Brief Report

Bilateral Diffuse Uveal Melanocytic Proliferation With Multiple Iris Cysts

Anthony Joseph, MD; Ehsan Rahimy, MD; David Sarraf, MD

Bilateral diffuse uveal melanocytic proliferation (BDUMP) is a rare paraneoplastic ocular syndrome with characteristic findings, including exudative retinal detachment, rapid cataract formation, and uveal melanocytic tumors. We report a case notable for bilateral iris and ciliary body cysts—a rare presentation of the disease.

OBSERVATIONS A woman in her 50s presented with bilateral decreased vision. Her medical history was significant for clear cell adenocarcinoma of the endometrium. Slitlamp examination revealed a contiguous ring of pigmented translucent iris cysts at the pupillary margin of each eye, confirmed with ultrasound biomicroscopy. Ophthalmoscopic examination of the left eye showed a geographic patch of subretinal fluid temporal to the macula that was associated with orange polygonal pigment. The patient underwent periocular injection of triamcinolone acetonide, with resolution of the subretinal fluid. Recurrent fluid was treated successfully with a second injection of triamcinolone.

CONCLUSIONS AND RELEVANCE Our case of BDUMP appears to be the first to demonstrate multiple iris and ciliary body cysts in high-quality color photography and ultrasound biomicroscopy. Involvement of the anterior uveal tract may be more common than reported in the literature because of its occult nature. Ultrasound biomicroscopy and anterior segment optical coherence tomography may be useful in patients with suspected BDUMP to identify anterior uveal tract involvement.

Report of a Case

A woman in her 50s presented with a 3-month history of bilateral decreased vision, worse in the right eye. Her medical history was significant for clear cell adenocarcinoma of the endometrium, initially diagnosed 7 months before presentation with the vision problems and treated with surgical resection of ectopic endometrial tissue as well as chemotherapeutic administration of 4 cycles of carboplatin and docetaxel. Metastatic lymph nodes were subsequently detected, warranting a new chemotherapeutic protocol. Review of medications was otherwise noncontributory, and the patient denied any history of ocular surgery or ophthalmic disorders.

On presentation, the woman’s Snellen visual acuity was 20/30 OD and 20/25 OS. Slitlamp examination revealed an anterior chamber that was shallow peripherally but deep centrally in both eyes. With dilation, both lenses demonstrated advanced anterior and posterior subcapsular cataracts (Figure 1A and B). In addition, a contiguous ring of pigmented translucent iris cysts was readily visible at the pupillary margins of each eye (Figure 1A and B). Ultrasound biomicroscopy confirmed the diffuse presence of iris and ciliary body cysts, as well as 360° of ciliary body thickening in both eyes, with resultant narrowing of the anterior chamber angle (Figure 1C and D). Contact of the cysts with the anterior lens capsule was also noted (Figure 1C).

Ophthalmoscopic retinal examination showed nothing remarkable in the right eye, but in the left eye revealed a geographic patch of subretinal fluid temporal to the macula that was associated with orange polygonal pigment presumably cor-
responding to hypertrophied retinal pigment epithelium (RPE) and adjacent foci of RPE atrophy (Figure 2A and B). Fluorescein angiography showed blocking defects corresponding to the orange polygonal lesions and hyperfluorescent window defects corresponding with the foci of RPE atrophy (Figure 2C); fundus autofluorescence demonstrated the reverse finding (Figure 2D). Finally, spectral-domain optical coherence tomography (OCT) through the temporal lesion in the left eye revealed subneurosensory fluid localized to the area of pigment changes described above as well as an irregular and thickened RPE (Figure 2E). B-scan ultrasonography excluded any underlying mass in either eye. On examination, no additional subretinal fluid was noted in the inferior periphery.

Approximately 1 month after presentation, the patient underwent periocular injection of triamcinolone acetonide, 40 mg, in the left eye. Enhanced depth imaging OCT performed before the injection showed persistent subretinal fluid and irregular RPE thickening, as described above (Figure 3A), and marked thickening of the choroid. Follow-up enhanced depth imaging OCT performed approximately 2 weeks later demonstrated nearly complete resolution of the fluid (Figure 3B), although choroidal thickness was essentially unchanged. Approximately 5 months after the initial injection in the left eye, the patient experienced a massive recurrence of subretinal fluid in the macula (Figure 3C), so an additional periocular injection of triamcinolone acetonide, 40 mg, was performed with significant reduction of subretinal fluid and persistent choroidal thickening noted at the 2-week (Figure 3D) and 1-month (Figure 3E) follow-up visits. Intraocular pressures in the left eye were consistently within normal limits and were essentially unchanged after the corticosteroid injections.

Discussion

With BDUMP first described by Machemer in 1966,2 more than 50 cases have since been reported in the literature. Patients are typically in the sixth to ninth decades of life and often present before diagnosis of a systemic malignant neoplasm.3 Cancers associated with BDUMP usually involve the reproductive tract in women (ovarian, uterine, or cervical cancer), whereas men are likely to have underlying lung, pancreatic, or colon cancer.4

Although our patient fit the usual demographic profile for BDUMP and her presentation included all 5 cardinal signs as described by Gass et al,1 her case was unique. Recently, Navajas et al5 described a patient very similar to ours with clear cell adenocarcinoma of the endometrium, characteristic RPE changes, and progressive iris nevi in both eyes. By comparison, our case was notable for the abundance of prominent pigmented iris and ciliary body cysts and diffuse thickening of the ciliary body and choroid in each eye. Iris and ciliary body cysts in association with BDUMP have rarely been reported,2,6-8 and the descriptions were published decades ago when imaging ca-
Figure 2. Multimodal Imaging of the Retina

A. Color fundus montage of the left eye showing a geographic patch of subretinal fluid temporal to the macula associated with orange polygonal pigment. B. Color fundus photograph of the left eye centered on this lesion. C. Fluorescein angiography of the left eye demonstrating blocking defects corresponding to the orange polygonal lesions and hyperfluorescent window defects corresponding to foci of presumed retinal pigment epithelium (RPE) atrophy. D. 30° fundus autofluorescence (Heidelberg Retina Angiograph; Heidelberg Engineering) demonstrating intensified levels of autofluorescence corresponding to the orange polygonal lesions and complete loss of the expected RPE autofluorescence pattern corresponding to foci of presumed RPE atrophy. E. Spectral-domain optical coherence tomography (OCT) through the temporal lesion in the left eye revealing subretinal fluid and an irregular and thickened RPE. The green line in the near infrared image is registered with the adjacent OCT.

Capabilities were less advanced. Some of these reports\textsuperscript{6,8} included low-resolution external photographs and abnormal tissue samples, but ours appears to be the first to demonstrate these findings with high-quality color photography and ultrasound biomicroscopy. It is possible that involvement of the anterior uveal tract is more common in BDUMP than previously...
Figure 3. Relationship of Subretinal Fluid to Periocular Corticosteroid Injections

A, Enhanced depth imaging optical coherence tomography (EDI-OCT) of the left macula on presentation. The previously noted area of subretinal fluid and irregularly thickened retinal pigment epithelium (Figure 2E) are seen temporally. B, EDI-OCT of the left macula 2 weeks after injection of periocular triamcinolone acetonide, 40 mg. The temporal subretinal fluid has almost completely resolved. C, EDI-OCT of the left macula 5 months after the initial injection of periocular triamcinolone shows a large neurosensory detachment. D, EDI-OCT of the left macula 2 weeks after a second injection of periocular triamcinolone acetonide, 40 mg, showing a decrease in subretinal fluid. E, EDI-OCT of the left macula 1 month after the second injection of periocular triamcinolone shows further reduction in subretinal fluid, especially temporally. The thickened choroid at each follow-up remains grossly unchanged with injection (about 500 μm). The green line in the near infrared image is registered with the adjacent OCT.
reported because of its occult nature. Ultrasound biomicroscopy and anterior-segment OCT may be useful in patients with suspected BDUMP to identify iris and ciliary body cysts, which may contribute to the rapid progression of cataracts in this population and also increase the risk of angle-closure glaucoma.

Another notable feature of this case was the marked reduction of the exudative detachment with periocular injection of triamcinolone. This finding carries potentially useful implications given the poor visual prognosis for patients with BDUMP. Reports of systemic corticosteroid therapy have shown highly variable results, but there is a paucity of information in the literature regarding topical or periocular corticosteroid therapy. Other interventions, such as ocular radiotherapy, drainage of subretinal fluid, and treatment of the underlying neoplasm, have variable and transient results, and the benefit of cataract surgery is often limited by the underlying retinal abnormality. Recent reports suggest that plasmapheresis may stabilize vision in patients with BDUMP, but it is a resource-intensive process that requires a healthy patient. Given the poor prognosis for patients with BDUMP, both systemically and locally, periocular corticosteroid injections may provide an immediate adjunctive intervention in those too ill to undergo plasmapheresis or other more-invasive therapies.

ARTICLE INFORMATION

Submitted for Publication: August 28, 2013; final revision received January 18, 2014; accepted January 22, 2014.

Published Online: April 24, 2014. doi:10.1001/jamaophthalmol.2014.311.

Author Contributions: Drs Joseph and Sarraf had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Sarraf. Acquisition, analysis, or interpretation of data: All authors. Drafting of the manuscript: Joseph, Rahimy. Critical revision of the manuscript for important intellectual content: Rahimy, Sarraf. Study supervision: Rahimy, Sarraf.

Conflict of Interest Disclosures: None reported.

Correction: This article was corrected on April 30, 2014, to fix triamcinolone acetone dosage errors.

REFERENCES