Clinical case of acute hydrops treatment using the method of penetrating keratoplasty

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Summary

The article is devoted to the description of a clinical case of acute keratoconus and its successful treatment using the method of penetrating keratoplasty. The diagnosis of acute hydrops was established on the bases of clinical and functional methods, including optical coherent tomography. Removed part of the pathologically changed cornea was subjected to light microscopy. Histological examinations revealed morphological changes in almost all layers of the cornea. Comparison of the OST data and morphological studies showed that the indications for penetrating keratoplasty in this concrete patient were chosen adequately. The article presents modern data on the epidemiology of this rare disease, its pathogenesis, which is based on the rupture of Descemete membrane and soaking of the corneal stroma with aqueous fluid. The existing methods of treatment of acute hydrops are aimed to remove inflammation of the cornea with the subsequent restoration of the integrity of endothelial-descemete layer and drainage of stromal cysts for optimization of corneal healing. In this particular case, penetrating keratoplasty was performed due to a significant thinning of the cornea and the danger of its total rupture. As donor material was used «Material for corneal graft» manufactured by eye bank «iLab».

Key words: keratoconus, penetrating keratoplasty, acute corneal hydrops, donor material for corneal grafting.

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A 27 years old male patient D. contacted the clinic with complaints of sudden decrease in visual acuity of the left eye, and low vision in the right eye.

The patient explained that his vision had deteriorated in both eyes for the past 5-6 years. Three years ago, he was diagnosed with grade 1-2 keratoconus in the right eye and grade 2 keratoconus in the left eye. The patient received no treatment; the refraction was corrected with soft contact lenses and glasses.

The patient reported that 7 week ago the visual acuity of his right eye decreased rapidly for no obvious reason, the anterior eye reddened and he had pain in the right socket, so the patient referred to the department of eye microsurgery for consultation and treatment.

At the initial visit, examination revealed moderate mixed ocular injection in OD, conjunctival hyperemia, corneal edema in all layers throughout the surface and thickening in optical zone. The anterior chamber preserved, the liquid was transparent, no deeper structures could be observed (Pic. 1).

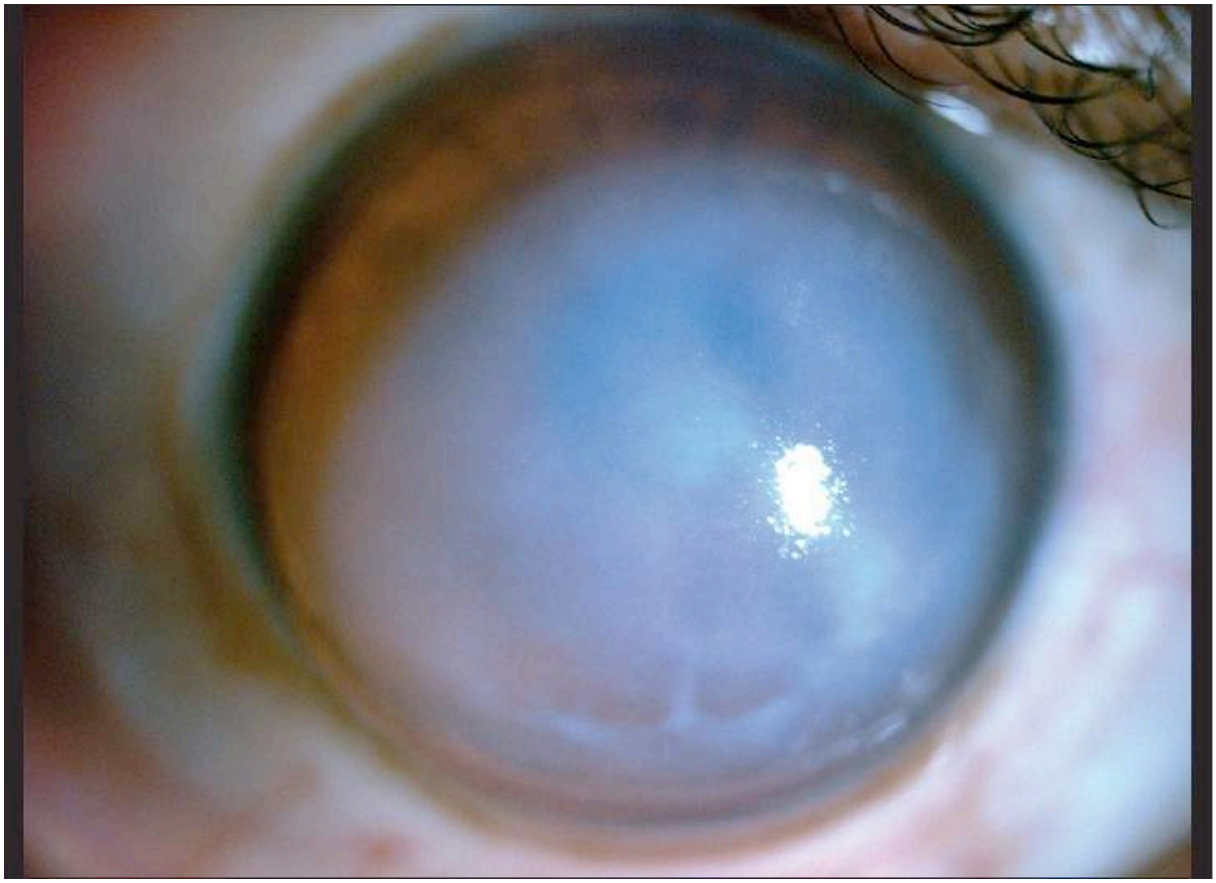

Fig. 1. Image of right eye anterior segment of patient D.

Left eye was undisturbed, ocular media were transparent with normal ocular fundus. His visual acuity was OD 0.01–0.02 incorrigible, OS 0.5 with sph -1.0 cyl -2.5 ax 160=0.9; IOP OD within the normal range by palpation, IOP OS: 19. Ultrasound keratopachymetric research showed central corneal thickness OD 923 μ, OS 480 μ. No corneal structural abnormalities revealed at OCT of the left eye, right eye examination discovered significant thickening of the cornea throughout the surface. Irregularly shaped massive cysts (cisterns) with acoustically transparent homogeneous content were observed mainly in central and paracentral areas of the corneal stroma, accompanied by stromal thinning over the cysts area (170-100 μ) (Pic. 2). The patient was diagnosed with OD acute keratoconus, OS grade 1-2 keratoconus.

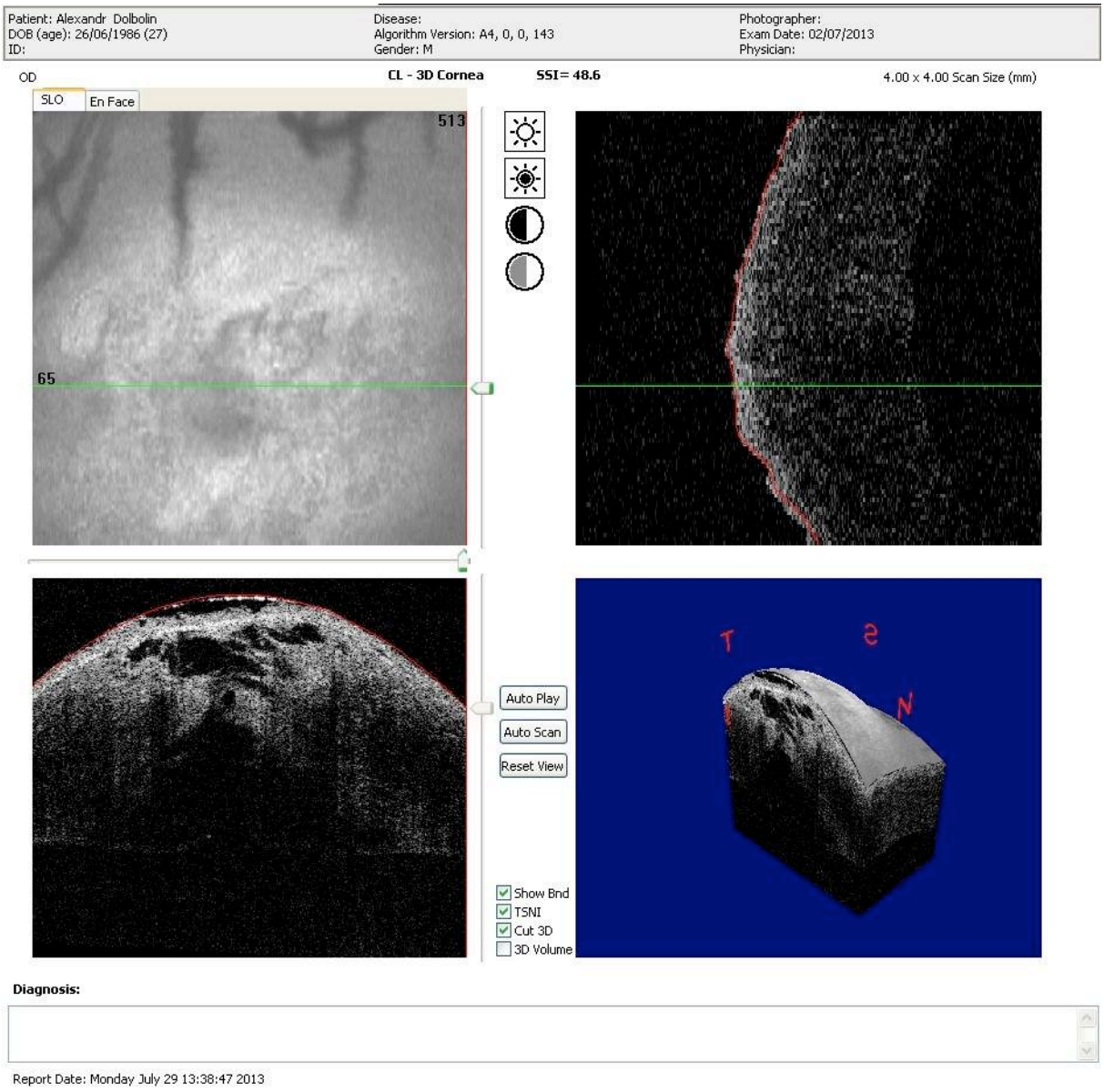

Fig. 2. Results of OCT- Optovue/ Corneal biomicroscopy software

The functional data presented indicate massive cisterns in corneal stroma, severe deformation of anterior surface of the cornea in central optical zone, edema and thickening of paracentral areas, and modified structure of stroma.

Taking into account significant thinning of right eye corneal stroma (to 100 μ) and increasing risk of its perforation in stromal cyst formation locus a decision was taken to perform penetrating keratoplasty.

The operation was performed on July 5, 2013 using the «Material for corneal graft» manufactured by eye bank «iLab». Surgery and immediate postoperative period passed normally. The eye condition at hospital discharge corresponded to duration and

severity of the intervention, corrected visual acuity was 0.2. The patient was transferred to outpatient treatment.

Histomorphological study of the patient's corneal tissue revealed acute corneal edema and morphological changes in all corneal layers (Pic. 3, 4 and 5).

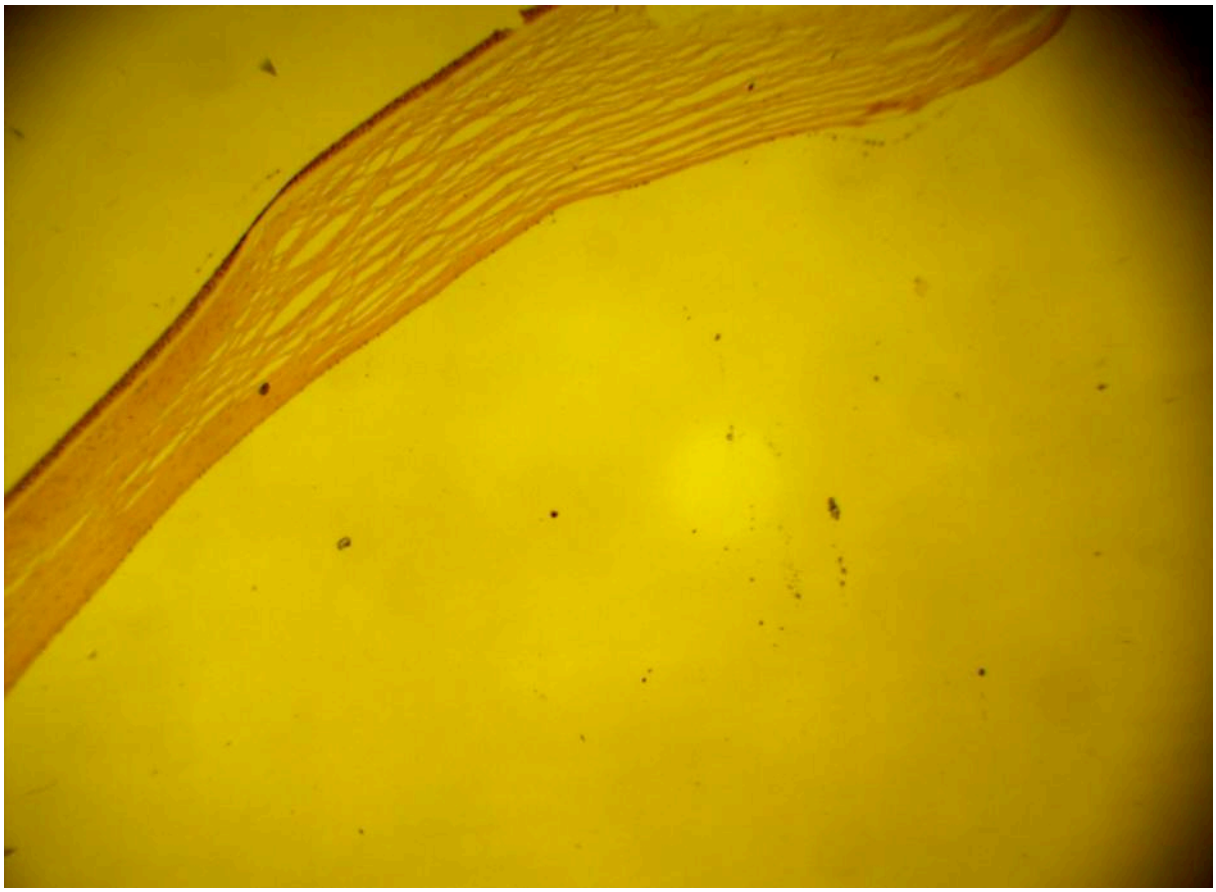

Fig. 3 Histomorphological image of corneal segment, Patient D., 27 years old.

Corneal stroma is extremely thinned closer to the optical zone. Bowman's membrane is strongly deformed. Magnification 100. Stained with haematoxylin-eosin.

Keratoconus manifestations in all corneal layers are particularly apparent at high magnification.

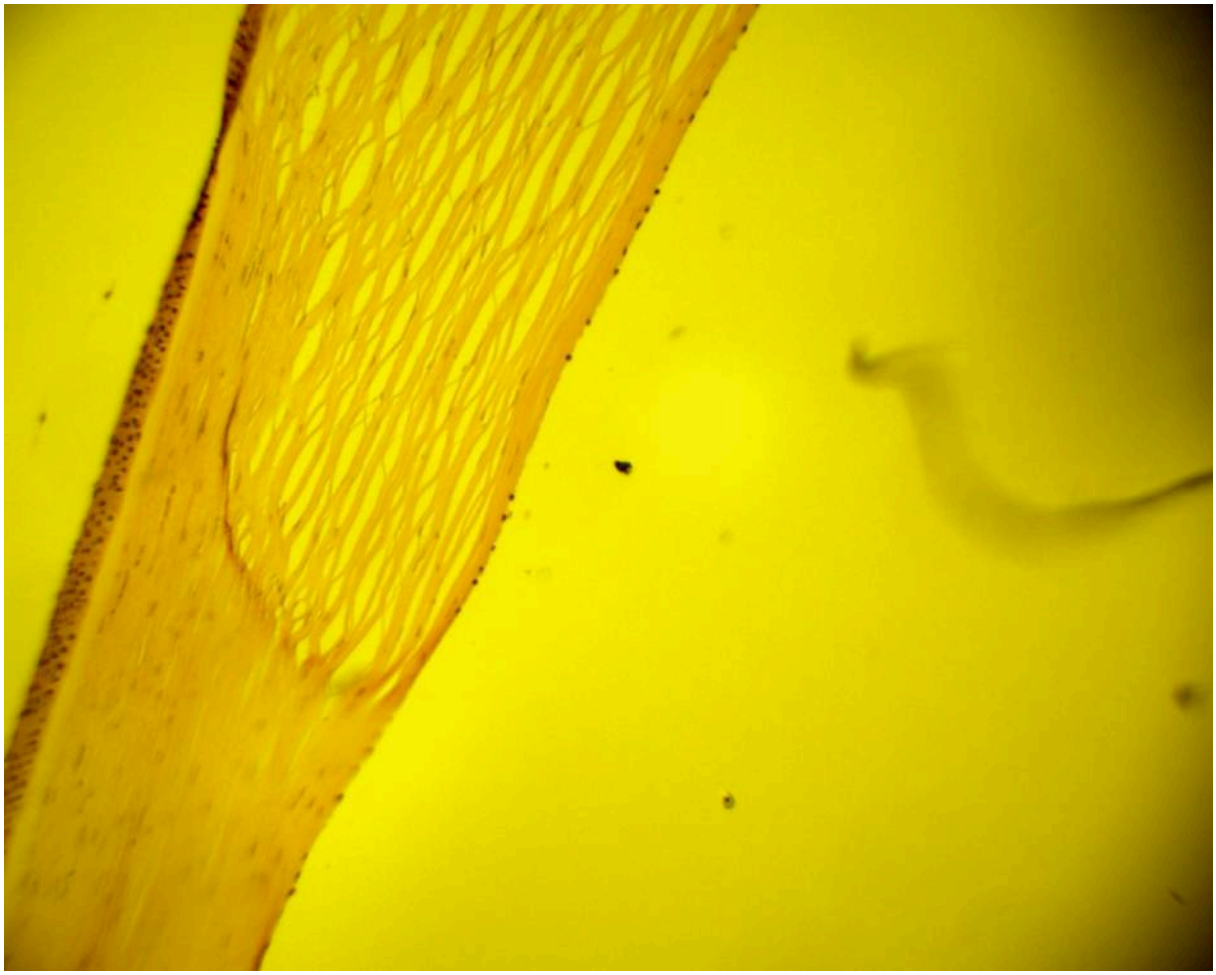

Fig. 4. Histomorphological image of corneal segment, Patient D., 27 years old.

Thinned epithelium, epithelial cells are deformed. Bowman's membrane is strongly deformed. Magnification 140. Stained with haematoxylin-eosin.

Corneal epithelium has few layers, especially closer to optical zone, with numerous deformed and dead cells. Bowman's membrane is deformed throughout, with uneven thickness and density. Corneal stroma contains areas of sparse collagen fibrils.

Picture 4 shows a segment of extremely hydropic stroma with collagen beams split into fibers. Cyst-like areas are observed interspersed with areas of integrity preserved to various extent. Stromal fibroblasts are deformed, in a number of layers apoptotic.

Lower layers of cornea have changes typical of keratoconus.

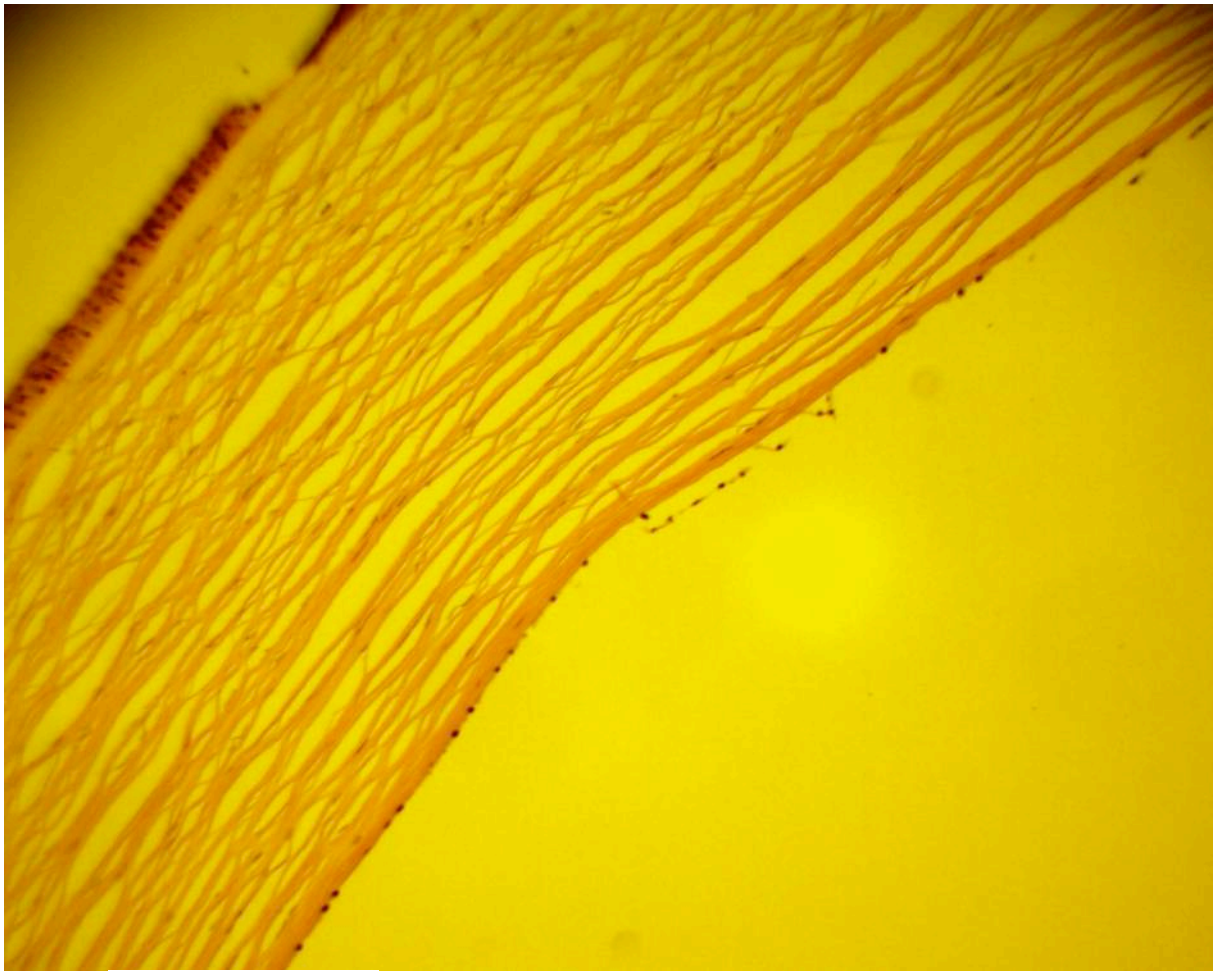

Fig. 5. Histomorphological image of corneal segment, Patient D., 27 years old. Endothelium is largely fragmented throughout, delaminated. Descemet's membrane is strongly deformed, in some places absent. Magnification 240. Stained with haematoxylin-eosin.

The structure of lower layers of cornea underneath the Descemet's membrane is completely destroyed. Endothelium at the preserved areas of the membrane is fragmented, yet mostly absent.

The surgically removed disc of the patient's cornea was morphologically and functionally defective.

Therefore, the morphological studies proved that the surgery was well justified and prompt, since conservative method of arresting the degenerative process in the cornea of the patient with keratoconus would not restore the transparency of the tissue due to gross changes in almost all layers of the cornea.

Acute keratoconus, acute corneal edema, or acute hydrops are uncommon complications for keratoconus. Such conditions are believed to develop in less than 3% of patients, and as a rule, in the eye with better visual functions. The influence of

patients' age or gender on the incidence of complications is not registered, and occurrence of the disease is more likely in residents of South-East Asia [1-2].

Hyperemia of the eye globe in patients with acute keratoconus is presumed to be caused not only by inflammation of the tissue with degenerative changes, but also by presence of *Serratia marcescens* in conjunctival cavity. It is a species of motile asporogenous rod-shaped Gram-negative bacteria that can cause expressed non-inflammatory or weakly inflammatory hyperemia of the eyelid skin, excretory ducts of the meibomian glands and the conjunctiva due to generated red pigment [3]. Besides, of all types of ectasias hydrops is only pathognomonic for primary (essential) keratoconus or secondary corneal ectasias [4].

The pathogenesis of acute keratoconus is based on the rupture of Descemet's membrane and soaking of the corneal stroma with aqueous humor of anterior chamber, which leads to formation of massive, filled with fluid cysts in stroma. The clinical manifestation of this process is emergence of thickened cataract, its area depending on location and size of Descemet's membrane rupture. Subsequently, in the absence of adequate treatment, the cataract vascularizes due to ingrowth vessels of the limbus area, unless the outer corneal tissues are ruptured possibly followed by infection. The reason for initial rupture of Descemet's membrane is unknown. It is assumed to be connected with the immune disorders, particularly, increased titer of IgG and IgE. The studies that prove statistically significant frequency of chronic rhinoconjunctivitis, asthma and eczema also associated with increasing titer of the specified antibodies, in patients with acute keratoconus, implicitly support that assumption [2, 5-6].

In this clinical case no ruptures of Descemet's membrane and endothelium were discovered. Although, pronounced decrease of endothelial cells density (Pic. 5) could result in the changes similar to those present in hydrops.

There are two current approaches to the treatment of acute keratoconus, surgical and conservative. The conservative approach is encouraged by the concept of possible self-induced recovery of Descemet's membrane defect followed by self-maintained dispersion of the cysts. In such cases, patients receive local anti-inflammatory therapy with steroid drops, prostaglandin inhibitors, as well as wearing SCL as a bandage. The literature contains descriptions of successful outcomes of such therapy. However, this

kind of therapy takes about 6-7 months and does not lead to full recovery of visual acuity due to residual corneal opacities [4-6].

Surgical treatment of hydrops involve the use of gas for tamponade of ruptures in Descemet's membrane and endothelium of the cornea from the anterior chamber side. The authors of the method consider plugging of slowly resorbable C_3F_8 gas bubble to be the most effective and rapid treatment. Tamponade can be supplemented by lamellar paracentesis of cornea to accelerate cyst drain. Though it should be noted, that injection of slowly resorbable gas can itself provoke the epithelial-endothelial dystrophy of cornea. Besides, there is histological evidence of specific character of Descemet's membrane ruptures; the edges of membrane curl up in scrolls and fail to unravel independently to seal the defect. Hence it was suggested that back keratoplasty would be reasonable for such patients, although opacity of the cornea serves as a technical obstacle to the surgery [1-3]. It should be particularly emphasized that corneal crosslinking is ineffective in treatment of hydrops due to emergence of expressed morphological changes and extensive stromal thinning (thinner than 400 μ), which may result in injuries of endothelium [7].

In the above case, we decided to conduct a penetrating keratoplasty for several reasons. First, thinning of cornea over the cyst area threatened with the rupture of stroma and corneal fistula. Secondly, extensive corneal cataract implied serious defect of Descemet's membrane, which was unlikely to occlude autonomously. Thirdly, either conservative or palliative treatment infers rather long-term recuperation and offers no guarantee of substantial improvement in visual acuity provided that residual opacities in central optical zone persist. Use of «Material for corneal graft» during penetrating keratoplasty operation has proven effective for elective corneal surgery. The presented clinical case confirms the effectiveness of this method in emergency situations as well.

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