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Cystoid macular edema in a child

Case

An 11-year-old Caucasian boy with a 9-month history of blurred vision in his right eye (OD) was found to have cystoid macular edema (CME) OD that failed to respond to Nepafenac eyedrops. Family and systemic history was unremarkable. Upon referral, best corrected visual acuity (BCVA) was 20/25 OD and 20/20 in his left eye (OS). Anterior segment was quiet and intraocular pressure was 12 mmHg in each eye (OU). Fundus examination of the left eye was normal. The right eye demonstrated tiny parafoveal pinpoint exudation and meticulous examination revealed subtle vascular misdirection with looping along the superonasal periphery [Fig. 1a, blue arrow]. There was no vitritis or retinitis.

What is Your Next Step?

A. Laboratory workup for uveitis
B. Laser photocoagulation to the exudation
C. Fundus fluorescein angiography (FFA)
D. Genetic analysis for von Hippel Lindau disease.

Findings

Fluorescein angiography demonstrated superonasal peripheral nonperfusion (white arrows) with tortuous looping vessels, telangiectasia (yellow arrows), and light bulb microaneurysms (blue arrows) [Fig. 1b and c]. There was additional mild leakage in the inferonasal foveal region. Optical coherence tomography (OCT) confirmed the presence of intraretinal macular exudation (white arrow) and mild, noncystoid edema in the papillomacular bundle [Fig. 1d]. Focal cystoid macular edema was noted inferior to the foveola (figure not shown). These findings were suggestive of Coats disease stage 2B with parafoveal exudation, a relatively mild stage. The patient was treated with laser photocoagulation to the telangiectasia and microaneurysms.

Diagnosis

Coats disease Stage 2B.

Correct Answer: C.

Discussion

Coats disease is an idiopathic retinal condition characterized by vascular telangiectasia, exudation, and subretinal fluid, mostly affecting young males.[1] Meticulous peripheral fundus examination and confirmatory diagnostic testing with FFA are necessary due to its resemblance to other vascular and exudative retinopathies.[2] This condition is classified into 5 stages based on clinical features of (1) telangiectasia, (2) exudation, (3) subretinal fluid, (4) glaucoma, and (5) phthisis bulbi.[1] Management options, based on staging, include laser photocoagulation, cryotherapy, external drainage of retinal detachment, and, in advanced end-stage, enucleation.[1,3] Outcomes reveal visual acuity ≥ 20/40 in stage 1 (100%), stage 2 (33%), stage 3 (20%), stage 4 (0%), and stage 5 (0%).[3] Therefore, earlier detection of Coats disease at a less advanced stage would allow improved visual outcomes.

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.

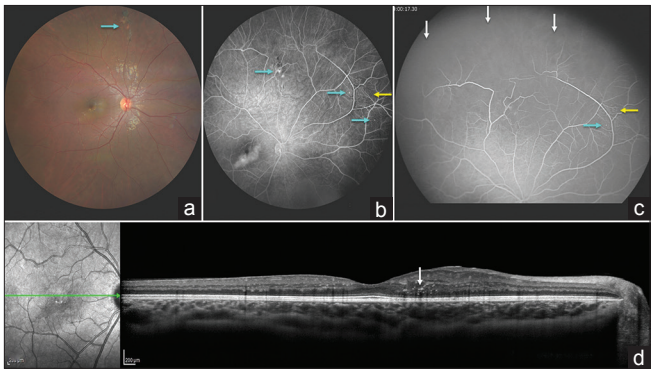

Figure 1: Fundus photography of the right eye demonstrated tiny parafoveal pinpoint exudation and subtle vascular misdirection with looping along the superonasal periphery [Fig. 1a, blue arrow]. Fluorescein angiography demonstrated superonasal peripheral nonperfusion (white arrows) with tortuous looping vessels, telangiectasia (yellow arrows), and light bulb microaneurysms (blue arrows) [Fig. 1b, c]. Optical coherence tomography confirmed the presence of intraretinal macular exudation (white arrow) and mild, noncystoid edema in the papillomacular bundle [Fig. 1d]

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Nil.

Conflicts of interest

There are no conflicts of interest.

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