JAMA Ophthalmology Clinical Challenge

A Case of a Bumpy Retina

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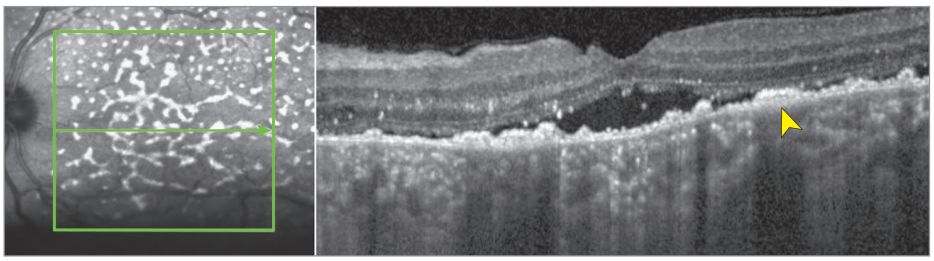
A Spectral-domain macular OCT



B Red-free ultra-widefield fundus image

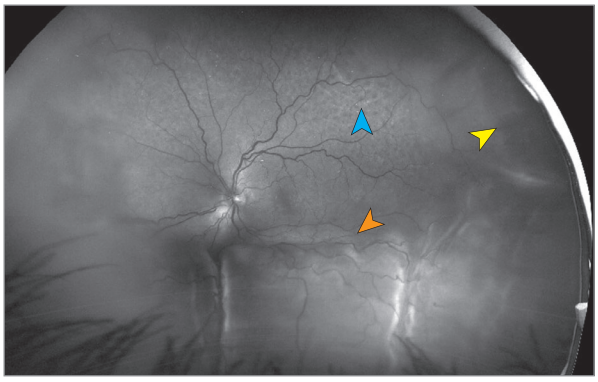


Figure 1. A, Spectral-domain macular optical coherence tomography of the left eye showing macula-involving subretinal fluid and nodular retinal pigment epithelium thickening (arrowhead). B, Red-free ultra-widefield fundus image of the left eye showing inferior subretinal fluid (orange arrowhead), leopard spotting pattern of pigmentary changes (blue arrowhead), and peripheral choroidal detachments (yellow arrowhead).

A 77-year-old male with history of hypertension and asthma presented with 3 months of progressive decline in vision in the left eye. He had no known ocular history. At presentation, best-corrected visual acuity was 20/100 OD and 20/50 OS. Intraocular pressures were normal bilaterally and pupils were equally reactive. Examination of the right eye was unremarkable except for moderate cataract. The left-eye slitlamp examination was remarkable for a shallow but quiet anterior chamber and moderate cataract. Dilated fundus examination of the left eye revealed shallow peripheral serous choroidal detachments, bullous inferior macula-involving retinal detachment with shifting fluid, and diffuse hyperpigmented lesions involving the macula and superior fundus that corresponded to areas of nodular retinal pigment epithelium thickening on optical coherence tomography (**Figure 1**). These hyperpigmented lesions were associated with hyperautofluorescence and blockage on fluorescein and indocyanine green angiography. No retinal breaks were seen on scleral depressed examination. Ultrasound biomicroscopy demonstrated 360° ciliochoroidal effusion. Axial length was 24.30 mm OD and 24.32 mm OS. The patient denied a history of known refractive error.

Results of inflammatory laboratory tests were negative for antineutrophil cytoplasmic antibodies, rheumatoid factor, and *Treponema pallidum* antibodies. Chest radiography and magnetic resonance imaging of the brain and orbits with and without contrast showed no extraocular disease. Treatment with 60-mg prednisone was initiated for 1 week but did not result in any changes in examination findings. The patient had a negative systemic review of systems.

WHAT WOULD YOU DO NEXT?

A. Scleral buckle with cryotherapy

B. Pars plana vitrectomy

C. Intravitreal steroid trial

D. Scleral windows surgery

CME Quiz at jamacmelookup.com

Diagnosis

Idiopathic uveal effusion syndrome

What to Do Next

D. Scleral windows surgery

Discussion

In the absence of a retinal tear, surgical options for rhegmatogenous retinal detachment (RD) repair (choices A and B) are inappropriate. Given no intraocular inflammation or response to oral steroids, choice C is unlikely to be of benefit. A diagnosis of uveal effusion syndrome (UES) was made, in which it is hypothesized that abnormal scleral thickness and/or composition decreases transscleral protein outflow and can compress vortex veins, leading to fluid accumulation in the choroidal and subretinal spaces.[1] Performing scleral windows surgery (choice D), which involves partial-thickness scleral excision in multiple quadrants, can improve transscleral outflow and help resolve the exudative RD.[1]

UES is a rare condition characterized by serous ciliochoroidal and RD associated with shifting subretinal fluid and leopard spotting.[1] UES has been classified into type 1, nanopthalmic eyes; type 2, non-nanophthalmic eyes with clinically abnormal sclera (defined by abnormal scleral thickness or histology); and type 3, non-nanophthalmic eyes with clinically normal sclera.[1,2] In a case series of 106 eyes from 66 Asian patients diagnosed with UES, the mean (SD) patient age was 35.9 (12.2) years for type 1 UES and 44.7 (8.2) years for type 2 UES. Fifty percent of affected patients were male.[3] UES in older individuals is uncommon, and this patient adds to the small number of published cases described in individuals 70 years or older.[4-6]

UES is a diagnosis of exclusion. Bullous central serous chorioretinopathy (CSCR) can present with shifting subretinal fluid and may mimic UES. Unlike UES, CSCR is associated with retinal pigment epithelium detachments, and steroid administration can exacerbate bullous CSCR.[7] Lack of inflammation or pain differentiates UES from inflammatory disorders such as posterior scleritis. Hypotony and medication adverse effects can mimic UES, but these did not apply to this patient. UES can be mistaken for choroidal melanoma or other neoplasm, but reassuring features for UES can include nonpigmented choroidal detachment often extending 360°, absence of orange pigment, lack of transillumination shadow, and absence of intrinsic vasculature on B-scan ultrasonography.[1] Bilateral diffuse uveal melanocytic proliferation is usually bilateral and tends to have red-orange lesions that are associated with hyperfluorescence on fluorescein angiography and hypoautofluorescence on fundus autofluorescence, in contrast to this patient's findings.[8]

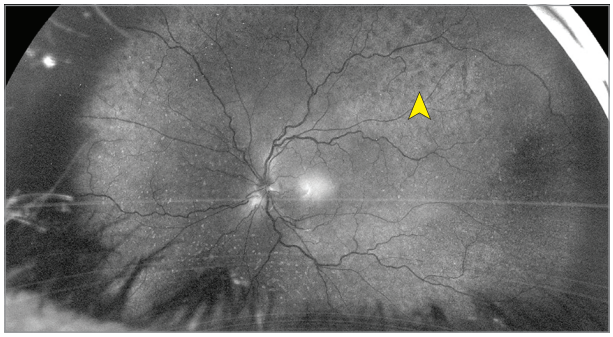


Figure 2. Red-free ultra-widefield fundus image of the left eye showing persistent leopard spotting pattern (arrowhead) but resolution of subretinal fluid and choroidal detachment after successful scleral windows surgery.

Although some authors have reported improvement of UES with steroids, acetazolamide, or latanoprost, surgery is often indicated for vision-threatening cases.[6,9] Scleral windows are typically created in 2 or more quadrants,[2] and serial follow-up initially at least every couple months is indicated to assess for RD resolution and possible recurrence.[10] Surgical management may be more effective for types 1 and 2 than type 3 UES.[2]

Patient Outcome

This patient underwent partial-thickness scleral windows in all 4 quadrants. Intraoperatively, the sclera was abnormally thick, suggestive of type 2 UES. Histopathology of the excised sclera showed patchy calcifications and glycosaminoglycan deposits and was negative for amyloid Congo red staining. Complete retinal reattachment was achieved by postoperative month 3 (**Figure 2**). After cataract surgery, best-corrected visual acuity was 20/30.

ARTICLE INFORMATION

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