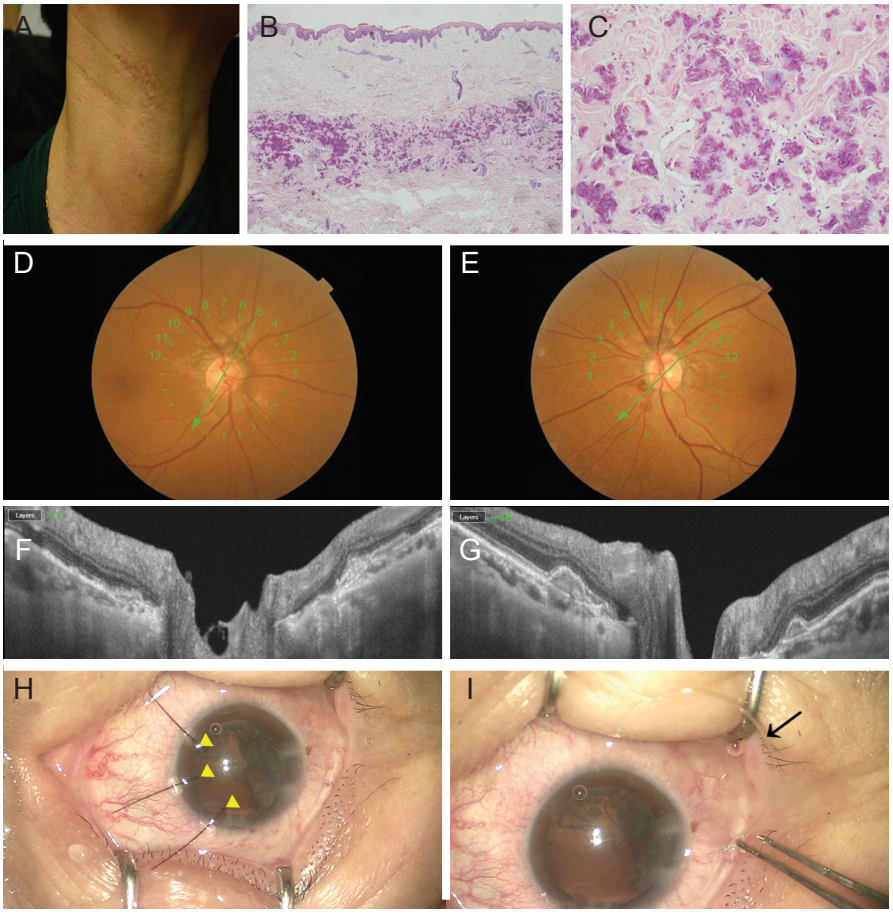

Fig. 1. (A) Photograph of the patient's neck showing multiple, yellowish, flat papules. (B,C) Biopsy of a skin lesion showing short, curled elastic fibers with basophilic calcification of the reticular dermis which are characteristics of pseudoxanthoma elasticum. Hematoxylin-eosin stain, ×40 original magnification (B), ×200 original magnification (C). (D,E) Fundus photographs (D: right, E: left) showing bilateral angioid streaks, which are narrow and irregular lines emanating from the optic disc. (F,G) Optical coherence tomographic images (F: right, G: left) showing pigment epithelial detachment and breaks in Bruch's membrane at the sites of angioid streaks. (H,I) Photographs during cataract surgery. (H) Iris retractors are placed at the capsulorrhexis edge (arrowheads) to support the area with zonular weakness. (I) A capsular tension ring (arrow) is inserted before intraocular lens implantation to stabilize the capsular bag.

Department of Ophthalmology, Dongsan Medical Center, Keimyung University School of Medicine, Daegu, Korea
E-mail (Yu Cheol Kim): eyedr@dsmc.or.kr

Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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