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CASE REPORT

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Secondary glaucoma due to progressive iris atrophy – case report

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ABSTRACT

Iridocorneal endothelial syndrome is a rare disease, the etiology of which is not clearly understood. It usually manifests in only one eye, causing corneal edema, secondary glaucoma, and iris anomalies. Depending on the occurrence of these symptoms, three different variants of the condition can be distinguished: Chandler syndrome, Cogan-Reese syndrome, and progressive iris atrophy.

We present a case report of a female patient with secondary glaucoma due to progressive iris atrophy, who underwent transscleral ciliary body treatment to reduce and stabilize intraocular pressure.

KEY WORDS: ICE, secondary glaucoma, TSCPC, MP-TLT, PIA.

INTRODUCTION

Iridocorneal endothelial syndrome (ICE) is a rare and slowly progressive disorder that may cause vision loss. ICE affects both eyes, but it is most commonly manifested in just one eye [1]. The condition occurs predominantly in women in the third to fifth decades of life [2]. The etiology of ICE is not entirely clear, but recent studies seem to provide evidence for a link with a history of HSV infection [3]. The mechanism of the syndrome involves the acquisition of epithelial features (histological: desmosomes, microvilli, filopodia [4-8] and immunohistochemical: vimentin and cytokeratins [9-11]) by the endothelium, resulting in the proliferation and migration of altered endothelial cells (ICE cells) towards the filtration angle and the iris, where they form a transparent membrane and induce a change in the function and architecture of the anterior chamber of the eye, which in turn causes secondary glaucoma, corneal decompensation, and structural anomalies of the iris. ICE is categorized into three variants:

- Chandler syndrome, characterized predominantly by corneal manifestations such as edema and the associated decline in visual acuity; intraocular pressure is usually normal and iris atrophy is rather mild [12], rarely leading to full-thickness hole formation [13];
- Cogan-Reese syndrome, also referred to as the iris naevus syndrome, is characterized by the presence of multiple iris nodules, which may be pedunculated, and the loss of surrounding crypts [13];
- Progressive iris atrophy presents as pupil displacement (corectopia) and additional pseudopupils (pseudopolycoria) resulting from iris thinning and the stresses induced by the shrinking membrane and peripheral anterior adhesions [14].

The three variants listed above may overlap in one patient [15-17]. Consequently, it may be difficult to identify one of them in the diagnostic process.

CASE REPORT

A 59-year-old woman presented to the A&E Department at the Professor Kornel Gibiński University Clinical Center of the Medical University of Silesia in Katowice (UCK) with diminished visual acuity and pain in the left eye. There was no pain in the right eye. The patient reported a history of "spots" on the iris of the left eye, persisting for many years. A few months before the current consultation, while doing physical exercise, the patient felt pain in her left eye. She also noticed a decrease in visual acuity and what she described as a "rupture" of the iris. After the incident the patient saw a specialist at the local ophthalmology outpatient clinic. At the time of presenting to the UCK, the patient used eye drops with latanoprost in both eyes (Rozaprost, Adamed) 1 × day, eye drops with brimonidine (Biprolast, Adamed) 2 × day in the left eye, eye drops with dorzolamide/timolol (Rozacom, Adamed) 2 × day in the left eye, and she took oral acetazolamide tablets (Diuramid, Polpharma) 1 tablet 2 × day. On examination, visual acuity (Visus) was found to be 1.0 and 0.2 in the right and left eyes, respectively. Intraocular pressure in the right and left eyes was 15 mmHg and 45 mmHg, respectively. The anterior segment of the right eye was normal,

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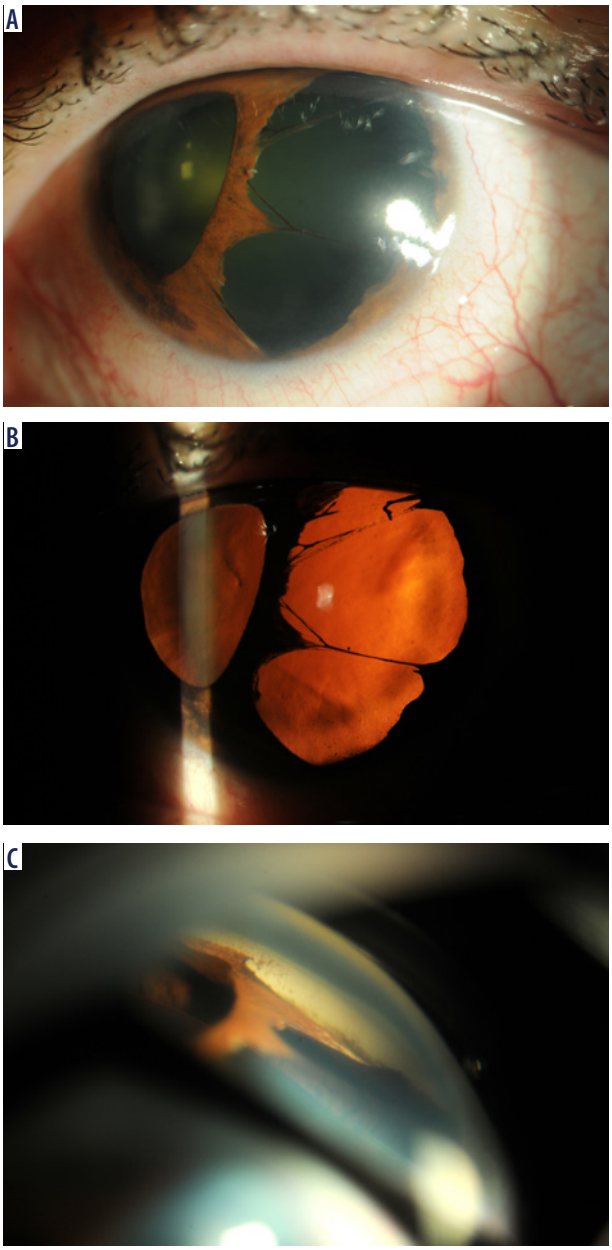

Figure 1. A) Anterior segment of the left eye; **B)** retroillumination of the anterior segment left eye; **C)** angle of filtration of the left eye

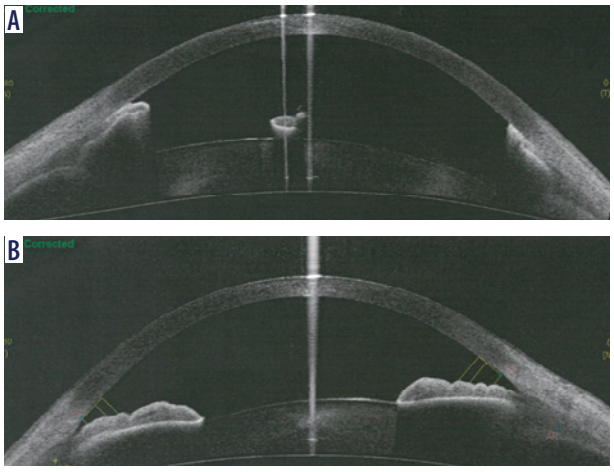

Figure 2. OCT of anterior segment: **A)** left eye; **B)** right eye

without signs of inflammation, with clear natural lens. Findings in the left eye included mixed congestion, transparent cornea, clear anterior chamber of medium depth, corectopia and pseudopolycoria with preserved light reflex, normal convergence and accommodation, incipient cataract, and small floaters in the vitreous chamber. The fundus in both eyes revealed no abnormalities. Gonioscopic evaluation of the right eye showed a wide angle in all quadrants, with a small amount of pigment. No pathological structures were detected. Left eye gonioscopy revealed a narrow angle, closed in sectors by peripheral anterior adhesions, with a moderate amount of pigment (Figure 1). A decision was made to admit the patient to the Ophthalmology Department to reduce and stabilize intraocular pressure in the left eye. During hospitalization, OCT evaluation of the anterior segment was performed, revealing wide angle of filtration in the right eye and filtration angle closure in the left eye (Figure 2).

Based on specular microscopy, the mean endothelial cell count was found to be 2,335/mm^2 and 1,137/mm^2 in the RE and LE, respectively. Field-of-view examination performed by static perimetry yielded the following values: right eye – MS 27.0 dB, MD 0.0 dB, LV 0.9 dB; left eye – MS 4.1 dB, MD 23.0 dB, LV 34.5 dB (Figure 3). Laboratory tests showed no major abnormalities.

The patient underwent MicroPulse Transscleral Laser Therapy (MP-TLT). The procedure was performed in a routine manner, without complications. Intraocular pressure was successfully reduced to 22 mmHg. The patient was discharged home with the following instructions: topical medications to be administered to the left eye: dorzolamide + timolol (Rozacom, Adamed) 2 × day; dorzolamide (Rozalin, Adamed) 1 × day; brimonidine (Briglau Free, Polfa Warszawa) 3 × day, dexamethasone (Dexafree, Thea) 2 × day; and oral acetazolamide (Diuramid, Polpharma) 1/2 tablet 2 × day. In the follow-up examination performed two weeks later, visual acuity (Visus) in the left eye was 0.3, and intraocular pressure was 30 mmHg. The following local findings were observed in the left eye: quiet eye, "beaten-metal" appearance of the corneal endothelium, pseudopolycoria with corectopia, clear anterior chamber without tyndallization, iridescent lens, and preserved fundus view. The patient was readmitted for the procedure of transscleral cyclophotocoagulation (TSCPC). The procedure was performed in a routine manner, without complications. At discharge, the patient's intraocular pressure was 17 mmHg. During subsequent follow-up examinations, intraocular pressure ranged from 12 to 18 mmHg.

DISCUSSION

In the case reported above, the diagnosis of progressive iris atrophy was made on the basis of the patient's sex, age, unilateral presentation of symptoms, absence of significant medical history, morphological features of the anterior segment of the eye and secondary glaucoma, following the exclusion of posterior polymorphous corneal dystrophy [18].

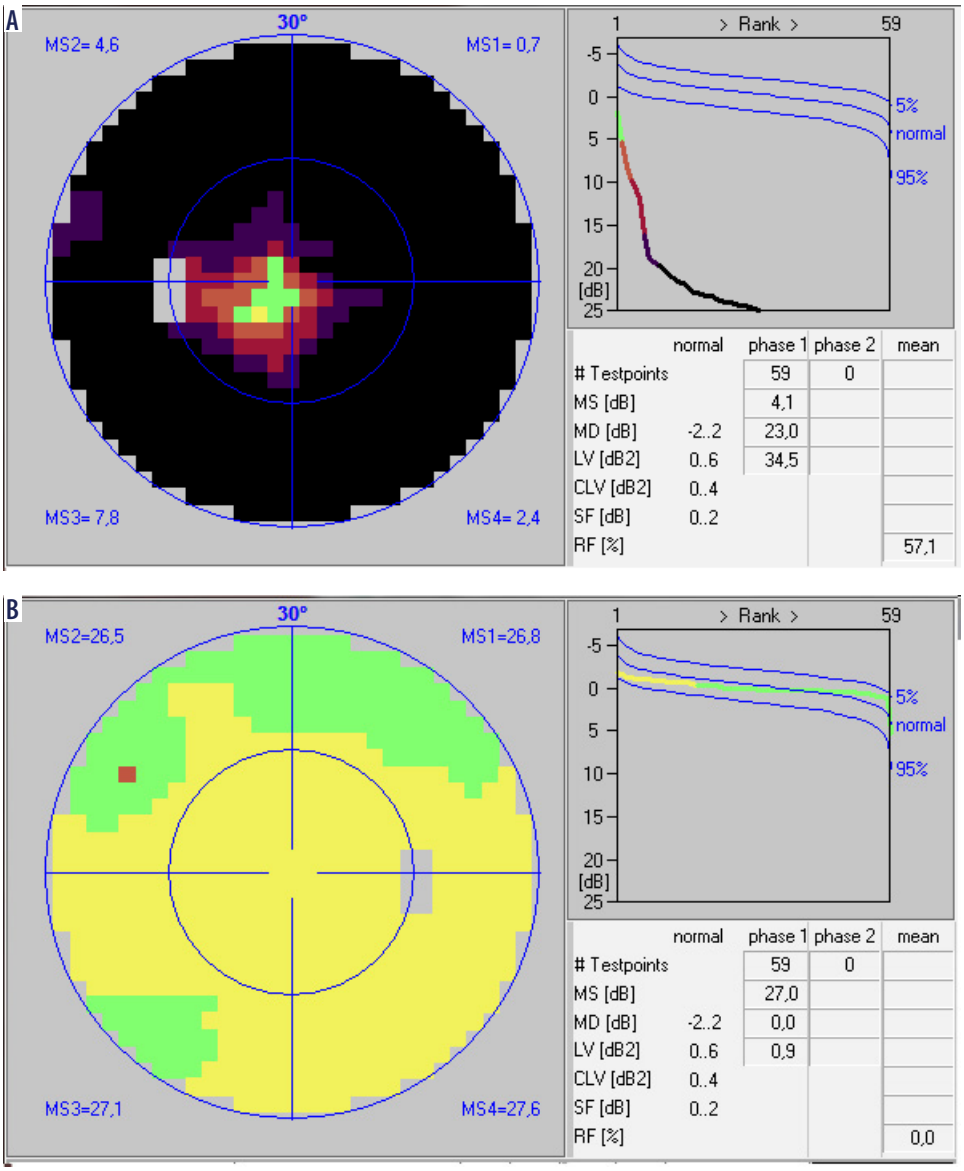

Figure 3. Field of view at first hospitalization: **A)** left eye; **B)** right eye

Secondary glaucoma coexists in approximately 46–82% of ICE cases [19]. Optimization of intraocular pressure in these patients is difficult, and ultimately they often require surgery [3, 13, 20, 21]. Topical treatment is associated with a high failure rate (60–80%). In the literature, there are several retrospective case reports on the surgical treatment of ICE-related glaucoma. The findings suggest that trabeculectomy is less effective in the treatment of glaucoma secondary to ICE than other glaucoma types, with an estimated success rate of 60% in the first year and 40% after two years [19, 22, 23]. Of note, the efficacy of the procedure decreases dramatically to under 20% in cases of revision surgery [22]. Cytostatic agents, especially mitomycin C, have been found to be therapeutically useful [23-25]. The failure of trabeculectomy is due to the pathological mechanism of ICE, namely the migration of ICE cells and overgrowth of the filter pad [26]. Consequently, approximately 12-54% of eyes with ICE that have been treated by trabeculectomy at some point require a secondary surgical intervention [27]. The efficacy of drainage implants is approximately 70% during the first year, and 40-70% after three years, up to 50% after five years [27, 28]. In about 20% of the eyes, endothelial proliferation was found, resulting in the occlusion of the implant lumen [28]. Taking into account the above-mentioned factors and the patient's general clinical condition, we decided against trabeculectomy and instead opted for MP-TLT. The method is relatively new and associated with a lower risk of complications compared to the standard tube shunt surgery. Unlike in conventional TSCPC, where the laser beam is continuous, MP-TLT delivers a series of repetitive short pulses of energy which, combined with the movement of the probe, maintain the temperature of the ciliary body at ca. 35°C, compared to 55°C in the conventional method [29]. The technique does not cause anatomical, UBM [30, 31] or histopathological changes [32],

which reduces inflammatory response and decreases damage to the surrounding tissues. Consequently, the treatment reduces the production of aqueous humor and increases its outflow via both conventional and episcleral routes. The effects of the procedure are considered to be satisfactory if the intraocular pressure is reduced by 30% of the baseline value. The method is very effective both in the treatment of primary and secondary glaucoma, e.g. after keratoplasty or vitreoretinal procedures. Unfortunately, the patient reported here failed to achieve an acceptable outcome after MP-TLT, so a decision was made to perform conventional TSCPC, which ultimately produced a satisfactory result. We found no reports related directly to ICE and cyclophotocoagulation, while published studies describing MP-TLT and conventional TSCPC do not reveal any significant differences, since the proportion of study participants with ICE was too small. Nonetheless, given the mechanism of development of secondary glaucoma in patients with ICE, and the mode of action of TSCPC, as well as the encouraging results of available studies [33, 34], this may be an interesting area for further research on a larger group of patients.

DISCLOSURE

Authors declare no conflict of interest.

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