Original Article

Clinical presentation and treatment outcomes of an algorithmic approach to uveal effusion syndrome

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Purpose: To describe the clinical features and treatment outcomes in spontaneous uveal effusion syndrome (UES). **Methods:** A 10-year retrospective chart review of UES patients from a tertiary eye center was carried out. Optical coherence tomography (OCT), fundus fluorescein angiography, and ultrasound biomicroscopy (UBM) scans were performed. UES was managed based on presenting best-corrected visual acuity (BCVA), symptoms, and fundus findings. Patients with secondary causes of uveal effusion were excluded. **Results:** Twenty-five eyes of 16 patients were included. Of the 16 patients, 14 (88%) were male and 9 (56%) had bilateral disease. Fifteen of 25 affected eyes had nanophthalmos (axial length (AL) <20.5 mm) and 6 had hyperopia with AL >20.5 mm. The presenting mean distance BCVA was 0.74 ± 0.64 logMAR (mean Snellen: 20/100). Eleven eyes had exudative retinal detachment, and 4 also had exudative choroidal detachment (CD). Choroidal thickness (CT) was increased in 11 eyes on B-scan ultrasonography, and the mean CT was 1.74 ± 0.38 mm. Sub-retinal fluid (SRF) and retinal folds were the most common OCT findings. UBM findings included shallow angles, peripheral CD, and supra-ciliary effusion. A combination of local and systemic corticosteroids was used to successfully treat 12 eyes, 6 needed surgery, and 7 were observed. Partial sclerectomy with anterior chamber maintainer-assisted SRF drainage was the favored surgery. The median period of follow-up was 6.5 months (0.1–76 months), and the mean distance BCVA at the last follow-up was 0.58 ± 0.42 logMAR (mean Snellen: 20/80). **Conclusion:** UES can be suitably managed both medically and surgically based on clinical presentation.

Key words: Anterior chamber maintainer, exudative RD, ocular ultrasonography, sclerectomy, sub-retinal fluid drainage, uveal effusion syndrome, UES

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Uveal effusion syndrome (UES) is a rare ocular disease that most commonly occurs in otherwise healthy middle-aged males.[1] A study from the United Kingdom reported the estimated annual incidence of UES as 1.2 per 10,000,000 population.[2] UES is characterized by fluid collection in the supra-choroidal space, leading to choroidal detachment (CD), secondary exudative retinal detachment (ERD), and ultimately degeneration of the retinal pigment epithelium (RPE) with vision loss in chronic cases.[3] Though there are several causes of uveal effusion, the term UES is reserved for idiopathic cases where the cause of uveal effusion is unknown. It is a diagnosis of exclusion.[3,4] UES has been further classified as type-1: associated with nanophthalmos, type-2: in non-nanophthalmic eyes with an abnormal sclera, and type-3: in non-nanophthalmic eyes with a normal sclera.[5]

The pathogenesis of fluid accumulation in UES is multifactorial and occurs due to a combination of vortex vein compression due to congenital scleral thickening, reduced scleral protein permeability due to abnormal deposition of glycosaminoglycan (GAG) like material in the sclera, altered scleral hydraulic conductivity, primary chronic hypotony, and increased choroidal permeability.[6-11] The relative contribution of each factor can be variable.

No consensus exists on the management strategy for UES in previous literature and different medical and surgical approaches have been attempted with variable success. Whereas the association of UES with nanophthalmos is well known, the diagnostic criteria for nanophthalmos vary with some even considering all eyes with an axial length less than 21 mm to be potentially nanophthalmic.[3] There are limited case series reported on this rare disease and even fewer cases reported from South Asia. It is also known that the ocular dimensions of South-Asian eyes are smaller than those of Caucasian eyes.[12] Our study aims to describe the clinical features and treatment outcomes in UES in a South-Asian population and compare these findings to what has been reported in different populations. We also describe a previously

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unreported surgical technique of partial (deep) sclerectomy with anterior chamber maintainer (ACM)-assisted sub-retinal fluid (SRF) drainage for the surgical management of UES.

Methods

This retrospective observational non-comparative study was conducted at a tertiary eye-care center in India after approval by the institute ethics committee (LEC-BHR-R-05-22-874). Medical records of patients of any age diagnosed to have UES from 2010 to 2020, that is, over 10 years, were accessed from the electronic medical records and institute medical records database. Demographic and clinical data of these patients were compiled and analyzed. Ocular ultrasonography (USG) [Accutome Inc., Malvern, PA, USA], ultrasound biomicroscopy (UBM) [Accutome Inc., Malvern, PA, USA], optical coherence tomography (OCT) [Cirrus HD-OCT, Carl Zeiss Meditec, Dublin, CA and DRI OCT Triton, Topcon, Tokyo, Japan], fundus fluorescein angiography (FFA) [Heidelberg HRA2, Heidelberg Engineering, Inc, Vista, CA], and optical biometry findings [IOL Master 500, Carl Zeiss Meditec, Jena, Germany] were studied. The management strategies used to treat these patients were studied.

The criteria for patient selection included the presence of peripheral ciliochoroidal/CD and/or increased choroidal thickness (CT), with or without associated ERD in at least one eye. Only patients without a clearly identifiable cause for the CD or increased CT were included. Ciliochoroidal detachment was detected either during dilated fundus examination on indirect ophthalmoscopy or on B-scan-USG or UBM of the eye. The choroid was said to be thickened if the CT exceeded 1.7 mm on USG as measured inferior to the optic disc.[13] Patients with intra-ocular inflammation (trauma, iatrogenic, and posterior uveitis), hypotony, medication intake, rhegmatogenous retinal detachment, Hunters syndrome, carotid-cavernous fistula, or uveal melanoma-induced uveal effusion were excluded.

Included patients were sub-divided into those with nanophthalmos and those without nanophthalmos. Patients with an axial length of less than 20.5 mm with high hyperopia (spherical equivalent of more than +7D) and/or a shallow central anterior chamber (less than 2.2 mm) with increased CT (>1.7 mm) were classified to have nanophthalmos.[13-15]

Partial-thickness (deep) sclerectomy in all four quadrants with ACM-assisted SRF drainage without vortex vein decompression was the preferred surgical technique in our series. The technique is described as follows: After creating a scleral window with partial-thickness sclerectomy using a crescent blade at a distance of 4 mm from the ora on the sclera, an ACM is secured into the anterior chamber. The infusion is then started at 30 mm Hg (high pressure should be avoided as it can result in lens subluxation). A partial-thickness scleral flap is raised and SRF is drained with the help of a 26-gauge needle or the sharp tip of a 7-0 vicryl suture needle [Fig. 1]. SRF drainage is performed in the quadrant in which the ACM is inserted to avoid touching the lens.

Results

Demographic and baseline ocular characteristics and dimensions [Table 1]

Twenty-five eyes of sixteen patients met the inclusion criteria of the study. Fifteen UES-affected eyes (60%) had nanophthalmos. The patient demographic and baseline ocular characteristics and dimensions of UES eyes are elaborated upon in Table 1.

Clinical features at presentation

Twelve of sixteen patients (75%) presented with a diminution of vision (DOV) in the affected eye. Of these, 6 had bilateral disease, that is, 18 of the 25 (72%) affected eyes presented with DOV. The mean duration of DOV was 3.22 ± 3.95 months. The other 7 eyes of 4 patients (3 bilateral UES) were diagnosed with UES at the time of a general eye check-up and did not complain of vision loss at presentation.

Eleven eyes in a total of the twenty-five included eyes (44%) had ERD. Four of these had co-existent CD. The other 14 eyes (56%) had no ERD. However, these were found to have retinal and/or choroidal folds, retinal pigmentary changes, cystoid macular edema (CME), and even a sub-retinal band in one case [Fig. 2].

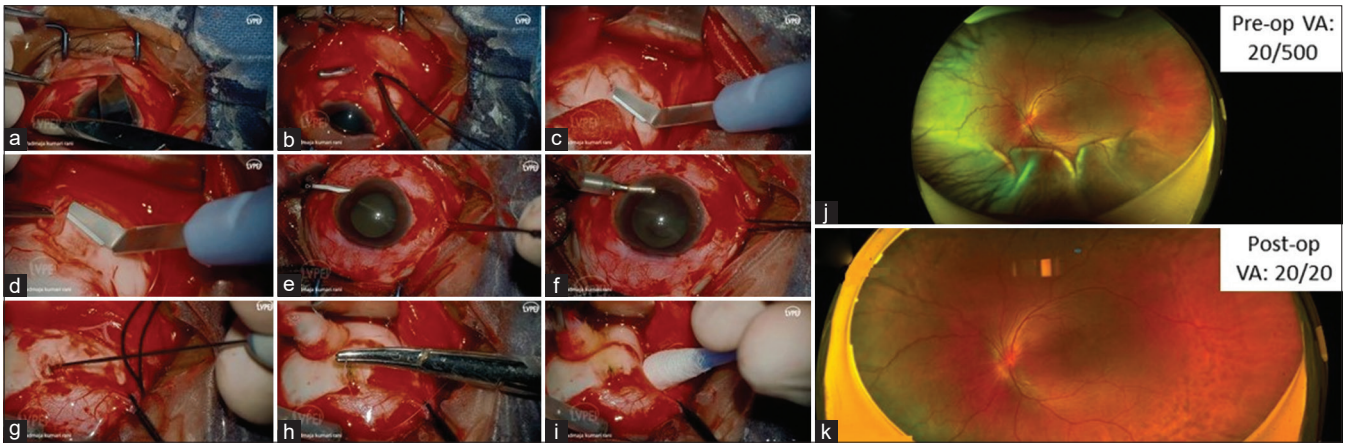

Figure 1: Partial-thickness (deep) sclerectomy with ACM-assisted SRF drainage (a) Peritomy and Tenon's capsule dissection with peritomy scissors, (b) inferior rectus hooked and tagged with muscle hook and 4-0 silk suture, (c and d) scleral window with partial-thickness sclerectomy using crescent blade, (e) entry into the anterior chamber with a MVR blade, (f) ACM in the anterior chamber, (g) sclerotomy with a 26G needle, (h) SRF drained with a sharp tip of 7-0 vicryl suture, (i) pressure on the globe with a sterile cotton bud, (j and k) pre- and post-operative color fundus photographs

Table 1: Summary of demographic and baseline ocular characteristics and dimensions of UES patients

Patient demographics	Summary
Patients recruited for the study (*n*)	16
Gender (Male/Female)	14/2 (87.5%/12.5%)
Age (years) *	43.25±14.77 years (13-70 years)
Laterality (UL/BL)	7/9 (43.75%/56.25%)
UES eyes	
Total eyes affected	25
Eye affected (Right/Left)	15/10 (60%/40%)
Distance BCVA (LogMAR) (*n*=25) *	0.74±0.64 (0.00-3)
Near BCVA (LogMAR) (*n*=23) *	0.78±0.3 (0.4-1.3)
Refractive error [Mean spherical equivalent] (dioptres) (*n*=24) *	6.29±6.77 (−3.25 - +15.25)
Myopia (mean spherical equivalent ≥ −1D)	4/24 (16.7%)
Low myopia: 3/4, moderate myopia: 1/4 {low myopia: −1 to−3D, moderate myopia−3D to−6D}	
Hyperopia (mean spherical equivalent ≥+1D)	18/24 (75%)
Hyperopia: 18/24 (75%)—low: 4/18, moderate: 2/18, high: 12/18 (66.6%) {low hyperopia: 1 to 2D, moderate myopia 2D to 5D, high hyperopia: >5D}	
Emmetropia	2/24 (8.3%)
IOP (mm Hg) (*n*=23) *#	16.65±10.96 (10–51)
Ocular dimensions of UES eyes^	
Corneal diameter (WTW) (mm) (*n*=12) *	11.55±0.66
Corneal thickness (CCT) (microns) (*n*=7) *	527.42±27.32
Anterior chamber depth (Endo ACD/aqueous depth) (mm) (*n*=13) *	2.48±0.46
Shallow AC (i.e. ACD<2.2 mm) - 3/13 (23.07%)	
Lens thickness (mm) (*n*=12) *	4.49±0.47
Axial length (mm) (*n*=21) *	18.9±3.18

Nanophthalmos (AL<20.5 mm, high hyperopia or shallow AC with increased CT>1.7 mm)—15/21 (71.4%). *expressed in terms of (mean±SD) (Range). #IOP as recorded by Goldman applanation tonometry, IOP was not recorded for 2 eyes but was noted to be digitally normal. ^ Ocular dimensions as measured by optical biometry except for axial length measured by A-scan ultrasonography

UES with nanophthalmos

Fifteen UES-affected eyes of nine patients had nanophthalmos (3 unilateral and 6 bilateral UES). Among these, 6 (40%) had ERD, of which 2 had co-existent CD. The other 9 (60%) eyes had an attached retina. Among these, 4 had retinal folds only, 1 had both retinal and choroidal folds along with OCT confirmed CME, 2 had retinal pigmentary changes, 1 had a sub-retinal band, and 1 had no other obvious retinal findings but had shallow angles on UBM with increased CT on USG. Vitreous cells were found in 2 eyes.

UES without nanophthalmos

Among the 10 eyes of 7 patients without nanophthalmos (4 unilateral and 3 bilateral UES), 5 (50%) eyes had ERD, of which 2 had co-existent CD. The other 5 eyes had an attached retina, and of these, one had retinal pigmentary changes. Vitreous cells were found in 2 eyes.

Investigations and imaging at presentation

Additional ophthalmic investigations [Fig. 3] were performed at presentation when necessary to help confirm the diagnosis of UES and exclude other causes of secondary uveal effusion.

USG

USG was performed in 17 affected eyes of 11 patients. Eleven eyes had increased CT (>1.7 mm); of these, 4 had ERD, of which 1 had co-existent CD. CT was found to be normal in 6 eyes; of these, 2 had ERD, of which 1 had co-existent CD. The overall mean CT was found to be 1.74 ± 0.38 mm and ranged from 1.2 to 2.4 mm.

OCT

OCT was performed for 17 affected eyes of 11 patients (6 bilateral UES). OCT findings included SRF, retinal folds, choroidal folds, sub-retinal hyperreflectivity, CME, retinal schisis, and RPE abnormalities. These findings were found to be either isolated or combined.

SRF and retinal folds were the most common OCT findings and were seen in 8/17 (47%) eyes each. Choroidal folds were seen in 7/17 (41%) eyes. Retinal folds without choroidal folds were seen in 5 eyes, of which 2 had associated SRF. Choroidal folds without retinal folds were seen in 4 eyes, all of which had associated SRF. Both retinal and choroidal folds were found in 3 eyes, none of which had SRF. The 2 other eyes with SRF had sub-retinal hyperreflectivity. The mean sub-foveal choroidal thickness (SFCT) was found to be 354.2 ± 49 microns and ranged from 311 to 459 microns.

FFA

FFA was performed for 13 affected eyes of 8 patients (5 bilateral UES). Diffuse pinpoint and patchy areas of hyperfluorescence due to window defects were the most common FFA findings seen in 8 eyes. In one patient hyperfluorescence due to leakage at the site of ERD was seen. In one of the patients with bilateral disease, no active leakage or staining was seen, and in another, the scan quality was too poor to interpret.

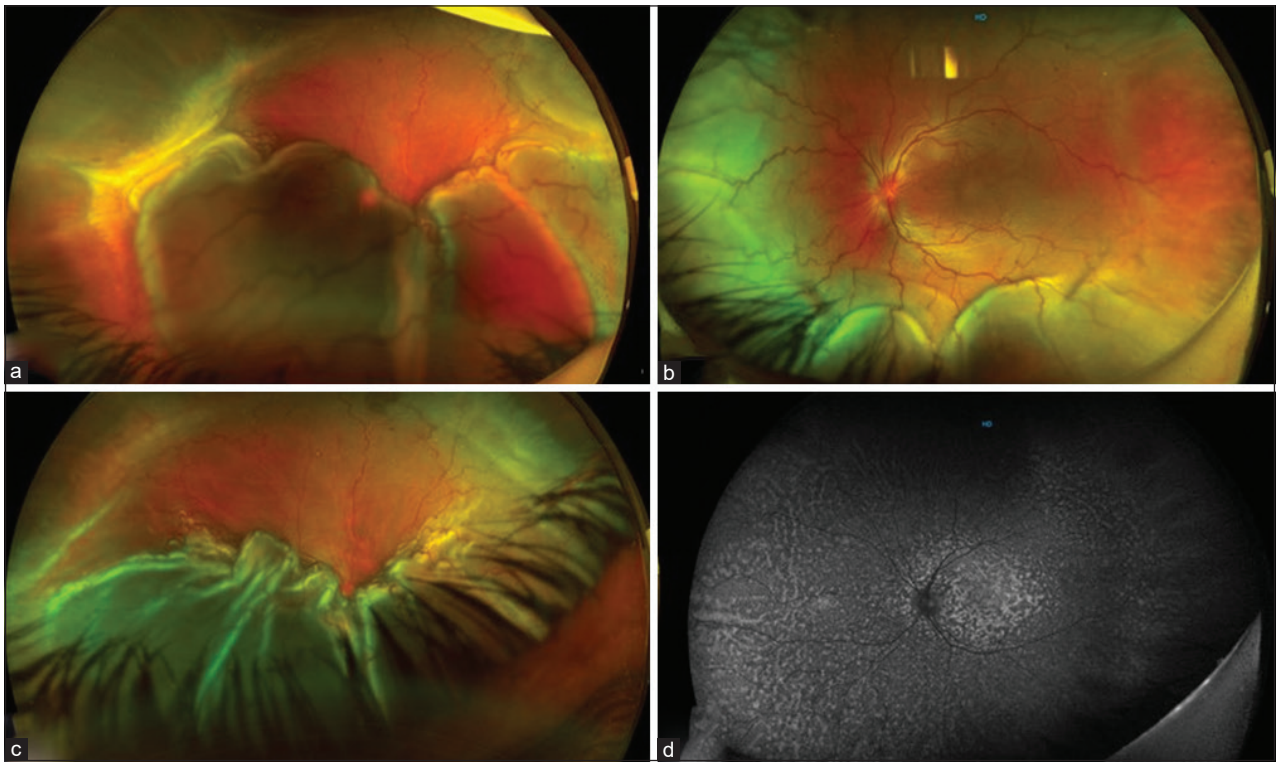

Figure 2: Ultra-widefield color fundus photographs of patient eyes affected with UES demonstrating (a) ERD, (b) ERD with CD, (c) ERD with retinal folds and RPE pigmentary changes, and (d) ultra-widefield fundus autofluorescence image demonstrating RPE pigmentary changes

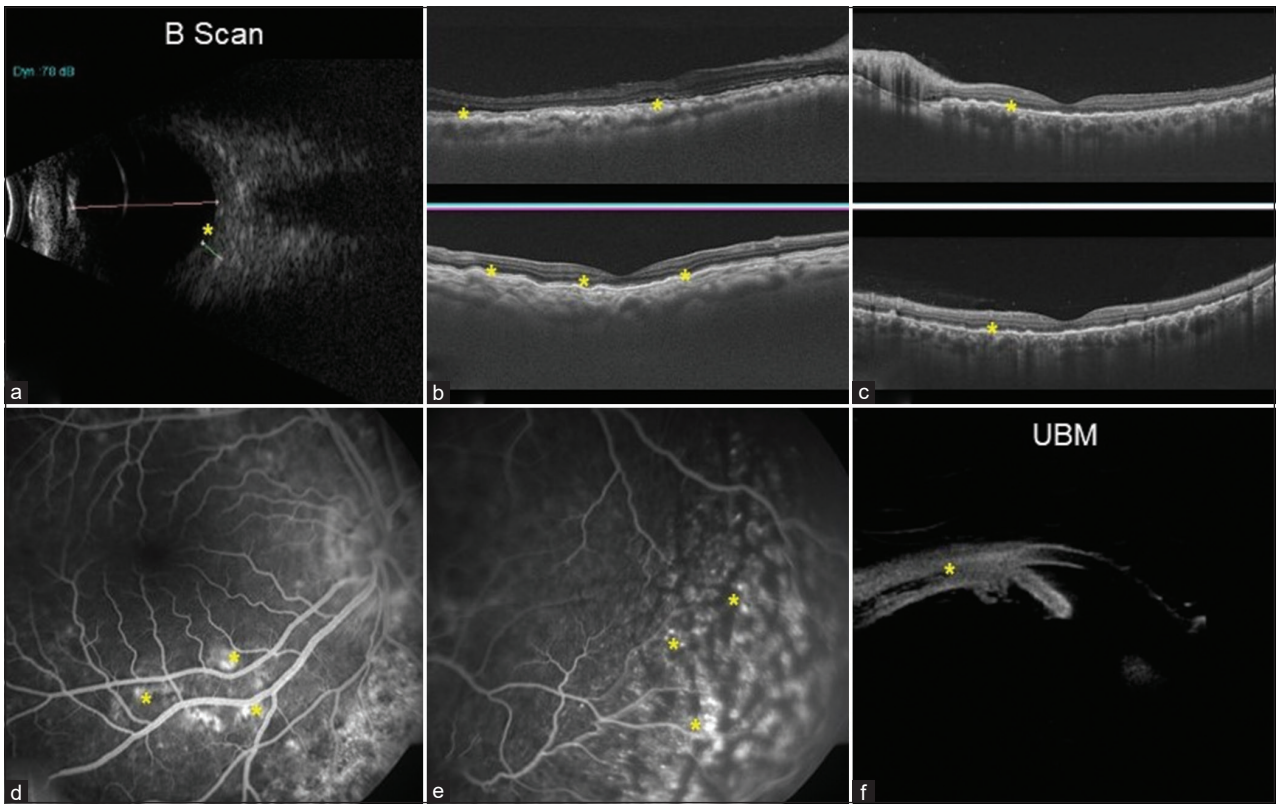

Figure 3: Common findings of ocular investigations of eyes affected with UES: (a) increased CT on B-scan ultrasonography, (b) swept-source OCT line scan images demonstrating SRF and sub-retinal folds, (c) swept-source OCT line scan images demonstrating increased RPE hyperreflectivity, (d and e) FFA images demonstrating pinpoint and patchy areas of hyperfluorescence, and (f) ultrasound biomicroscopy scan demonstrating peripheral CD and ciliary body effusion. (Findings indicated by a yellow asterisk in images)

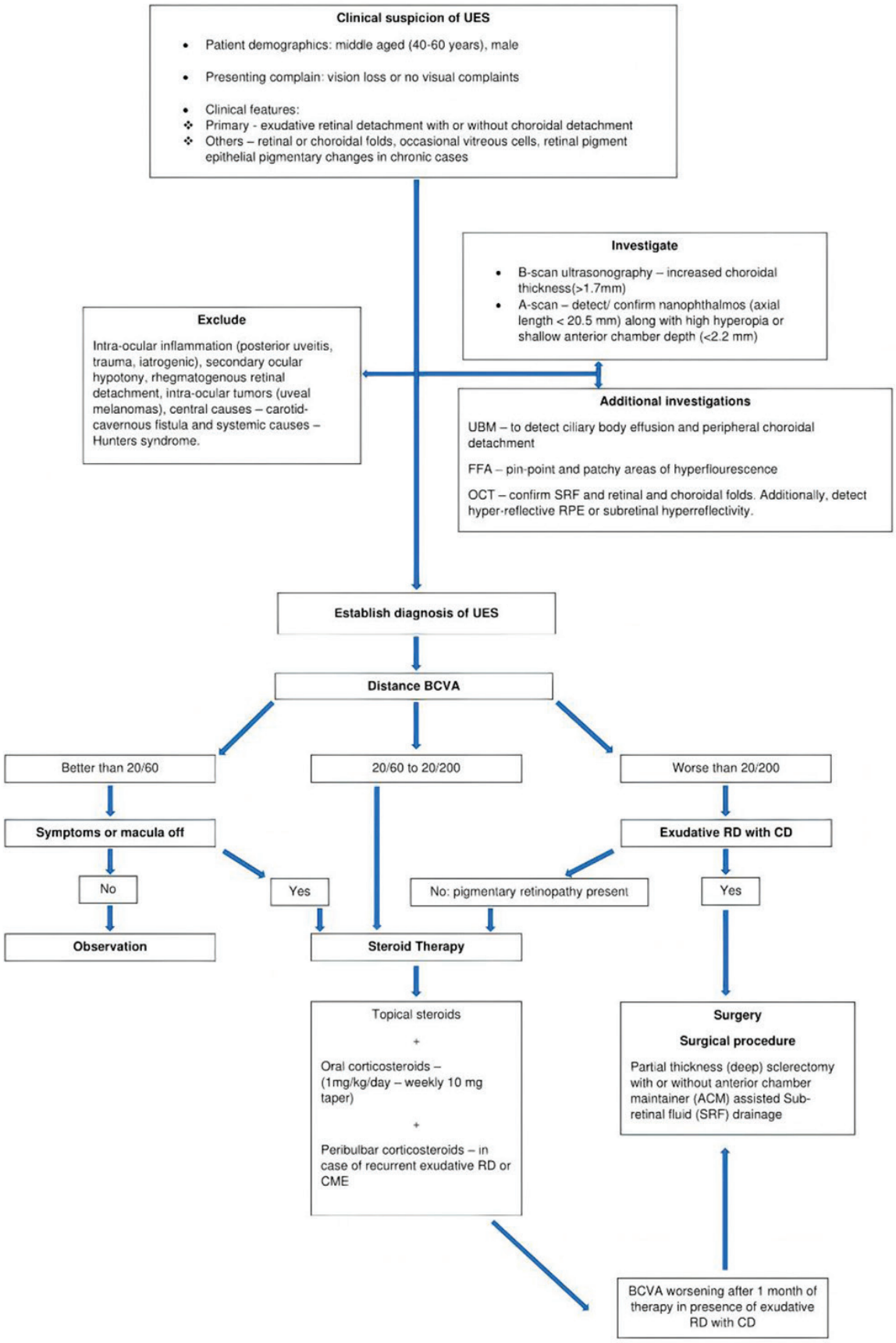

Figure 4: Algorithmic approach for the management of UES

UBM

UBM was performed in 7 affected eyes of 5 patients. Peripheral ciliochoroidal effusion, shallow angles, and shallow angles with ciliary body effusion were seen in 2 eyes each, whereas findings were normal in 1 eye.

Management and outcome

The management of UES could be divided into an observation group and an intervention group consisting of medical management and surgical management. Management was based on the clinical judgment of the treating ophthalmologist as elaborated upon in Fig. 4 which shows an algorithm approach for the management of UES based on our experience. Overall, the median follow-up period was 6.5 months and ranged from 3 days to 76 months.

Observation

Eight affected eyes of four patients with the bilateral disease were observed at presentation. The mean presenting BCVA for this group was 0.27 ± 0.17 logMAR (mean Snellen: 20/40; range 20/20 to 20/50). Two of these patients were detected to have UES during a general eye check-up. The other 2 patients (both nanophthalmos) presented with DOV; one had bilateral shallow inferior ERD, and the other had retinal folds.

Only one eye of one patient (retinal folds at presentation) ultimately needed surgery in the form of partial-thickness sclerectomy with external SRF drainage 48 months after presentation after he developed ERD with CD and his BCVA worsened. This patient was followed up for 76 months, and the BCVA for his other affected eye was maintained without any intervention. The other patients did not follow up after their 2-month follow-up visit, but during their last visit, BCVA and retinal status remained unchanged from that at presentation.

Intervention

Fourteen affected eyes of ten patients (4 bilateral UES) were managed by medical therapy alone at presentation in the form of topical and oral corticosteroids. All eyes were administered topical steroids—1% prednisolone acetate was used in a weekly tapering dose started 6–8 times/day. Supplementary oral corticosteroids in tapering doses starting at 1.0 mg/kg body weight were administered in 7 patients; 4 with bilateral disease, that is, a total of 11 affected eyes. Additionally, periocular corticosteroid injection (1 ml—40 mg/ml triamcinolone acetonide) was also needed in 2 eyes of 2 patients with unilateral disease, one of whom had recurrent ERD and the other had CME confirmed on OCT.

Surgical intervention was planned for 3 affected eyes of 2 patients. One of these patients underwent surgery after 1 month of pre-operative medical therapy, whereas the other with bilateral disease underwent surgery in both eyes over 2 weeks.

Medical management

Twelve affected eyes of nine patients were managed with medical therapy alone. Eight eyes had nanophthalmos. The mean BCVA at presentation for this group was 0.7 ± 0.32 logMAR (mean Snellen: 20/100), and the mean duration of DOV was 4.4 ± 4 months. Oral corticosteroids were administered in all patients with ERD and RPE pigmentary changes.

The duration of follow-up was variable for these eyes. Five affected eyes of three patients were followed for more than 18 months (mean follow-up: 38 ± 19 months), whereas the mean follow-up duration of the rest was 3 ± 1 months. In the longer follow-up sub-group, the mean BCVA at the last follow-up was 0.8 ± 0.2 logMAR (mean Snellen: 20/125). Inferior exudative but stable RD persisted in 1 affected eye, whereas in the other 4, the retina was spontaneously re-attached and retinal pigmentary changes had developed in 2 of these eyes. In the other group, the mean BCVA at the last follow-up was 0.7 ± 0.3 logMAR (mean Snellen: 20/100) and the retina was attached in all but 1 affected eye where a stable ERD with CD persisted at the last visit. BCVA reduction in the longer follow-up sub-group was explained by the onset of new cataract formation.

Surgical management

Surgical intervention was needed for 6 affected eyes of 5 patients (3 bilateral UES). Four eyes had nanophthalmos, and the other two eyes were myopic—1 low myope (axial length: 21.99 mm, mean spherical equivalent: 2.5D) and 1 moderate myope (axial length: 21.9 mm, mean spherical equivalent: 3.25D). The pre-operative BCVA in this group was 1.6 ± 0.9 logMAR (mean Snellen: 20/800), and all eyes had ERD involving the posterior pole, along with co-existent CD. Partial-thickness (deep) sclerectomy with SRF drainage was the procedure performed for all cases. Partial-thickness (deep) sclerectomy with ACM-assisted SRF drainage was advantageous in achieving favorable anatomical and functional outcomes in eyes with bullous SRF and was performed in 2 eyes. Two eyes (both nanophthalmos) required resurgery as there was a post-operative recurrence of ERD involving the macula at 1 month. Pars plana lensectomy and pars plana vitrectomy with endolaser and silicon oil injection was performed in both cases. Silicon oil removal was not performed during the course of follow-up in these patients. The mean duration of follow-up in this group was 11 ± 5 months, and the final BCVA was 1 ± 1.29 logMAR (mean Snellen: 20/200). All eyes which underwent surgical intervention had an attached retina with resolved ERD at the last follow-up visit.

Other interventions

Four eyes underwent cataract surgery. In one case, cataract surgery with IOL implantation was performed before surgical intervention for UES as it had a total white cataract which precluded fundus visualization. The other 3 underwent cataract surgery with IOL implantation during follow-up after the cataract became visually significant.

YAG-laser peripheral iridotomy was performed for 5 eyes that had occludable angles with raised IOP. None of the eyes underwent any vitreoretinal surgical intervention post iridotomy.

Discussion

To the best of our knowledge, this is the largest case series of UES to be reported in Indian eyes. Our findings differ in certain aspects from those reported previously in the medical literature. In our series, the mean age of the patients at presentation was 43.25 ± 14.77 years which is close to the mean age of 45 years reported by Johnson and Gass in a series of UES patients who required surgical intervention.[15] However, this is younger than the mean age reported in a more recent case series: 70 years reported by Shields *et al*.[16] and 62 years reported separately by Sharma *et al*.[2] and Claeys *et al*.[17] Another older series by Uyama *et al*. reported the mean age as 52 years across all types

of UES.[5] Eighty-eight percent of our patients were male, which is higher than the 56% reported by Uyama *et al*.,[5] as well as in other recent case series.[2,16,17] Additionally, 56% of the patients in our series had bilateral disease which is similar to that reported by Sharma *et al*.[2] and Claeys *et al*.[17] but is much higher than the 13–18% reported by Uyama *et al*.[4] and Shields *et al*.[16] Sixty percent of the affected eyes in our series had nanophthalmos which is also higher than 31–33% previously reported.[2,5]

Although the demographic and baseline characteristics of the patients in our series differ from what has been reported previously in the literature, the most common presenting complaints of vision loss along with the common findings of ERD and increased CT are similar.[2,16]

The response to treatment in our patients also differed. Except for one patient from the observation group who underwent surgery, the documented follow-up duration for the remaining 3 patients (6 eyes) was too short to definitively comment on the natural history of UES. This suggests that either the disease was stable or the patients sought care elsewhere when the disease worsened. Observation may thus be initially appropriate in UES with BCVA better than 20/60 (Snellen) without symptoms or ERD involving the macula.

Treatment with corticosteroid therapy was effective in controlling the disease and maintaining vision even in nanophthalmic UES. This is a useful treatment option in eyes detected to have UES with BCVA of 20/60 and worse but better than 20/200 (Snellen) or with ERD involving the macula. The role of steroids in the management of UES has previously been reported by Shields *et al*.[16] However, unlike our series, the use of corticosteroids in nanophthalmic UES was found to be ineffective and not recommended.

UES eyes that required surgery in our series (24%) were lesser than 45% reported by Sharma *et al*.[2] Various surgical techniques have been described to surgically manage UES. These include vortex vein decompression,[18] quadrantic partial-thickness sclerectomies without vortex vein decompression,[15] sub-scleral sclerectomy,[5] and extensive circumferential partial-thickness sclerectomy.[19] The surgical technique used to manage our patients is also one which has not been previously described in the literature.

Partial-thickness (deep) sclerectomy in all four quadrants with ACM-assisted SRF drainage without vortex vein decompression was the preferred technique in our series. ACM-assisted SRF drainage is a safe technique that allows for more controlled SRF drainage and protects against globe collapse due to sudden hypotony during the procedure.

UES patients with an AL >20.5 mm are a subset of patients in whom conventional surgical management with partial-thickness sclerectomy has been reported to have limited success.[20] More radical treatment in the form of full-thickness sclerectomy or USG-guided placement of sclerotomies subjacent to the area of maximal choroidal swelling is needed to treat these cases.[20,21] Our same technique of ACM-assisted SRF drainage was successful in one such patient and did not need a different approach. More experience with additional cases by other surgeons will help to evaluate this technique further because surgical intervention for UES is not common.

This study, though, does have limitations. The retrospective nature of our study is an obvious limitation. All the ophthalmic imaging studies and investigations discussed in our study had not been performed for all included patients. We also did not have data on the scleral thickness of patients included. Magnetic resonance imaging (MRI) could have been useful in this regard in addition to conclusively ruling out a central cause of UES.[20,22] Histopathological analysis of excised scleral tissue, from the cases who underwent surgery would have also added value to the study and helped confirm the disease pathogenesis. Due to these limitations, we could not classify our patients into type 1, type 2, and type 3 UES[5] and we instead classified them into UES patients with and without nanophthalmos. As our data set did not have biometry values for all cases, it was not possible to clearly differentiate between nanophthalmos and posterior microphthalmos in the affected eyes.[14] However, as the biometry criteria for nanophthalmos and posterior microphthalmos can overlap and the association of nanophthalmos with UES is well known, we classified these eyes to have nanophthalmos.[23]

Conclusion

To conclude, UES is more common in middle-aged males and bilateral in more than half the cases. Nanophthalmos is a common ocular association. UES can be suitably managed both medically and surgically based on clinical presentation. Integrated care across ophthalmic sub-specialties should be sought to manage associated cataract and glaucoma in these patients. Given the rarity of UES, a multi-center prospective study is needed to help formulate treatment guidelines and accurately compare different treatment options.

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Conflicts of interest

There are no conflicts of interest.

References

1. Yeh PT, Yang CM, Yang CH, Lin CP. Nonrhegmatogenous retinal detachment. In: Schachat AP, editor. Ryan's Retina. 6th ed. Beijing: Elsevier; 2018. p. 1830-1.
2. Sharma R, Foot B, Jackson TL. A prospective, population-based, surveillance (BOSU) study of uveal effusion syndrome in the UK. Eur J Ophthalmol 2021;31:2451-6.
3. Elagouz M, Stanescu-Segall D, Jackson TL. Uveal effusion syndrome. Surv Ophthalmol 2010;55:134-45.
4. Gass JD, Jallow S. Idiopathic serous detachment of the choroid, ciliary body, and retina (uveal effusion syndrome). Ophthalmology 1982;89:1018-32.
5. Uyama M, Takahashi K, Kozaki J, Tagami N, Takada Y, Ohkuma H, *et al*. Uveal effusion syndrome: Clinical features, surgical treatment, histologic examination of the sclera, and pathophysiology. Ophthalmology 2000;107:441-9.
6. Shaffer RN. Discussion of: Calhoun FP Jr. The management of glaucoma in nanophthalmos. Trans Am Ophthalmol Soc 1975;73:96-112.
7. Gass JD. Uveal effusion syndrome: A new hypothesis concerning pathogenesis and technique of surgical treatment. 1983. Retina 2003;23 (6 Suppl):159-63.
8. Ward RC, Gragoudas ES, Pon DM, Albert DM. Abnormal scleral findings in uveal effusion syndrome. Am J Ophthalmol 1988;106:139-46.
9. Jackson TL, Hussain A, Morley AM, Sullivan PM, Hodgetts A, El-Osta A, *et al*. Scleral hydraulic conductivity and macromolecular diffusion in patients with uveal effusion syndrome. Invest Ophthalmol Vis Sci 2008;49:5033-40.
10. Daniele S, Schepens CL. Can chronic bulbar hypotony be

responsible for uveal effusion? Report of two cases. Ophthalmic Surg 1989;20:872-5.

11. Kumar A, Kedar S, Singh RP. The indocyanine green findings in idiopathic uveal effusion syndrome. Indian J Ophthalmol 2002;50:217-9.
12. Qin B, Tang M, Li Y, Zhang X, Chu R, Huang D. Anterior segment dimensions in Asian and Caucasian eyes measured by optical coherence tomography. Ophthalmic Surg Lasers Imaging 2012;43:135-42.
13. Hoffman RS, Vasavada AR, Allen QB, Snyder ME, Devgan U, Braga-Mele R, *et al*. ASCRS cataract clinical committee, challenging/complicated cataract surgery subcommittee. Cataract surgery in the small eye. J Cataract Refract Surg 2015;41:2565-75.
14. Relhan N, Jalali S, Pehre N, Rao HL, Manusani U, Bodduluri L. High-hyperopia database, part I: Clinical characterisation including morphometric (biometric) differentiation of posterior microphthalmos from nanophthalmos. Eye (Lond) 2016;30:120-6.
15. Johnson MW, Gass JD. Surgical management of the idiopathic uveal effusion syndrome. Ophthalmology 1990;97:778-85.
16. 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;65:1093-104.
17. Claeys E, Stalmans P, Van Calster J, Casteels I, Stalmans I, Vandewalle E. A retrospective case series of uveal effusion syndrome. J Glaucoma 2020;29:995-8.
18. Casswell AG, Gregor ZJ, Bird AC. The surgical management of uveal effusion syndrome. Eye (Lond) 1987;1:115-9.
19. Mansour A, Stewart MW, Shields CL, Hamam R, Fattah MA, Sheheitli H, *et al*. Extensive circumferential partial-thickness sclerectomy in eyes with extreme nanophthalmos and spontaneous uveal effusion. Br J Ophthalmol 2019;103:1862-7.
20. Maggio E, Polito A, Prigione G, Pertile G. Uveal effusion syndrome mimicking severe chronic posterior uveitis: A case series of seven eyes of four patients. Graefes Arch Clin Exp Ophthalmol 2016;254:545-52.
21. Ghazi NG, Richards CP, Abazari A. A modified ultrasound-guided surgical technique for the management of the uveal effusion syndrome in patients with normal axial length and scleral thickness. Retina 2013;33:1211-9.
22. Andrijević Derk B, Benčić G, Corluka V, Zorić Geber M, Vatavuk Z. Medical therapy for uveal effusion syndrome. Eye (Lond) 2014;28:1028-31.
23. Carricondo PC, Andrade T, Prasov L, Ayres BM, Moroi SE. Nanophthalmos: A review of the clinical spectrum and genetics. J Ophthalmol 2018;2018:2735465.