UVEAL EFFUSION SYNDROME. A CASE REPORT

Konrád A., Penčák M., Veith M., Studený P.

Department of Ophthalmology, University Hospital Královské Vinohrady and 3rd Faculty of Medicine, Charles University Prague, Czech Republic

The authors of the study declare that no conflict of interests exists in the compilation, theme and subsequent publication of this professional communication, and that it is not supported by any pharmaceuticals company. The study has not been submitted to any other journal or printed elsewhere, with the exception of congress abstracts and recommended procedures.

Received: 28 December 2020
Accepted: 7 May 2021
Available on-line: 15 August 2021

MUDr. Aleš Konrád
Oftalmologická klinika
FN Královské Vinohrady
Šrobárova 1150/50
100 34 Praha 10
E-mail: ales.konrad@fnkv.cz

SUMMARY

Purpose: Purpose of this article is to present a case report of a patient with uveal effusion syndrome who underwent deep posterior sclerotomy.
Case report: A 73-year-old patient with unilateral decrease in the best corrected visual acuity, ablation of choroid and secondary retinal detachment in the right eye was admitted to our clinic for examination in November 2017. At the first examination, the best corrected visual acuity in the right eye was 0.3, in the left eye 1.0. Intraocular pressure was 16 mmHg in the right eye and 21 mmHg in the left eye. After performing ultrasound biomicroscopy of the anterior segment (Accutome, Keeler, USA), ultrasound sonography of the affected eye (Accutome, Keeler, USA), magnetic resonance imaging, computed tomography, abdominal ultrasound and blood tests, we concluded the finding as uveal effusion syndrome. We initiated a conservative treatment consisting of oral administration of carbonic anhydrase inhibitor in combination with topical use of prostaglandin analogue. Despite conservative treatment the best corrected visual acuity of the affected eye decreased to 0.05 so we proceeded to a surgical procedure – deep posterior sclerotomy with perioperative scleral sampling for histological examination (detection of glycosaminoglycans in the sclera wall by Alcian blue staining), which was negative. This histological result ranks the patient as the third type of uveal effusion syndrome (ie, non-nanophthalmic with a normal sclera). After the operation both the ablation of choroid and retinal detachment reattached and the best corrected visual acuity in the right eye improved to 0.3. After the subsequent cataract surgery, the ablation of choroid and retinal detachment occurred again, this time with spontaneous recovery. Postoperatively, the best corrected visual acuity in the right eye was 0.5 and at the last check-up at our clinic 0.6.
Conclusion: Deep posterior sclerotomy is a method of choice of surgical treatment for uveal effusion syndrome that does not respond to conservative therapy.
Key words: uvea, uveal effusion syndrome, deep posterior sclerotomy, secondary retinal detachment

Čes. a slov. Oftal., 77, 2021, No.4, p. 202–206

INTRODUCTION

Uveal effusion syndrome (UES) ranks among rare ocular pathologies and is characterised by a leakage of transudate from the choriocapillaries into the suprachoroidal space, where the transudate accumulates and may lead to ablation of the choroid. If the fluid enters the subretinal space, secondary serous retinal detachment may occur. In some cases, UES is linked with nanophthalmos. As a rule, it has a protracted course, with relapses and remissions, and may lead to a reduction of visual acuity [1] as far as blindness, or glaucoma. In treatment both conservative and surgical methods of treatment are used. In our case report we present a patient in whom, following the failure of conservative therapy, an improvement of the finding was achieved after surgical solution by deep posterior sclerotomy.

Uyama et al. [1] divide UES into three types. The first type is UES associated with nanophthalmos. The second type is non-nanophthalmic UES with an abnormal sclera, in which changes in the sclera are characterised by irregular organisation of collagen fibres with accumulation of an extracellular matrix, predominantly glycosaminoglycans. The third type is non-nanophthalmic UES with a normal sclera.

The clinical picture of UES is typified by minimal inflamma-

tory changes, ablation of the choroid, exudative retinal detachment, dilation of the episcleral vessels, blood in the Schlemm's canal and changes of the retinal pigment epithelium with the appearance of "leopard spots". This concerns diagnosis per exclusionem.

Diagnosis of UES is based on criteria according to Uyama et al. [1]: 1) identification of retinal detachment in the lower periphery without the presence of retinal crack. 2) presence of subretinal fluid, shifting with head movement. 3) no demonstration of leakage of fluid from choroidal membrane into subretinal space with the aid of fluorescein angiography. 4) retinal detachment accompanied with flat or circular ablation of the choroid. 5) detectable ora serrata without compression of sclera. 6) exclusion of other causes of choroidal ablation such as reduction of intraocular pressure, intraocular tumour, rhegmatogenous retinal detachment or intraocular inflammation.

The examination methods used in the diagnosis of UES include ophthalmological examination, ultrasonography (USG), ultrasound biomicroscopy of the anterior segment (UBM), optical coherence tomography (OCT) and fluorescein angiography (FAG).

In differential diagnostics of UES it is necessary to consider choroidal melanoma, Vogt-Koyanagi-Harada syndrome, choroiditis, posterior scleritis, central serous retinopathy, hypertension and metastases [2].

CASE REPORT

A seventy-three-year-old patient was sent to Královské Vinohrady University Hospital in November 2017 due to retinal detachment in his right eye (RE), with ablation of the choroid. Subjectively the patient stated a deterioration of best corrected visual acuity (BCVA) in the RE. Upon admittance, BCVA in the RE was 0.3, in the left eye (LE) 1.0. Intraocular pressure (IOP) in the RE was 16 mmHg, LE 21 mmHg. Upon examination on a slit lamp, on the anterior segment (AS) there was an evident pterygium passing to the limbus from the nasal side, with bilateral presence of corticonuclear cataract, otherwise the finding on the AS was physiological. On the ocular fundus of the RE in the lower temporal and nasal quadrant there was balloon buckling retinal detachment extending towards the lower temporal arcades, no crack was perceptible, and the macula was attached. In the centre there was also presence of small yellowish deposits on the ocular fundus, with the character of leopard spots. On the upper periphery of the retina there was presence of ablation of the choroid with maximum in the upper nasal and upper temporal quadrant (Fig.1). The finding on the ocular fundus of the LE was physiological. USG (Accutome, Keeler, USA) of the eyeball of the RE, B scan, showed serous ablation of the choroid, no solid deposit was evident (Fig. 2 A). During hospitalisation an internal examination was conducted, with sampling of tumour markers, which were without a pathological finding. UBM (Accutome, Keeler, USA) was also performed, without demonstration of a solid lesion of the corpus ciliare. Magnetic resonance imaging (MRI) of the head was performed, focusing on the orbit with the use of a contrast substance (Fig. 2 B), in which there was perceptible ablation of the choroid and retinal detachment, without perceptible solid lesions intraocularly and extraocularly, and computer tomography (CT) of the chest and stomach was performed within the framework of oncological screening in order to exclude a primary tumour with metastatic affliction of the eye, with a negative finding. The performed examinations excluded other possible causes of serous retinal detachment and choroidal ablation, and the finding was concluded per exclusionem as UES.

With regard to the patient's good BCVA and the small extent of retinal detachment without affliction of the macula, we embarked upon conservative therapy. According to research of the literature, first of all a prostaglandin analogue was applied to the RE – Xalatan gtt. (Latanoprost 0.005%), with oral administration of a carboanhydrase inhibitor – Diluran tbl. (Acetazolamide) and potassium – KCl tbl. (Kalium chloratum). However, despite the application of conservative therapy the patient experienced a progressive deterioration of BCVA to 0.05, as well as the progression of retinal detachment with detachment of the macula verified on OCT (Spectralis, Heidelberg Engeneering, Germany) (Fig. 3). As a result, we decided to proceed to a surgical solution – deep posterior sclerotomy.

This surgical procedure was performed at our centre in March 2018 according to the schema published by Kong et al. [3]. Peritomy of the conjunctiva was performed during the operation, afterwards 2 scleral flaps of 5 x 2 mm were created in the full thickness of the sclera 7 mm beyond the limbus, with subsequent suture with a single stitch and suture of the conjunctiva (Fig. 4). Perioperatively a sample of the sclera was taken, which was taken for a histological examination. Alcian blue staining used to identify an accumulation of glycosaminoglycans in the sclera was negative. Our case therefore concerned a patient with the third

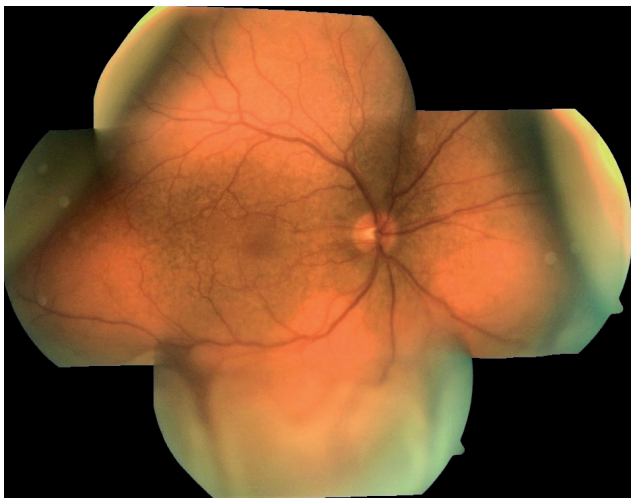

Figure 1. Fundus photo of the right eye, January 2018. Ablation of choroid from above and retinal detachment from below with macula off

type of UES. A video of the posterior deep sclerotomy was made on 16.3.2018 [4].

At the first postoperative follow-up examination in April 2018 there was an improvement of BCVA to 0.3 and an improvement of the finding on the ocular fundus with reattachment of the choroid and persistent serous retinal detachment (Fig. 5). At further follow-up examinations there was an improvement of the finding with complete reattachment of the retina and choroid. BCVA deteriorated to 0.2 with the influence of progression of the cataract. As a result, uncomplicated cataract surgery was performed on both eyes at our centre in January 2019, with implantation of an artificial intraocular lens into the sac. The operations on both eyes were performed on the same day. Postoperative an improvement of BCVA was recorded to 0.5, on the ocular fundus of the RE there was again a finding of minor serous retinal detachment from below with ablation of the choroid, and flat ablation of the choroid was also

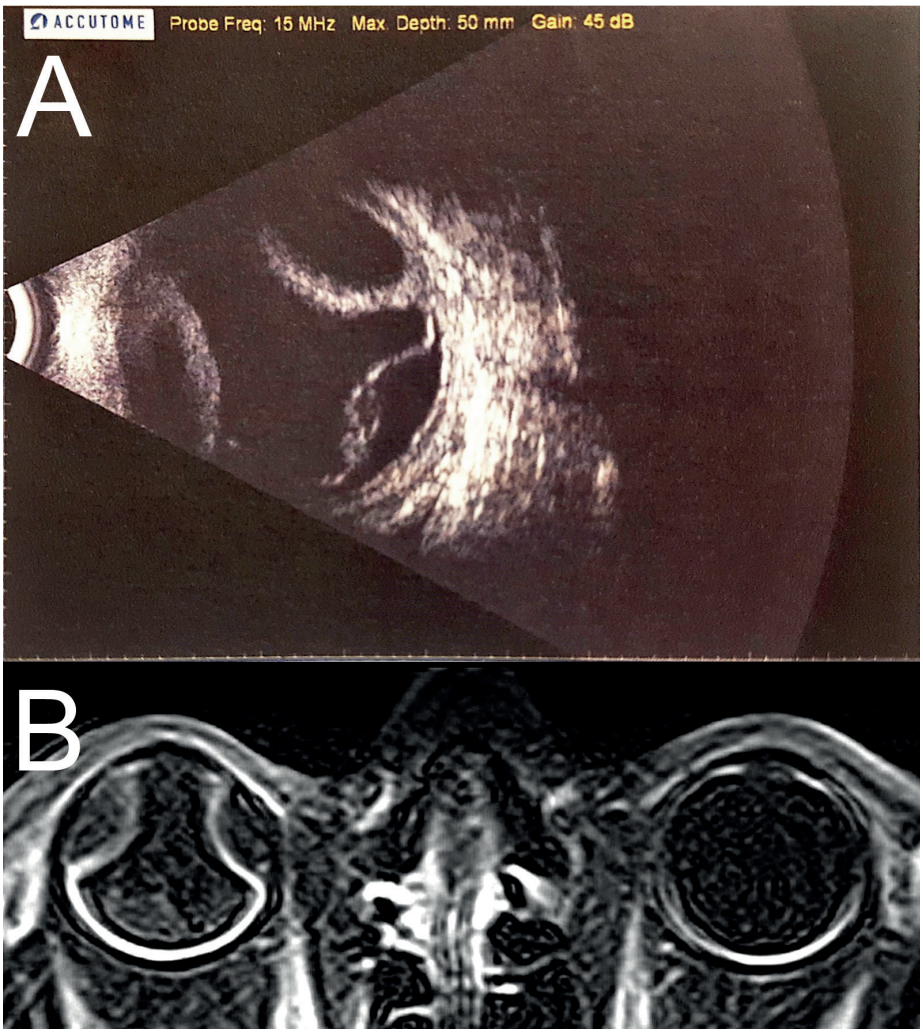

Figure 2. (A) Ultrasound Sonography of the right bulb, B scan, November 2017. Ablation of choroid from above, serous retinal detachment from below. **(B)** Magnetic resonance imaging of the orbits of both eyes, T1- weighted image, November 2017. Ablation of choroid and serous retinal detachment

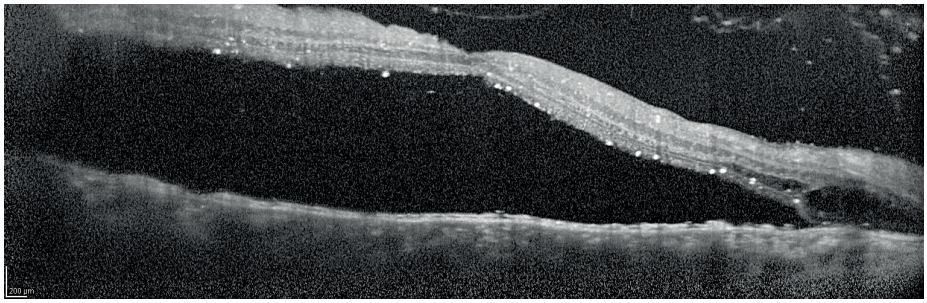

Figure 3. Optical coherence tomography, macula of the right eye, January 2018. Macula off

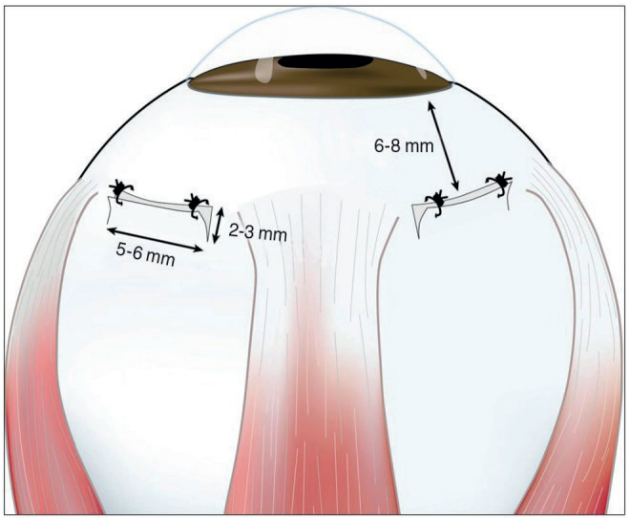

Figure 4. Deep posterior sclerotomy according to a scheme published by Kong et al. [3]. Scheme of the operation with numbers

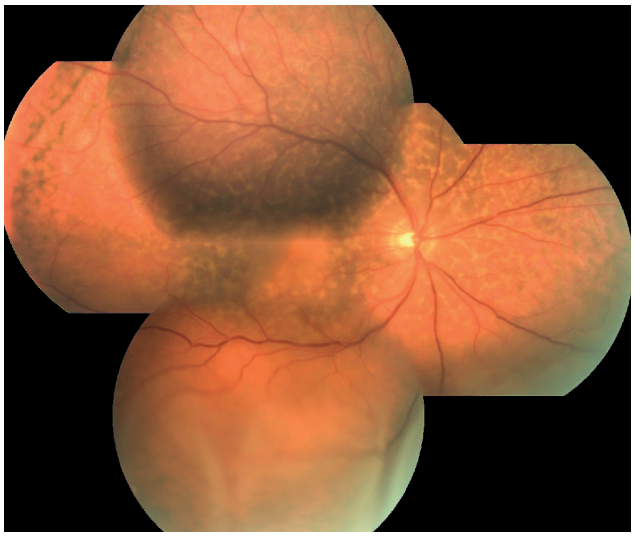

Figure 5. Fundus photo of the right eye after the surgery (deep posterior sclerotomy), April 2018.Without ablation of choroid, persisting serous retinal detachment from below and„leopard spots"

newly recorded in the LE, where BCVA was 1.0. Healing of the finding on the ocular fundus of both eyes occurred spontaneously, without the necessity of a further intervention. At the last follow-up examination at our centre in July 2019, the patient had BCVA in the RE of 0.6 and LE 1.0. IOP in RE was 18 mmHg, LE 12 mmHg. Yellowish deposits with the character of leopard spots persisted on the retina of the RE.

DISCUSSION

Conservative therapy of UES covers oral administration of a carboanhydrase inhibitor in combination with local use of a prostaglandin analogue. The method of choice may be corticosteroid therapy or oral non-steroid anti-inflammatory drug with slow release.

It is possible to consider application of corticosteroid therapy orally, periocularly, topically or in combination in the case of UES with absence of nanophthalmos or abnormality of the thickness of the sclera [5]. In isolated cases, successful therapy with high doses of systemic corticosteroids has also been described [6].

Kumar et al. used an oral non-steroid anti-inflammatory drug (NSAID) with slow release (Indomethacin) and laser photocoagulation of the retina in the treatment of UES. The patients manifested a slight improvement of vision and the finding on the retina, but it is not possible to exclude the possibility that the improvement would have also occurred spontaneously [6].

Another type of local conservative therapy for patients with UES is prostaglandins, about which it is known that they increase scleral macromolecular permeability. UES may be linked with reduced transscleral permeability of albumin, which decreases with age probably as a consequence of exogenous non-enzymatic cross-linking inside the scleral substrate [6]. Moldow et al. [7] stated that acetazolamide stimulates membrane transport of cells forming the blood-retinal barrier, reduces the level of intraocular pressure by an inhibitive effect on the production of intraocular fluid, and also influences other mechanisms of cellular transport and choroidal circulation, which could contribute to a positive clinical response in patients with UES. A combination of topical prostaglandin analogues and acetazolamide led to an improvement of the finding in some patients. However, it was not possible to differentiate which of these two drugs, or indeed spontaneous development of the pathology, was responsible for the final result [8].

In our case conservative therapy had no effect, during this therapy there was a further progression of the finding, and we therefore proceeded to the performance of a surgical procedure – deep posterior sclerotomy.

A number of types of surgical procedures have been described in the literature, the aim of which is (1) direct (internal or external) drainage of subretinal fluid, (2) reduction of scleral resistance, enabling evacuation of fluid from the subarachnoid space, (3) decompression of the vorticose veins or (4) a combination thereof [9].

In 1980 Brockhurst described a surgical method consisting in scleral resection with the release or decompression of the vorticose veins combined with sclerotomy [6]. In 1986 Yue et al. published a report on 2 nanophthalmic eyes with UES successfully treated by this method [10]. However, this surgical technique is highly demanding, with a high risk of complications, above all haemorrhage [11,12], and as a result is no longer used.

The preferred surgical method at present is sclerotomy or sclerectomy without decompression of the vorticose veins, due to its simplicity, manageability and satisfactory result. Various types of these procedures are described in the Czech and foreign literature, for example sclerotomy in the full thickness of the sclera [3,12,13,14], quadrant sclerectomy in partial thickness

of the sclera [15], sclerectomy in the full thickness of the sclera [16] and subscleral sclerectomy [11]. The use of an ExPRESS filtration implant (Alcon, Switzerland) has also been described in order to maintain the patency of sclerotomy or a combination of pars plana vitrectomy with gas tamponade (C3F8), drainage of the subretinal fluid and sclerectomies in the partial thickness of the sclera [9]. Mitomycin C, alleviating the postoperative cicatricial effect, topically applied to the scleral bed, may be a useful supplement in the surgical treatment of UES [17,18,19]. Sclerotomy or sclerectomy may be performed with the assistance of a CO2 laser guided by an optic fibre, which with the precision of incision and simultaneous inducement of haemostasis eases the creation of a scleral flap and thereby averts a frequent complication of this surgical procedure, namely haemorrhage from the highly vascularised uveal tissue [20].

It is therefore possible to state that, in accordance with the foreign literature, the standard and recommended therapy of UES remains surgical treatment. Conservative therapy may be considered as the first therapeutic step, and in case of its failure it is possible to proceed to a surgical procedure.

CONCLUSION

Through the performance of a surgical procedure – deep posterior sclerotomy – in a patient with uveal effusion syndrome, we attained a subjective and objective improvement in the operated eye, as well as a correction of the intraocular finding. Deep posterior sclerotomy is an effective and safe method of surgical treatment of UES.

LITERATURE

1. Chan W, Fang-tian D, Hua Z, You-xin C, Rong-ping D, Ke T. Diagnosis and treatment of uveal effusion syndrome: a case series and literature review. Chin Med Sci J. 2011;26(4):231-236.
2. Palestine A, Cao J, Tripathy K, Skvortsova NA. Uveal Effusion Syndrome. American Academy of Ophthalmology. Available from: https://eyewiki.aao.org/Uveal_effusion_syndrome
3. Kong M, Kim JH, Kim SJ, Kang SW. Full-thickness sclerotomy for uveal effusion syndrome. Korean J Ophthalmol. 2013;27(4):294-298.
4. https://youtu.be/B4Ltyg6XV6I
5. Shields CL, Roelofs K, Di Nicola M, Sioufi K, Mashayekhi A, Shields JA. Uveal effusion syndrome in 104 eyes: Response to corticosteroids – The 2017 Axel C. Hansen lecture. Indian J Ophthalmol. 2017;65(11):1093-1104.
6. Elagouz M, Stanescu-Segall D, Jackson TL. Uveal effusion syndrome. Surv Ophthalmol. 2010;55(2):134-145.
7. Moldow B, Sander B, Larsen M, Lund-Andersen H. Effects of acetazolamide on passive and active transport of fluorescein across the normal BRB. Invest Ophthalmol Vis Sci. 1999;40(8):1770-1775.
8. Derk BA, Benčić G, Corluka V, Geber MZ, Vatavuk Z. Medical therapy for uveal effusion syndrome. Eye (Lond). 2014;28(8):1028-1031.
9. Bodnar Z, Mruthyunjaya P. Management of Uveal Effusion Syndome. Retina Today. 2018 September. Available from: https://retinatoday.com/articles/2018-sept/management-of-uveal-effusion--syndrome
10. Yue BYJT, Duvall J, Goldberg MF, Puck A, Tso MOM, Sugar J. Nanophthalmic sclera. Morphologic and tissue culture studies. Ophthalmology. 1986;93(4):534-541.
11. Uyama M, Takahashi K, Kozaki J, et al. Uveal effusion syndrome: clinical features, surgical treatment, histologic examination of the sclera, and pathophysiology. Ophthalmology. 2000;107(3):441-449.
12. Gass JDM, Jallow S. Idiopathic serous detachment of the choroid, ciliary body, and retina (uveal effusion syndrome). Ophthalmology. 1982;89:1018-1032.
13. Říhová E, Boguszáková J, Gajdošíková Z, Šišková A, Šplíchal L. Syndrom idiopatické uveální efuze [Idiopathic Uveal Effusion Syndrome]. Cesk Slov Oftalmol. 2000;(1):11-17. Czech.
14. Šišková A, Říhová E, Dotřelová D, Adam P. Perforující pars plana sklerotomie u syndromu idiopatické uveální efúze [Perforating Pars Plana Sclerotomy in Idiopathic Uveal Effusion Syndrome]. Cesk Slov Oftalmol. 2005;(3):172-178. Czech.
15. Johnson MW, Gass JDM. Surgical management of the idiopathic uveal effusion syndrome. Ophthalmology. 1990;97:778-785.
16. Jackson TL, Hussain A, Morley AMS, et al. Scleral hydraulic conductivity and macromolecular diffusion in patients with uveal effusion syndrome. Invest Ophthalmol Vis Sci. 2008;49(11):5033-5040.
17. Sabrosa NA, Smith HB, MacLaren RE. Scleral punch method with topical mitomycin C for safe revision of failed deep sclerectomy in nanophthalmic uveal effusion syndrome. Graefes Arch Clin Exp Ophthalmol. 2009;247(7):999-1001.
18. Akduman L, Adelberg DA, Del Priore LV. Nanophthalmic uveal effusion managed with scleral windows and topical mitomycin-C. Ophthalmic Surg Lasers. 1997;28(4):325-327.
19. Suzuki Y, Nishina S, Azuma N. Scleral window surgery and topical mitomycin C for nanophthalmic uveal effusion complicated by renal failure: case report. Graefes Arch Clin Exp Ophthalmol. 2007;245(5):755-757.
20. Bausili MM, Raja H, Kotowski J, et al. Use of fiberoptic-guided CO2 laser in the treatment of uveal effusion. Retin Cases Brief Rep. 2017;11(3):191-194.