Featured Original Article

Uveal effusion syndrome in 104 eyes: Response to corticosteroids – The 2017 Axel C. Hansen lecture

Carol L Shields, Kelsey Roelofs, Maura Di Nicola, Kareem Sioufi, Arman Mashayekhi, Jerry A Shields

Purpose: The purpose of the study was to investigate the corticosteroids for uveal effusion syndrome (UES). **Methods:** Retrospective series of 104 eyes with UES treated with oral corticosteroids (OCS), periocular corticosteroids (PCS), topical corticosteroids (TCS), or observation (OBS). Main outcome measure was UES resolution. **Results:** Of 104 eyes, treatment included OCS (n = 27), PCS (n = 12), TCS (n = 11), and OBS (n = 54). A comparison of the four groups (OCS vs. PCS vs. TCS vs. OBS) revealed differences in those managed with OCS versus OBS as younger (66 vs. 72 years, P = 0.049), PCS versus OBS as male (100% vs. 54%, P = 0.002), PCS versus OBS with decreased visual acuity (VA)/visual field (91% vs. 51%, P = 0.018), and OBS versus OCS as asymptomatic (28% vs. 0%, P = 0.001). Of the 59 with follow-up information, management included OCS (n = 21), PCS (n = 12), TCS (n = 6), and OBS (n = 20). There were differences in initial VA <20/400 in PCS versus OBS (42% vs. 5%, P = 0.018), effusion thickness in TCS versus OCS (7 vs. 3 mm, P = 0.004), and serous retinal detachment in PCS versus OBS (100% vs. 30%, P < 0.001) and PCS versus OCS (100% vs. 57%, P = 0.012). Regarding outcomes, VA showed less worsening in OCS versus OBS (0% vs. 30%, P = 0.008) and OCS versus PCS (0% vs. 33%, P = 0.012). There was no difference in rate of effusion resolution or effusion recurrence. Overall, using combination of corticosteroid therapies, effusion resolution was achieved in 56/59 (95%) cases and the need for surgical management with scleral windows was necessary in only 3/59 (5%) cases. Complications included cataract (n = 9) and no instance of steroid-induced glaucoma. **Conclusion:** Management of UES is complex and depends on disease severity. Using various corticosteroid delivery routes, UES control was achieved in 95%, and scleral window surgery was required in only 5%. A trial of corticosteroids can benefit patients with UES.

Key words: Choroid, corticosteroids, detachment, effusion, melanoma, pseudomelanoma, uvea, uveal effusion syndrome

Access this article online

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_752_17

Quick Response Code:

Uveal effusion syndrome (UES) was first reported by Schepens and Brockhurst in a seminal paper in 1963 in which they described the clinical features in 17 male patients who demonstrated choroidal detachment, often with secondary retinal detachment, optic disc swelling, and minimal signs of uveitis.[1] They noted that UES was "insidiously progressive over a period varying between several months (up to) 7 or 8 years". Of the nine patients with severe UES, total retinal detachment, often with lenticular touch, occurred within 1 year, and slowly resolved in five but remained permanent in four cases.[1]

Several theories on the pathophysiology of UES have been speculated, including vortex vein obstruction, increased choroidal permeability, intrinsic choroidal alterations, and decreased scleral permeability.[2-9] The latter has been supported by histopathologic studies revealing thicker sclera with disorganized bundles of collagen fibrils, increased amounts of glycosaminoglycans, and decreased permeability to albumin resulting in osmotic gradient with fluid retention in the suprachoroidal and supraciliary space. There are several inflammatory and hydrostatic conditions that can result in uveal effusion, but the term "UES" is reserved specifically for idiopathic cases.[2,3]

Based on the literature, therapy for UES generally involves surgical creation of scleral windows to decompress suprachoroidal fluid, and more recently, implantation of Express Valve.[3,4,10-20] There is very little information on the role of systemic or periocular corticosteroids (PCS) for UES and PubMed search for keywords "uvea," "choroid," "effusion," and "steroid" yielded one case series in the English literature.[21] A major survey on UES indicated that "treatment with systemic steroids does not appear to be effective."[4] The mechanism of corticosteroids for UES is unclear, but some speculate that there could be generalized reduction in inflammatory factors or control of transudation and edema by membrane stabilization. Herein, we evaluate the largest published cohort of UES cases (n = 104 eyes), mostly referred for suspicion of uveal tumor, and we specifically investigate the role of local and systemic corticosteroids as a primary therapy for this rare condition.

Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University, Philadelphia, Pennsylvania, USA

Correspondence to: Dr. Carol L Shields, Ocular Oncology Service, 840 Walnut Street, Suite 1440, Philadelphia 19107, Pennsylvania, USA. E-mail: arolshields@gmail.com

Manuscript received: 18.08.17; Revision accepted: 19.09.17

Cite this article as: 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.

Methods

A review of the computerized coding for patients evaluated on the Ocular Oncology Service at Wills Eye Hospital between October 1, 1975, and January 1, 2017, with the diagnosis of idiopathic UES was retrospectively performed. Eyes classified as idiopathic UES demonstrated serous uveal detachment, with axial length >19 mm (nonnanophthalmic, Type 3), and no evident inflammatory, vascular, or tumor-related condition and normal scleral appearance and thickness by ultrasonography and/or magnetic resonance imaging (MRI). The patient demographic and clinical features were evaluated, and therapeutic intervention was recorded. Institutional review board approval was obtained.

The demographic data included patient age at presentation, race, gender, effusion laterality, presenting symptom with related duration, and referring diagnosis. The clinical features included visual acuity (VA), intraocular pressure (mmHg), anterior segment features (eyelid edema, conjunctival, episcleral and scleral injection, scleral inflammation), UES location, extent, and circumferential diameter (mm) and thickness (mm). The presence and extent of an associated serous retinal detachment was recorded. The results of testing including transillumination, ocular ultrasonography, and MRI, when available, were recorded.

Specific treatment strategies were evaluated including primary treatment (oral, periocular, or topical corticosteroids (TCS) or observation [OBS]) and secondary treatment (additional oral, periocular, or TCS, intravitreal corticosteroid, or surgical sclerectomy [scleral window]). The dose and duration of medication was recorded.

Treatment outcomes were evaluated for patients who maintained follow-up in our clinic regarding VA (improvement [≥2 Snellen lines increase], stable, and worsening [≥2 Snellen lines decrease]), UES outcomes including UES any response, complete resolution, worsening, and recurrence. Assessment regarding the need for secondary treatment, type of treatment, and complications was recorded.

Results

There were 104 eyes of 97 patients with idiopathic UES included in this analysis, and all were classified as Type 3, based on the lack of nanophthalmos and lack of scleral wall abnormality.[3] The demographic features are listed in Table 1. The mean age was 70 years (median: 71, range: 27–94 years); majority were Caucasian (92%), male (64%), and with unilateral findings (87%). The referring diagnosis was choroidal melanoma (47%), choroidal metastasis (3%), choroidal tumor nonspecified (34%), and UES (16%). The chief complaints included decreased vision or visual field (VF) loss (56%), pain (11%), other symptoms (19%), or no symptoms (14%). The therapeutic intervention included OBS (n = 54, 52%), oral corticosteroids (OCS) (n = 27, 26%), PCS (n = 12, 12%), and TCS (n = 11, 11%). There was no difference among the treatment groups regarding race, UES laterality, or symptom duration. There was significant difference in treatment by mean age for OBS versus OCS (72 vs. 66 years, P = 0.042), gender male for PCS versus OBS (100% vs. 54%, P = 0.002), and PCS versus TCS (100% vs. 64%, P = 0.037), decreased vision/VF loss in PCS versus OBS (83% vs. 50%, P = 0.018), and lack of symptoms in OCS versus OBS (0% vs. 28%, P = 0.001).

A comparison of all 104 eyes based on therapeutic regimen regarding clinical features is listed in Table 2. In general, eyes receiving OCS, PCS, and TCS showed more advanced features than those treated with OBS. Compared to eyes managed with OBS, those treated with OCS (vs. OBS) demonstrated higher rate of associated retinal detachment (59% vs. 31%, P = 0.029), those treated with PCS (vs. OBS) showed poorer initial VA of 20/20–20/40 (0% vs. 30%) and higher rate of associated retinal detachment (100% vs. 31%, P < 0.001), and those treated with TCS (vs. OBS) showed poorer initial VA of 20/200–20/400 (45% vs. 15%, P = 0.034).

A comparison of the 59 eyes with follow-up in our department is listed in Table 3. The mean axial length of the globe was 22 mm (median 22 mm, range 19–25 mm). Of the 59 eyes, treatment was OBS (n = 20, 34%), OCS (n = 21, 36%), PCS (n = 12, 20%), and TCS (n = 6, 10%). Analysis of the demographic and clinical features of this cohort, compared to OBS, revealed patient mean age at treatment with OCS (vs. OBS) (66 vs. 77, P < 0.001), and PCS (67 vs. 77, P = 0.021) was younger, more likely male with PCS (100% vs. 60%, P = 0.013), with symptoms in OCS (100% vs. 40%, P = 0.001), with decreased (VA/VF) in PCS (83% vs. 35%, P = 0.011), and with initial VA of 20/400 or less in PCS (42% vs. 5%, P = 0.018).

Outcomes analysis of the 59 eyes is listed in Table 4 [Figs. 1 and 2]. Regarding VA outcome, worsening vision was less in OCS versus OBS (0% vs. 30%, P = 0.008) and in OCS versus PCS (0% vs. 33%, P = 0.012). However, it should be noted that OCS required occasional secondary treatments (usually further corticosteroids) to achieve these outcomes [Table 4]. Regarding effusion outcomes, there was no significant difference among the four treatment groups. Regarding the need for secondary treatment, greater need was found in OCS versus OBS (86% vs. 10%, P < 0.001), OCS versus PCS (86% vs. 42%, P = 0.016), and OCS versus TCS (86% vs. 0%, P < 0.001). Overall, the median time to effusion response was 3 months and to complete resolution was 11 months [Table 4]. In the entire group of 59 eyes, only three (5%) needed scleral window surgery (2 following OCS and 1 following PCS). Regarding complications, cataract was more commonly found in OCS versus OBS (33% vs. 5%, P = 0.044). There was no case of corticosteroid-induced glaucoma.

Discussion

In our practice of ocular oncology, we are commonly referred patients with intraocular conditions for evaluation to rule out malignancy.[22-26] In a comprehensive report on 1739 patients with choroidal pseudomelanomas, the top 10 conditions included choroidal nevus (49%), peripheral exudative hemorrhagic chorioretinopathy (8%), congenital hypertrophy of the retinal pigment epithelium (RPE) (6%), retina/RPE hemorrhagic detachment (5%), choroidal hemangioma (5%), age-related macular degeneration (4%), RPE hyperplasia (2%), optic disc melanocytoma (2%), choroidal metastasis (2%), and choroidal hemorrhage (2%).[22,23] Uveal effusion was #14 in the list of pseudomelanomas. Differentiation of uveal effusion from ciliochoroidal melanoma involves several clinical features suggestive of effusion such as ophthalmoscopic evidence of subtle choroidal folds or deep cleft between elevated lobes

Table 1: Uveal effusion syndrome in 104 eyes of 97 patients with comparative response to corticosteroids (oral, periocular injection, or topical) or observation: Demographics

	All cases (*n*=104), *n* (%)	OBS (*n*=54), *n* (%)	OCS (*n*=27), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=11), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
Age (years), mean (median, range)	70 (71, 27-94)	72 (75, 27-94)	66 (67, 45-86)	0.042†	67 (70, 44-90)	0.255†	0.737†	71 (69, 56-87)	0.750	0.200	0.325
Race (*n*)	104	54	27		12			11			
Caucasian	96 (92)	50 (93)	25 (93)	1.00	10 (83)	0.298	0.564	11 (100)	1.00	1.00	0.478
African American	4 (4)	3 (6)	0	0.547	1 (8)	0.561	0.250	0	1.00	1.00	1.00
Asian	3 (3)	0	2 (7)	0.112	1 (8)	0.181	1.00	0	1.00	1.00	1.00
Hispanic	1 (1)	1 (2)	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Gender	104	54	27		12			11			
Male	67 (64)	29 (54)	19 (70)	0.230	12 (100)	0.002	0.041	7 (64)	0.741	0.714	0.037
Female	37 (36)	25 (46)	8 (30)		0			4 (36)			
Laterality	104	54	27		12			11			
Unilateral	90 (87)	48 (89)	21 (78)	0.201	10 (83)	0.630	1.00	11 (100)	0.647	1.00	1.00
Bilateral	14 (13)	6 (11)	6 (22)		2 (17)			0			
Referring diagnosis	104	54	27		12			11			
Choroidal melanoma	49 (47)	28 (52)	11 (41)	-	5 (42)	-	-	5 (45)	-	-	-
Uveal effusion syndrome	17 (16)	9 (17)	6 (22)		1 (8)			1 (9)			
Choroidal metastasis	3 (3)	3 (6)	0		0			0			
Choroidal tumor, nonspecified	35 (34)	14 (26)	10 (37)		6 (50)			5 (45)			
Chief complaint	104	54	27		12			11			
Decreased VA/VF	58 (56)	27 (50)	15 (56)	0.813	10 (83)	0.018	0.059	6 (55)	1.00	1.00	0.193
Pain	12 (11)	5 (9)	6 (22)	0.169	1 (8)	1.00	0.648	0	0.578	0.153	1.00
Redness	7 (7)	3 (6)	2 (7)	1.00	0	1.00	1.00	2 (18)	0.196	0.564	0.217
Photopsia	6 (6)	2 (4)	2 (7)	0.600	0	1.00	1.00	2 (18)	0.129	0.564	0.217
Floaters	5 (5)	2 (4)	2 (7)	0.600	1 (8)	1.00	1.00	1 (9)	0.432	1.00	1.00
Metamorphopsia	1 (1)	0	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Asymptomatic	15 (14)	15 (28)	0	0.001	0	0.102	1.00	0	1.00	1.00	1.00
Symptom duration (months), mean (median, range)	6 (2, 0-240)	11 (2, 0-240)	2 (1, 0-18)	0.263†	4 (2, 0-24)	0.548†	0.271†	3 (2, 0-12)	0.540	0.932	0.258

Fisher's exact test was used to calculate *P* value for categorical variables, †Student *t*-test was used to calculate *P* value. VA: Visual acuity, VF: Visual field, OBS: Observation, OCS: Oral corticosteroid, PCS: Periocular corticosteroid, TCS: Topical corticosteroid

Table 2: Uveal effusion syndrome in 104 eyes of 97 patients with comparative response to corticosteroids (oral, periocular injection, or topical) or observation: Clinical features at presentation

Feature	All cases (*n*=104), *n* (%)	OBS (*n*=54), *n* (%)	OCS (*n*=27), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=11), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
VA (*n*)	104	54	27		12			11			
20/20-20/40	27 (26)	18 (33)	8 (30)	0.804	0	0.027	0.041	1 (9)	0.153	0.237	0.478
20/50-20/150	36 (35)	18 (33)	9 (33)	1.00	4 (33)	1.00	1.00	5 (45)	0.499	0.712	0.680
20/200-20/400	19 (18)	8 (15)	3 (11)	0.743	3 (25)	0.405	0.348	5 (45)	0.034	0.031	0.659
<20/400	22 (21)	10 (19)	7 (26)	0.563	5 (42)	0.124	0.455	0	0.190	0.084	0.037
Intraocular pressure (mmHg) (*n*)	104	54	27		12			11			
Mean (median, range)	15 (15,6-42)	16 (15,6-42)	16 (15,9-26)	0.828†	14 (14,7-24)	0.456	0.238†	16 (17,6-23)	0.965	0.910	0.491
Anterior segment features (*n*)	104	54	27		12			11			
Eyelid edema	3 (3)	1 (2)	2 (7)	0.256	0	1.00	1.00	0	1.00	1.00	1.00
Conjunctival injection	32 (31)	15 (28)	13 (48)	0.085	0	0.054	0.003	4 (36)	0.717	0.721	0.037
Episcleral/scleral injection	42 (40)	18 (33)	16 (59)	0.033	4 (33)	1.00	0.176	4 (36)	1.00	0.287	1.00
Sentinel vessels	7 (7)	2 (4)	4 (15)	0.176	1 (8)	0.458	1.00	0	1.00	0.302	1.00
Scleritis (anterior or posterior)	10 (10)	3 (6)	5 (19)	0.101	0	1.00	0.298	2 (18)	0.196	1.00	0.217
Posterior segment features (*n*)	104	54	27		12			11			
Number of quadrants involved											
1 quadrant	15 (14)	11 (20)	2 (7)	0.201	1 (8)	0.187	1.00	1 (9)	0.673	1.00	1.00
2 quadrants	15 (14)	10 (19)	1 (4)	0.089	4 (33)	0.672	0.018	0	0.190	1.00	0.093
3 quadrants	10 (10)	4 (7)	4 (15)	0.219	1 (8)	0.539	1.00	1 (9)	1.00	1.00	1.00
4 quadrants	64 (62)	29 (54)	20 (74)	0.146	6 (50)	1.00	0.272	9 (82)	0.104	1.00	0.193
Effusion anteroposterior location (*n*)	104	54	27		12			11			
Anterior margin											
Equator to ora serrata	47 (45)	26 (48)	10 (37)	0.477	5 (42)	1.00	0.721	6 (55)	0.750	0.471	0.684
Ciliary body	43 (41)	22 (41)	11 (41)	1.00	6 (50)	1.00	1.00	4 (36)	1.00	1.00	0.680
Iris	14 (13)	6 (11)	6 (22)	0.202	1 (8)	1.00	0.648	1 (9)	1.00	0.648	1.00
Posterior margin											
Macula	15 (14)	9 (17)	3 (11)	0.731	2 (17)	1.00	0.615	1 (9)	1.00	1.00	1.00
Macula to equator	55 (53)	30 (56)	10 (37)	0.154	7 (58)	0.747	0.166	8 (73)	0.337	0.074	0.666
Equator to ora serrata	34 (33)	14 (26)	14 (52)	0.024	3 (25)	0.715	0.077	2 (18)	0.717	0.077	1.00
Effusion circumferential diameter (mm) (*n*)	104	54	2		12			11			
Mean (median, range)	55 (72,6-72)	51 (72,6-72)	63 (72,6-72)	0.025†	50 (45,18-72)	0.804†	0.039†	60 (72,11-72)	0.517	0.406	0.275
Effusion thickness (mm) (*n*)	76	37	22		10			7			
Mean (median, range)	5 (4,1-11)	5 (4,2-11)	4 (3,2-7)	0.021†	5 (4,1-12)	0.912†	0.182†	5 (4,2-8)	0.729	0.050	0.952
Associated retinal detachment	50 (48)	17 (31)	16 (59)	0.029	12 (100)	<0.001	0.016	5 (45)	0.514	0.487	0.004
Subretinal fluid involving macula	47 (45)	16 (94)	16 (100)	1.00	12 (100)	1.00	1.00	3 (60)	0.116	0.047	0.073
Subretinal fluid involving fovea	35 (34)	11 (65)	13 (81)	0.438	8 (67)	1.00	0.661	3 (60)	1.00	0.552	1.00

Contd...

Table 2: Contd...

Feature	All cases (*n*=104), *n* (%)	OBS (*n*=54), *n* (%)	OCS (*n*=27), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=11), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
Choroidal folds	37 (36)	17 (31)	13 (48)	0.153	3 (25)	0.743	0.291	4 (36)	0.736	0.721	0.666
Location of folds (*n*)	37	17	13		3						
Posterior margin of effusion	8 (22)	5 (29)	2 (15)	0.426	1 (33)	1.00	0.489	0	0.532	1.00	0.428
Over apex of effusion	1 (3)	0	1 (8)	0.433	0	1.00	1.00	0	1.00	1.00	1.00
Anterior margin of effusion	1 (3)	1 (6)	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Macula	27 (72)	11 (65)	10 (77)	0.698	2 (66)	1.00	1.00	4 (100)	0.280	0.541	0.428
Transillumination (*n*)	81	43	19		10			9			
Transmission	75 (93)	39 (91)	19 (100)	0.302	9 (90)	1.00	0.344	8 (89)	1.00	0.321	1.00
Blockage of light	6 (7)	4 (9)	0		1 (10)			1 (11)			
Ultrasound density (*n*)	97	49	26		11			11			
Acoustically hollow	70 (72)	32 (65)	20 (77)	0.430	8 (67)	0.736	1.00	10 (91)	0.147	0.648	0.586
Acoustically solid	27 (28)	17 (35)	6 (23)		3 (27)			1 (9)			
Ultrasound surface (*n*)	92	46	26		10			10			
Smooth surface	62 (67)	36 (78)	15 (58)	0.104	4 (40)	0.024	0.462	7 (70)	0.682	0.706	0.369
Undulating surface	30 (33)	10 (22)	11 (42)		6 (60)			3 (30)			
Axial length (mm) (*n*)	44	15	18		9			2			
Mean (median, range)	22 (22,16-25)	22 (22,16-25)	22 (22,18-25)	0.970†	22 (22,18-23)	0.952†	0.916†	23 (23,22-24)	0.445	0.377	0.379
Magnetic resonance imaging (*n*)	19	5	7		5			2			
Choroidal effusion not visualized	7 (37)	2 (40)	4 (57)	1.00	0	0.444	0.08	1 (50)	1.00	1.00	1.00
T1 hyper intense	10 (53)	2 (40)	3 (43)	1.00	4 (80)	0.523	0.292	1 (50)	1.00	1.00	1.00
T2 hypo intense	7 (37)	2 (40)	1 (14)	0.052	3 (60)	1.00	0.222	1 (50)	1.00	1.00	1.00
T1 fat suppression gadolinium enhanced	6 (32)	1 (20)	1 (14)	1.00	3 (60)	0.523	0.222	1 (50)	1.00	1.00	1.00

Fisher exact test was used to calculate *P* value for categorical variables, †Student *t*-test was used to calculate *P* value. OBS: Observation, OCS: Oral corticosteroid, PCS: Periocular corticosteroid, TCS: Topical corticosteroid, VA: Visual acuity

Table 3: Uveal effusion syndrome with comparative response to corticosteroids or observation: Analysis of demographics and clinical characteristics in 59 cases*

Outcome	All cases (*n*=59), *n* (%)	OBS (*n*=20), *n* (%)	OCS (*n*=21), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=6), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
Age (years)											
Mean (median, range)	70 (71, 44-94)	77 (77, 56-94)	66 (67, 45-77)	<0.001	67 (70, 44-90)	0.021	0.613	68 (67, 56-81)	0.064	0.617	0.962
Race											
Caucasian	54 (92)	19 (95)	19 (90)	1.00	10 (83)	0.540	0.610	6 (100)	1.00	1.00	0.529
African American	2 (3)	1 (5)	0	0.487	1 (8)	0.363	0.363	0	1.00	1.00	1.00
Asian	3 (5)	0	2 (10)	0.487	1 (8)	1.00	1.00	0	1.00	1.00	1.00
Hispanic	0	0	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Gender											
Male	45 (76)	12 (60)	17 (81)	1.00	12 (100)	0.013	0.271	4 (67)	1.00	0.587	0.098
Female	14 (24)	8 (40)	4 (19)		0			2 (33)			
Laterality											
Unilateral	49 (83)	18 (90)	15 (71)	0.237	10 (83)	0.619	0.677	6 (100)	1.00	0.284	0.529
Bilateral	10 (17)	2 (10)	6 (29)		2 (16)			0			
Referring diagnosis											
Choroidal melanoma	29 (49)	13 (65)	8 (38)	-	5 (42)	-	-	3 (50)	-	-	-
Uveal effusion syndrome	9 (15)	3 (15)	5 (24)		1 (8)			0			
Choroidal metastasis	1 (2)	3 (15)	0		0			0			
Choroidal tumor, nonspecified	19 (32)	1 (5)	7 (33)		6 (50)			3 (50)			
Preseptal cellulitis	1 (2)	0	1 (5)		0			0			
Chief complaint											
Decreased VA/VF	33 (56)	7 (35)	13 (62)	0.121	10 (83)	0.011	0.259	3 (50)	0.644	0.661	0.268
Pain	4 (7)	0	3 (14)	0.231	1 (8)	0.375	1.00	0	1.00	1.00	1.00
Redness	3 (5)	1 (5)	1 (5)	1.00	0	1.00	1.00	1 (17)	0.415	0.401	0.333
Photopsia	6 (10)	2 (10)	2 (10)	1.00	0	0.516	0.522	2 (33)	0.218	0.061	0.028
Floaters	4 (7)	2 (10)	2 (10)	1.00	0	0.516	0.522	0	1.00	1.00	1.00
Metamorphopsia	1 (2)	0	0	1.00	1 (8)	0.375	0.363	0	1.00	1.00	1.00
Asymptomatic	8 (14)	8 (40)	0	0.001	0	0.013	1.00	0	0.131	1.00	1.00
Symptom duration (months)											
Mean (median, range)	3 (1, 0-24)	3 (1, 0-10)	2 (1, 0-18)	0.663†	4 (2, 0-24)	0.613†	0.346†	1 (1, 0-2)	0.314†	0.576†	0.388†
VA											
20/20-20/40	15 (25)	7 (35)	7 (33)	1.00	0	0.028	0.031	1 (17)	0.629	0.633	0.333
20/50-20/150	23 (39)	9 (45)	8 (38)	0.755	4 (33)	0.712	0.712	2 (33)	1.00	1.00	1.00
20/200-20/400	10 (17)	3 (15)	1 (5)	0.343	3 (25)	0.343	1.00	3 (50)	0.100	0.024	0.344
<20/400	11 (19)	1 (5)	5 (24)	0.183	5 (42)	0.018	0.434	0	1.00	0.555	0.114
Intraocular pressure (mmHg)											
Mean (median, range)	16 (15, 6-42)	17 (17, 10-42)	15 (14, 9-26)	0.295†	14 (14, 7-24)	0.157†	0.399†	14 (13, 6-23)	0.252†	0.419†	0.865†

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Table 3: Contd...

Outcome	All cases (*n*=59), *n* (%)	OBS (*n*=20), *n* (%)	OCS (*n*=21), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=6), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
Anterior segment features (*n*)	59	20	21		12			6			
Eyelid edema	2 (3)	0	2 (10)	0.487	0	1.00	0.522	0	1.00	1.00	1.00
Conjunctival injection	11 (19)	2 (10)	8 (38)	0.067	0	0.516	0.013	1 (17)	1.00	0.627	0.333
Episcleral/scleral injection	20 (34)	5 (25)	10 (48)	0.197	4 (33)	0.696	0.486	1 (17)	1.00	0.349	0.614
Sentinel vessels	7 (12)	2 (10)	4 (19)	0.662	1 (8)	1.00	0.630	0	1.00	0.543	1.00
Scleritis (anterior or posterior)	0	0	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Posterior segment features (*n*)	59	20	21		12			6			
Number of quadrants involved											
1 quadrant	8 (14)	4 (20)	2 (10)	0.409	1 (8)	0.626	1.00	1 (17)	1.00	0.543	1.00
2 quadrants	11 (19)	6 (30)	1 (5)	0.044	4 (33)	1.00	0.047	0	0.283	1.00	1.00
3 quadrants	4 (7)	0	3 (14)	0.231	1 (8)	0.375	1.00	0	1.00	1.00	1.00
4 quadrants	36 (61)	10 (50)	15 (71)	0.208	6 (50)	1.00	0.274	5 (83)	0.197	1.00	0.315
Effusion anteroposterior location (*n*)	59	20	21		12			6			
Anterior margin											
Equator to ora serrata	28 (47)	11 (55)	8 (38)	0.354	5 (42)	0.716	1.00	4 (67)	1.00	0.357	0.618
Ciliary body	24 (41)	7 (35)	9 (43)	0.751	6 (50)	0.473	0.737	2 (33)	1.00	1.00	0.638
Iris	7 (12)	2 (10)	4 (19)	0.662	1 (8)	1.00	0.630	0	1.00	0.543	1.00
Posterior margin											
Macula	6 (10)	1 (5)	2 (10)	1.00	2 (17)	0.540	0.610	1 (17)	0.415	0.543	1.00
Macula to equator	33 (56)	14 (70)	8 (38)	0.061	7 (58)	0.702	0.300	4 (67)	1.00	0.357	1.00
Equator to ora serrata	20 (34)	5 (25)	11 (52)	0.121	3 (25)	1.00	0.160	1 (17)	1.00	0.181	1.00
Effusion circumferential diameter (mm) (*n*)	59	20	21		12			6			
Mean (median, range)	54 (72, 6-72)	49 (54, 6-72)	62 (72, 6-72)	0.064†	50 (45, 18-72)	0.919†	0.099†	53 (72, 11-72)	0.733†	0.374†	0.790†
Effusion thickness (mm) (*n*)	45	13	18		10			4			
Mean (median, range)	4 (4, 1-12)	5 (4, 2-11)	3 (3, 2-7)	0.162†	5 (4, 1-12)	0.818†	0.163†	7 (7, 4-8)	0.193†	0.004†	0.407†
Associated retinal detachment	34 (58)	6 (30)	12 (57)	0.118	12 (100)	<0.001	0.012	4 (67)	0.162	1.00	0.098
Subretinal fluid involving macula	34 (100)	6 (100)	9 (75)	0.514	12 (100)	1.00	0.217	3 (75)	0.400	1.00	0.250
Subretinal fluid involving fovea	25 (74)	3 (50)	7 (58)	1.00	8 (67)	0.626	1.00	3 (75)	0.571	1.00	1.00
Choroidal folds	23 (39)	9 (45)	10 (48)	1.00	3 (25)	0.451	0.277	1 (17)	0.352	0.349	1.00
Location of folds											
Posterior margin of effusion	6 (26)	3 (33)	2 (20)	0.628	1 (33)	1.00	1.00	0	1.00	1.00	1.00
Over apex of effusion	1 (4)	0	1 (10)	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Anterior margin of effusion	0	0	0	1.00	0	1.00	1.00	0	1.00	1.00	1.00
Macula	16 (70)	6 (67)	7 (70)	1.00	2 (66)	1.00	1.00	1 (100)	1.00	1.00	1.00
Transillumination (*n*)	46	17	14		10			5			

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Table 3: Contd...

Outcome	All cases (*n*=59), *n* (%)	OBS (*n*=20), *n* (%)	OCS (*n*=21), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=6), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
Transmission	43 (93)	16 (94)	14 (100)	1.00	9 (90)	1.00	0.416	4 (80)	0.411	0.263	1.00
Blockage of light	3 (7)	1 (6)	0		1 (10)			1 (20)			
Ultrasound density (*n*)	56	18	21		11			6			
Acoustically hollow	43 (77)	13 (72)	16 (76)	1.00	8 (73)	1.00	1.00	6 (100)	0.281	0.555	0.514
Acoustically solid	13 (23)	5 (28)	5 (24)		3 (27)			0			
Ultrasound surface (*n*)	53	16	21		10			6			
Smooth surface	34 (64)	13 (81)	13 (62)	0.284	4 (40)	0.046	0.441	4 (67)	0.585	1.00	0.608
Undulating surface	19 (34)	3 (19)	8 (38)		6 (60)			2 (33)			
Axial length (mm) (*n*)	31	6	15		9			1			
Mean (median, range)	22 (22, 18-25)	22 (22, 21-25)	22 (22, 18-25)	0.496†	22 (22, 19-23)	0.371†	0.889†	24	-	-	-
Magnetic resonance imaging (*n*)	16	3	7		5			1			
Choroidal effusion not visualized	5 (31)	1 (30)	4 (57)	1.00	0	0.375	0.080	0	1.00	1.00	1.00
T1 hyperintense	9 (56)	1 (30)	3 (43)	1.00	4 (80)	0.464	0.292	1 (100)	1.00	1.00	1.00
T2 hypointense	6 (38)	1 (30)	1 (14)	1.00	3 (60)	1.00	0.222	1 (100)	1.00	1.00	1.00
T1 fat suppression, gadolinium-enhanced	6 (38)	1 (30)	1 (14)	1.00	3 (60)	1.00	0.22	1 (100)	1.00	1.00	1.00

*Exclusion criteria for this subgroup analysis: (a) Concurrent diagnosis of scleritis (b) patients with no follow-up in our service, Fisher's exact test was used to calculate *P* value for categorical variables, †Student *t*-test was used to calculate *P* value. OBS: Observation, OCS: Oral corticosteroid, PCS: Periocular corticosteroid, TCS: Topical corticosteroid, VA: Visual acuity, VF: Visual field

Table 4: Uveal effusion syndrome with comparative response to corticosteroids versus observation: Analysis of outcomes in 59 cases*

Outcome	All cases (*n*=59), *n* (%)	OBS (*n*=20), *n* (%)	OCS (*n*=21), *n* (%)	*P* (OCS vs. OBS)	PCS (*n*=12), *n* (%)	*P* (PCS vs. OBS)	*P* (PCS vs. OCS)	TCS (*n*=6), *n* (%)	*P* (TCS vs. OBS)	*P* (TCS vs. OCS)	*P* (TCS vs. PCS)
VA outcomes** (*n*)	59	20	21		12			6			
Improvement	25 (42)	8 (40)	9 (43)	1.00	3 (25)	0.464	0.456	5 (83)	0.160	0.164	0.043
Stable	23 (39)	6 (30)	12 (57)	0.118	5 (42)	0.702	0.481	0	0.280	0.020	0.114
Worsening	11 (19)	6 (30)	0	0.008	4 (33)	1.00	0.012	1 (17)	1.00	0.222	0.614
Effusion outcomes (*n*)	59	20	21		12			6			
Effusion response (improvement)	46 (78)	13 (65)	19 (90)	0.067	8 (67)	1.00	0.158	6 (100)	0.145	1.00	0.245
Mean (median, range) months to improvement	7 (3, 1-66)	6 (4, 1-20)	10 (3, 1-66)	0.460†	3 (3, 1-6)	0.198†	0.294†	6 (4, 0-18)	0.987	0.618	0.077
Effusion complete resolution	29 (49)	10 (50)	13 (62)	0.535	4 (36)	0.470	0.157	3 (50)	1.00	0.661	0.626
Mean (median, range) months to resolution	54 (11, 1-289)	12 (5, 0-39)	67 (12, 1-289)	0.131†	77 (19, 3-267)	0.139†	0.879†	99 (18, 6-274)	0.089	0.673	0.194
Effusion worsening	5 (8)	1 (5)	1 (5)	1.00	3 (25)	1.00	1.00	0	1.00	1.00	1.00
Effusion recurrence	2 (3)	0	2 (10)	0.487	0	1.00	0.534	0	1.00	1.00	1.00
Mean (median, range) months to recurrence	162 (162, 8-316)	-	162 (162, 8-316)	-	-	-	-	-	-	-	-
Secondary treatment (*n*)	59	20	21		12			6			
No secondary treatment	33 (56)	18 (90)	3 (14)	<0.001	7 (58)	0.073	0.016	6 (100)	1.00	<0.001	0.114
Yes secondary (rescue) treatment	26 (44)	2 (10)	18 (86)		5 (42)			0			
Oral steroid	7 (12)	1 (5)	4 (19)	0.447	2 (17)	1.00	0.576	0	-	-	-
Periocular steroid injection	12 (20)	0	11 (52)	0.189	1 (8)	1.00	0.617	0	-	-	-
Topical steroid	1 (2)	0	0	1.00	0	1.00	0.353	0	-	-	-
Intravitreal steroid injection	2 (3)	0	1 (5)	1.00	1 (8)	1.00	0.504	0	-	-	-
Surgical management (scleral window)	3 (5)	0	2 (10)	1.00	1 (8)	1.00	1.00	0	-	-	-
Intravitreal anti-VEGF injection	1 (2)	1 (5)	0	0.100	0	0.285	1.00	0	-	-	-
Complications (*n*)	59	20	21		12			6			
Cataract	11 (19)	1 (5)	7 (33)	0.044	3 (33)	0.136	0.709	0	1.00	0.154	0.514
Steroid-induced glaucoma	0	0	0	-	0	-	-	0			

*Exclusion criteria for this subgroup analysis: (a) Concurrent diagnosis of scleritis (b) patients with no follow-up, **VA categories defined as: improved: Improvement in VA by ≥2 Snellen lines, Stable: Change in VA pre and posttreatment ≤1 Snellen line, Worse: Decline in VA by ≥2 Snellen lines, Fisher's exact test was used to calculate *P* value for categorical variables, †Student *t*-test was used to calculate *P* value. VEGF: Vascular endothelial growth factor, OBS: Observation, OCS: Oral corticosteroid, PCS: Periocular corticosteroid, TCS: Topical corticosteroid, VA: Visual acuity

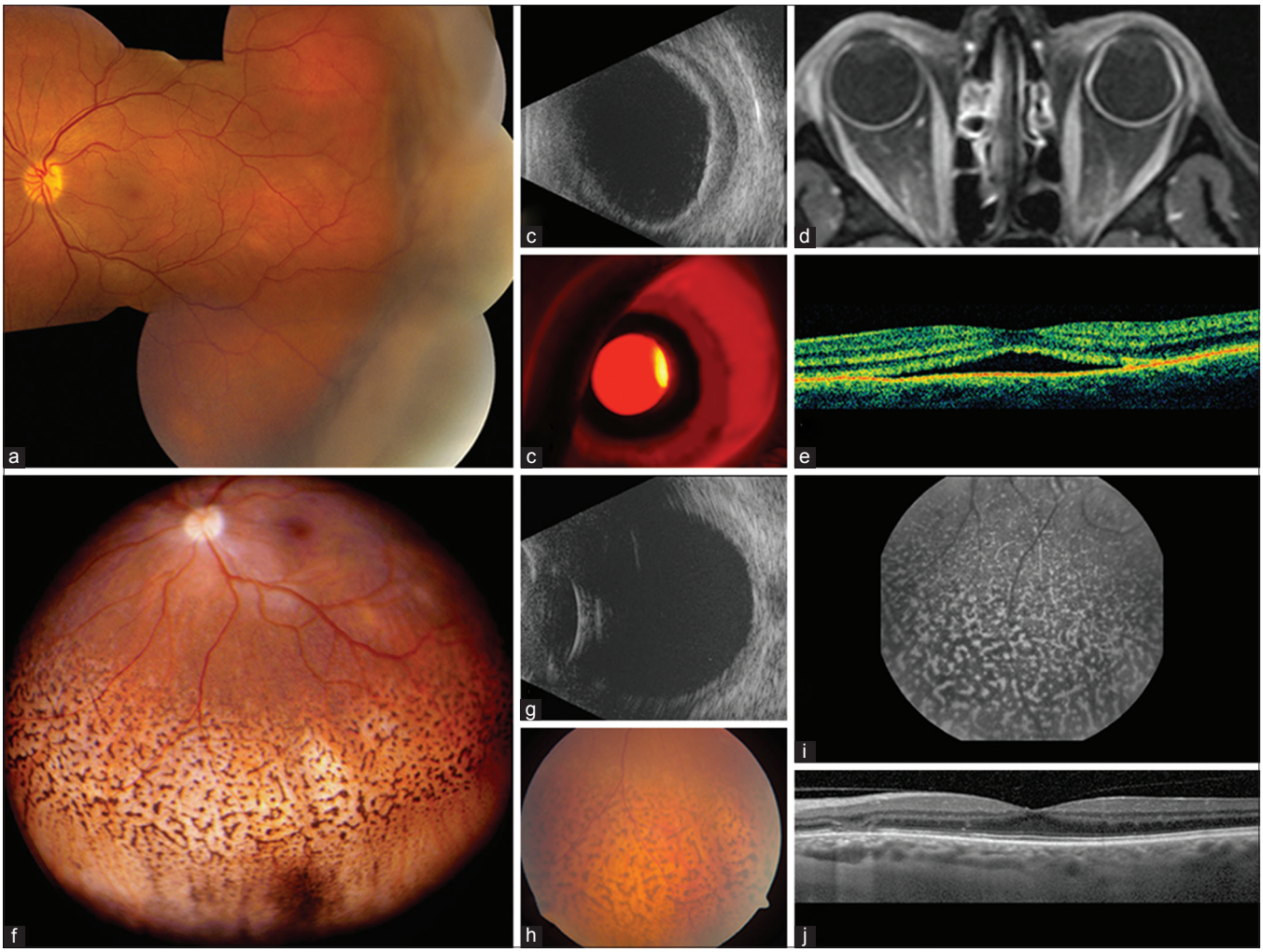

Figure 1: (a) Idiopathic uveal effusion, documented on (b) ultrasonography and demonstrating (c) no transillumination shadow. (d) Magnetic resonance imaging (T1) with gadolinium showing peripheral ciliochoroidal detachment and (e) optical coherence tomography confirmed subfoveal fluid. Following oral and periocular corticosteroids, there was (f) effusion resolution with retinal pigment epithelial mottling, (g) ultrasonographic flat retina, lipofuscin deposition (h) clinically and (i) on autofluorescence, (j) fluid resolution on optical coherence tomography, and final visual acuity of 20/40

of choroid, often extending for 360° in the peripheral uvea, lack of shadow on transillumination, angiographic lack of hyperfluorescence or "double circulation", ultrasonographic complete lucency and lack of intrinsic vascular pulsation, and MRI lack of gadolinium enhancement. This complex array of features can suggest uveal effusion, but the clinician should understand that underlying choroidal melanoma and metastasis can produce secondary choroidal effusion as a side effect, so the diagnosis can be enormously challenging.[27,28]

The management of UES typically has involved surgical sclerectomies (scleral window surgery), using partial or full-thickness scleral resection to allow for transscleral drainage of suprachoroidal and supraciliary fluid.[10-19] Based on published data, systemic or local corticosteroids are not often employed for the management. In one comprehensive review, the authors wrote that "treatment with systemic steroids does not appear to be effective" and "the most common treatment is full-thickness sclerotomies to provide an exit for choroidal fluid".[4] Surgical decompression of the vortex veins at their scleral emissary site or full-thickness sclerectomies has been the treatment of choice, particularly for uveal effusion related to nanophthalmos or scleral wall abnormalities, that is, those classified as Type 1 or Type 2 uveal effusion.[3,4] Type 3 uveal effusion typically does not respond to surgical scleral windows, according to Uyama *et al*.[3] Our experience with Type 3 UES, as shown in this report, is that there often is a response to systemic (oral), local (periocular or topical) corticosteroids or a combination of them, and some cases can show spontaneous resolution without intervention. In this analysis of 59 eyes with follow-up on our service, we were successful in controlling UES in 95% of cases and only needed to resort to scleral window surgery in 5%.

There are few reports in the literature that have evaluated a considerable number of case series of UES. In 2010, a major survey of published reports on UES highlighted the challenge in diagnosis, emphasizing that this condition can simulate central serous chorioretinopathy and choroidal melanoma.[4] In our series, most patients were referred for ocular oncology evaluation with suspicion of choroidal melanoma (47%), metastasis (3%), or unspecified tumor (34%), and only 16%

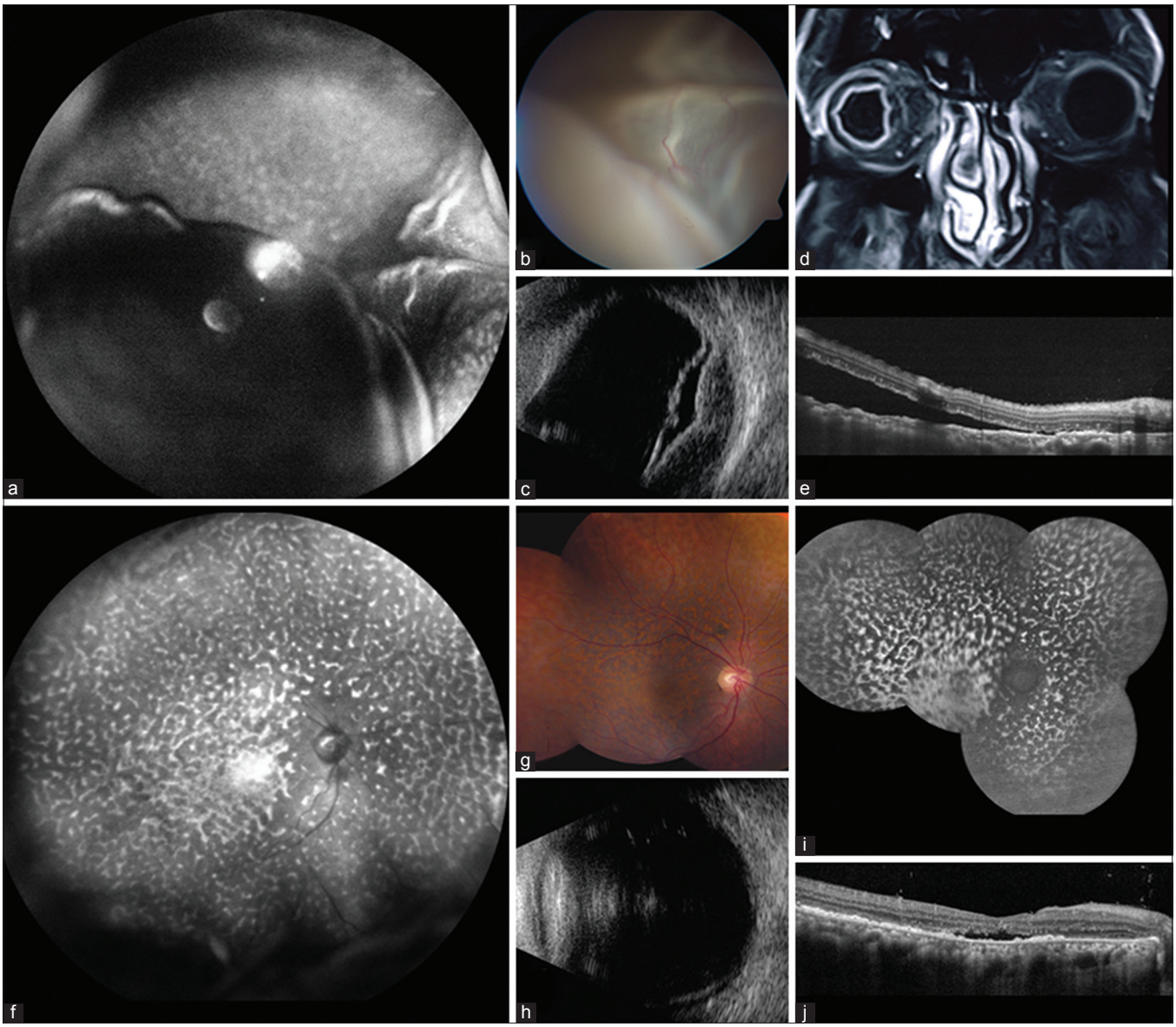

Figure 2: Uveal effusion by (a) wide-angle and (b) macular image, with (c) retinal and choroidal elevation on ultrasonography. (d) Magnetic resonance imaging (T1) with gadolinium, showed uveal detachment. (e) Optical coherence tomography confirmed submacular fluid. Following periocular and intravitreal corticosteroids, there was (f) fluid resolution with retinal pigment epithelial mottling, (g) flat retina, (h) confirmed on ultrasonography, and (i) lipofuscin deposition on autofluorescence. (j) Optical coherence tomography showed trace subfoveal fluid with outer retinal atrophy. Final visual acuity was 20/150

were correctly diagnosed with UES. Further points in that review suggested that this condition could be classified as hypermetropic/nanophthalmic versus idiopathic.[4] The suggested therapies included vortex vein decompression, sclerectomy or sclerotomy, long-term oral nonsteroidal anti-inflammatory drugs, or prostaglandin analogs to increase uveoscleral outflow.[4] These authors indicated that "treatment with high-dose systemic steroids has been described with rare success".[4] A more recent review on 19 eyes with UES suggested classification into three groups based on axial length and scleral thickness, including Type 1 (nanophthalmic eye, axial length <19 mm), Type 2 (nonnanophthalmic eye with abnormal thickened sclera documented on MRI), and Type 3 (nonnanophthalmic eye without scleral abnormality).[3] These authors found that sclerectomy was effective for Types 1 and 2, but not Type 3. They commented that Type 3 had no detectable abnormality, classified as truly "idiopathic" and represented two cases (11%) in their series, both with no effect from surgical sclerectomy and eventuating in VA of hand motions and 20/200.[3] In our study, we specifically evaluated Type 3 UES in nonnanophthalmic eyes and with normal sclera by ultrasound or MRI. We found that corticosteroids were beneficial for eyes with more advanced features, worse initial VA, or more extensive serous retinal detachment, and those with less advanced disease were managed conservatively with OBS, often with spontaneous resolution.

There has been one large case series studying medical management for uveal effusion, in 12 patients from a glaucoma center, in which 8 (67%) had angle closure and

6 (50%) had serous retinal detachment.[21] However, this case series did not represent true idiopathic UES as 3 (25%) had scleritis, 2 (17%) were drug-induced, 1 (8%) was related to Vogt–Koyanagi–Harada syndrome, and 1 (8%) had scleral amyloidosis. The authors indicated that only three patients had idiopathic UES. They noted seven patients (58%) responded to medical treatment, but this included an array of therapies and rarely corticosteroids alone. The three eyes with idiopathic UES were all treated with surgical sclerectomy.[21] They indicated that management of uveal effusion should consider the underlying cause, whether it be inflammatory that might respond to systemic corticosteroids, drug-induced that relies on cessation of the causative drug, or abnormal sclera-related effusion that necessitates surgical scleral thinning procedures.[21]

In our practice, we are typically referred patients with UES simulating an intraocular tumor and without underlying nanophthalmos or scleral abnormalities. We have found that medical treatment with corticosteroids (oral, periocular, topical, or a combination) can be successful, implying that there could be an underlying, subclinical mild inflammatory component, despite the absence of frank scleritis. In our series, 11% of eyes demonstrated low-grade pain, and 7% showed episcleral injection.

Conclusion

In summary, oral, periocular, topical, or a combination of corticosteroids can be considered for therapy of UES, in the absence of nanophthalmos and/or scleral thickness abnormalities. In this series, corticosteroids provided control of UES in 95% of cases. If control is not achieved, then consideration for scleral window surgery is an option.

Financial support and sponsorship

This study was financially supported by Eye Tumor Research Foundation, Philadelphia, PA (CLS, JAS).

Conflicts of interest

There are no conflicts of interest.

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