Bilateral secondary angle-closure glaucoma and ciliochoroidal effusion as an initial manifestation of systemic lupus erythematosus

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Uveal effusion with secondary angle-closure glaucoma (SACG) has been described as an initial manifestation in myelodysplastic syndrome, IgA nephropathy, and human immunodeficiency virus. Nanophthalmic eyes, with thick abnormal sclera and short anteroposterior diameters are predisposed to uveal effusion with SACG. Few retinal interventions like scleral buckle, pan-retinal photocoagulation, and ocular conditions like central vein occlusion, arteriovenous fistula, uveitis, hemorrhagic ciliochoroidal detachments, and posterior scleritis can also result in uveal effusion and angle closure. We report a rare instance of bilateral SACG and ciliochoroidal effusion as an initial manifestation of SLE; only few case reports have been published so far. Our patient is the youngest one diagnosed to have lupus-associated ACG and ciliochoroidal effusion in the published literature so far.

A 19-year-old female presented to us with acute onset defective vision in both eyes (OU) for the past 5 days. Her best-corrected visual acuity (BCVA) OU was 6/6 with −4 D spherical correction, and intraocular pressure (IOP) by applanation tonometry was 24 mmHg OD and 22 mmHg OS. Anterior segment evaluation OU was unremarkable except for lid edema and shallow peripheral anterior chamber depth of VH1 using Van Herick’s method [Fig. 1a]. Gonioscopy OU showed 360 degrees of appositional angle closure without any peripheral anterior synechiae. Undilated fundus examination OU showed normal disc with internal limiting membrane folds (ILM) folds in the macula [Fig. 1b] which was confirmed on OCT image [Fig. 1c]. B-mode ultrasonogram (USG) OU revealed 360 degree choroidal effusion [Fig. 1d]. Ultrasound biomicroscopy (UBM-Accutome by Keeler plus) OU showed anterior rotation of the ciliary body with crowding of the angle structures and suprachoroidal effusion [Fig. 1e and f]. Fundus fluorescein angiography OU was done to exclude lupus choroidopathy which was unremarkable.

Detailed probing of the history revealed that she had had angioedema-like bilateral lid swelling and skin rash over the face two weeks ago for which she consulted a physician. Blood investigations including hemoglobin, random blood sugar, serum urea and creatinine were within normal limits, whereas spot urine test showed increased urine protein-creatinine ratio (250 mg/g). Her antinuclear antibody (ANA) titre levels were positive (1:100). USG abdomen showed moderate ascites and grade 1 medical renal disease. Additionally, chest computed tomography revealed bilateral pleural effusion. Hence, the patient was diagnosed with systemic lupus erythematosus (SLE) with lupus nephritis and polyserositis based on ACR criteria (American College of Rheumatology), and started on intravenous methylprednisolone sodium succinate 500 mg in 1000 ml normal saline twice daily for 5 days, followed by oral prednisolone 30 mg once daily and oral mycophenolate mofetil 500 mg twice daily. There was no other drug intake which could potentiate SACG in our patient. Hence, she was diagnosed to have lupus-associated ciliochoroidal effusion and SACG of non-pupillary block mechanism. She was started on aqueous suppressant (timolol maleate 0.5%) and advised to continue the treatment given by the nephrologist.

At the one-month review visit after treatment with topical aqueous suppressant, systemic steroids and immunosuppressants, her uncorrected visual acuity OU improved to 6/6 without any myopic refraction with normal IOP of 14–16 mmHg OU. B-mode USG and UBM OU showed completely resolved suprachoroidal and choroidal effusion with deepening of the anterior chamber depth [Fig. 1g and h]. During the subsequent year, the secondary acute angle closure did not recur and renal parameters remained under control.

Discussion

SLE is a multisystemic, chronic, idiopathic, autoimmune disease caused by the combination of auto-antibodies and abnormally activated immune complexes. Ocular manifestations of SLE include blepharitis, keratoconjunctivitis sicca, scleritis, retinal vascular disease, choroidopathy, and optic neuropathy. Uveal effusion with secondary glaucoma can be a presenting sign for SLE and is usually accompanied by polyserositis and...
The anterior rotation of the ciliary body, thickening of the lens, supraciliary space from the choroidal capillaries. Ultimately, complex deposition would have caused fluid to enter the edema of ciliary body and choroid resulting from immune response. The disease activity and renal involvement. A previous study reported that four of 28 patients with choroidopathy died from lupus-related complications. Prompt and aggressive immunosuppressive therapy should be used. The limitation of this case report is that indocyanine green angiography (ICG), which is the primary investigation of choice to exclude choroidal lesions, was not done in our patient.

Most of the patients can be effectively treated using topical glaucoma medications and intense immunosuppressive therapy. Partial-thickness sclerectomies, linear sclerostomies, and choroidal drainage may be needed in individuals with persistent choroidal effusions.

Individuals with systemic lupus erythematosus can develop ciliochoroidal effusion, which can lead to acute secondary angle-closure glaucoma, and this manifestation is often accompanied by polyserositis and nephritis. Meticulous systemic evaluation is warranted if antecedent ocular abnormalities are not present while assessing a patient with uveal effusions and secondary angle-closure glaucoma. The dose of oral steroids and mycophenolate should be rationalized according to body weight and appropriate liver function test monitoring. Hence, the treating ophthalmologists should be aware of this rare manifestation of SLE. Prompt referral to a rheumatologist and aggressive high doses of steroids and immunosuppressive therapy are strongly recommended.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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