Caso Clínico

Type I Uveal Effusion Syndrome: A case report

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RESUMO

Introdução: O síndrome de efusão uveal (SEU) envolve um raro e idiopático descolamento exsudativo da retina, mais provavelmente causado por alterações esclerais. O tratamento pode ser desafiante devido ao curso frequentemente recorrente da doença e com fraca resposta ao tratamento. Os autores apresentam uma caso de SEU tipo I bilateral, resolvido com sucesso cirurgicamente.
Métodos: Os autores apresentam um caso de um homem de 61 anos com antecedentes de hipermetropia elevada bilateral desde a infância e nanoftalmia. Ao exame oftalmológico observou-se descolamente bilateral exsudativo da retina.
Resultados: O doente foi tratado medicamente e, uma vez que não foi obtida resposta, foi submetido a cirurgia que incluiu realização de *flaps* esclerais inferonasais e inferotemporais e esclerectomia bilateralmente. Trinta meses após a primeira cirurgia no olho direito e seis meses após cirurgia no olho esquerdo o doente apresentou uma acuidade visual de 20/100 e resolução do descolamento de retina em ambos os olhos.
Conclusão: O caso apresentado salienta a importância da abordagem cirúrgica em alguns casos de SEU, frequentemente refractários à terapêutica médica. O objectivo da cirurgia é reduzir a resistência ao fluxo de saída coroideu, facilitando a drenagem e resolução do descolamento de retina.

Palavras-chave
Síndrome de efusão uveal (SEU), descolamento de retina exsudativo, nanoftalmia.

ABSTRACT

Introduction: Uveal effusion syndrome (UES) is a rare idiopathic serous detachment of the retina, most probably caused by scleral abnormality. Treatment may be challenging due to the frequent relapsing course and unresponsiveness of the disease. The authors report a case of a type I bilateral UES, successfully resolved with surgery.
Methods: The authors report a case of a 61-year-old man with a history of high bilateral hyperopia since childhood and nanophthalmic eyes. On examination bilateral exudative retinal detachment was evident.
Results: Patient was treated medically and since there was no clinical response, underwent bilateral inferonasal and inferotemporal scleral flaps and full-thicknes sclerectomy. Thirty months after the first surgery on the right eye and six months after surgery on the left eye, patient had a visual acuity of 20/100 and resolution of the retinal detachment on both eyes.
Conclusion: The case reported highlights the importance of surgical approach in some cases of UES, which are frequently refractory to medical therapy. Surgical intervention aimed to decrease the overall resistance to choroidal fluid outflow, facilitating drainage and resolution of retinal detachment.

Keywords
Uveal effusion syndrome (UES), exudative retinal detachment, nanophthalmos.

INTRODUCTION

Uveal effusion syndrome (UES) is an idiopathic serous detachment of the retina, most probably caused by scleral abnormality. The increased thickness of the latter impedes transcleral intraocular fluid outflow and compresses the vortex vein, leading to congestion of the choroidal veins. Intraocular fluid then accumulates in the choroid leading to ciliochoroidal detachment.[1] This may also be associated with fluid in the subretinal space, resulting in secondary serous retinal detachment.[2] Ciliochoroidal detachment caused by hypotony, intraocular inflammation and intraocular tumor should be excluded from uveal effusion syndrome, as forms of secondary ciliochoroidal detachments.[1]

Idiopathic UES has a male preponderance and a relapsing-remitting clinical course. Often there is a severe visual loss due to chronic submacular fluid and secondary retinal pigment epithelium (RPE) changes described as a leopard spot fundus. Approximately half of the patients who require surgical treatment in one eye eventually require surgery in the second eye and some involvement of both eyes occurs in a majority of patients (up to 65% of all cases). The incidence of UES is not reported, but it is undoubtedly an extremely rare disease.[2]

Several surgery procedures have been proposed and include vortex vein decompression, sclerotomy, partial or full-thickness sclerectomy, ultrasound-guided sclerotomy or ExPRESS shunt for choroidal fluid drainage.[2,3,4,5] Medical therapy have been described as a possible first step before surgical approach and include topical prostaglandins analogs and/or oral carbonic anhydrase inhibitors.[2,6] Despite treatment this disease often follows a relapsing course.[7]

CASE REPORT

The authors report a case of a 61-year-old man who presented to our clinic with decreased vision in both eyes for years. He had a history of high hyperopia since childhood, with +19D in the right eye (RE) and of +18D in his left eye (LE). Ophthalmological examination revealed visual acuity (VA) of 20/200 bilaterally, intraocular pressure of 18 and 17 mmHg in his RE and LE respectively, shallow anterior chamber bilaterally and on fundoscopy exudative detachment involving the macula on both eyes, as shown in Figure 1.

B-scan ultrasonography demonstrated a thickened sclera, axial length of 14mm in the RE and 15mm in the LE and confirmed bilateral retinal detachment (Figure 2).

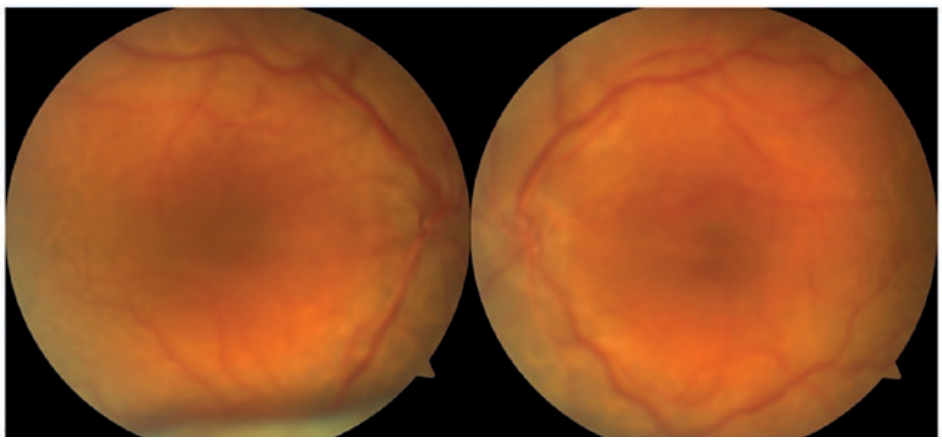

Fig. 1 | Retinography showing exudative detachment on both eyes.

Patients with UES are divided into three groups: type I, nanophthalmic eye, axial length of the eyeball of less than 19.0mm, high grade of hyperopia in refraction and, at the time of surgery, the sclera is rigid and thick; type II, non-nanophthalmic eye with abnormal sclera, axial length of the eyeball is not small, there are no remarkable refractive errors, but the sclera is rigid and thick; type III, non-nanophthalmic eye with normal sclera, normal size eyeball and the sclera has a normal thickness.[1]

Macular optical coherence tomography (OCT) confirmed a thickened sclera and retinal detachment. Initially the patient was treated with oral corticosteroids and oral carbonic anhydrase inhibitors, with no clinical response. Six months later, it was decided to proceed to surgery in his LE.

Patient underwent sclerectomy and sclerotomy procedure. At the equator of the inferotemporal and inferonasal

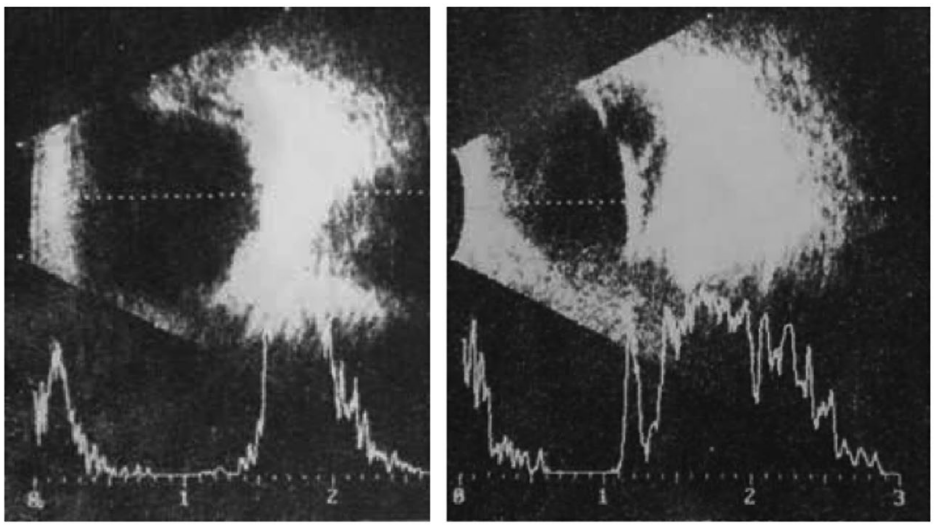

Fig. 2 | Ocular ultrasonography showing nanophthalmic eyes and retinal detachment.

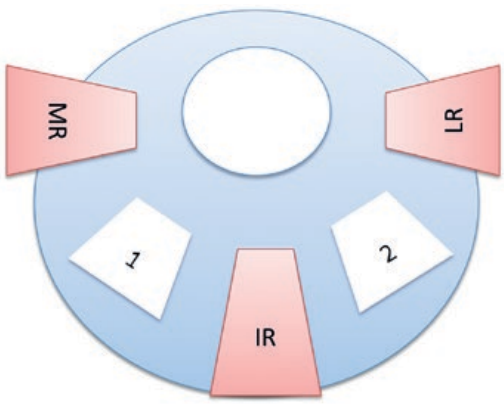

Fig. 3 | Graphic representation of the surgical procedure performed on both eyes. MR, medial rectus muscle; IR, inferior rectus muscle; LR, lateral rectus muscle; 1 and 2, inferotemporal and inferonasal flaps.

quadrants the authors performed a two-thirds thickness scleral flap measuring 4mm x 5mm. Under the scleral flap, the remaining thickness of the sclera was excised in pieces measuring 3mm x 4mm and the choroid was exposed (Figure 3). The scleral flap was not sutured, but Tenon's capsule and the bulbar conjunctiva were closed tightly.

The surgery was uncomplicated and during follow-up there was a slow but significant improvement in the subretinal fluid resolution. Six months after surgery, the choroidal effusions disappeared and VA improved to 20/100 in the LE. Two years after the initial surgery, patient underwent the same procedure on his RE. The results were similar with resolution of the retinal detachment and VA improvement to 20/100 within 6 months (Figure 4).

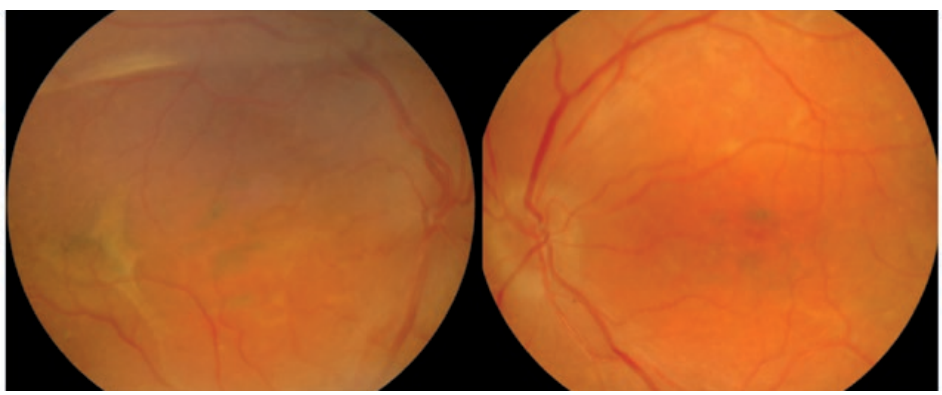

Fig. 4 | Postoperative retinography showing resolution of retinal detachment bilaterally and leopard skin spots involving both foveas.

CONCLUSION

Effective surgical results are described when sclerotomy and sclerectomy are performed in type I and II UES, since the abnormal sclera and increased resistance to the transcleral outflow of intraocular fluid are thought to be the main causes of these disorders. However this technique was not effective in type III UES, which develops in non-nanophthalmic eyes with normal eyeball size and normal scleral thickness.[7]

The authors present a case of a patient with type I bilateral UES, initially treated with medical therapy. Since retinal exudative detachments where not responsive to conservative treatment, patient underwent inferonasal and inferotemporal scleral flaps with a full thickness sclerectomy on both eyes. Follow-up of 30 months and 6 months after surgery respectively on his RE and LE revealed anatomical good results. Despite that, functional improvement may be considered modest, with final visual acuity of 20/100 on both eyes. The reason is that after a long period of serous retinal detachment, photoreceptor and retinal pigment epithelial damage may lead to pigmentary changes, described as leopard skin spots, with permanent visual loss.

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