# Macular Star, Optic Disc Swelling, and Lethargy in a Child

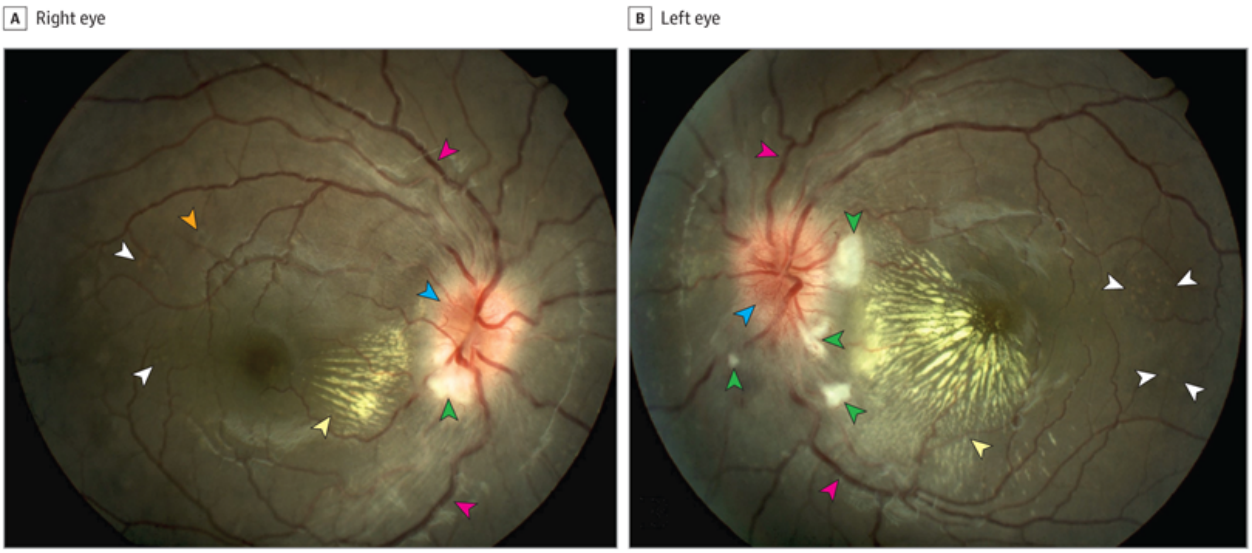
Case

A 13-year-old girl with a 7-day history of painless vision loss and central scotoma in her left eye was referred to the department of ophthalmology. Three weeks prior, she had presented with mild fever and flulike symptoms, associated with a severe frontal headache that was mildly relieved by analgesics. Her medical history was positive for relapsing urinary tract infections since early infancy without adequate follow-up. Findings of an ophthalmological examination 1 year prior were reported as normal.

At initial examination, she was conscious but somewhat lethargic. Her best-corrected visual acuity was 20/20 OD and 20/80 OS. Pupillary light responses, extrinsic ocular motility, anterior segment biomicroscopy, and intraocular pressure were normal in both eyes. Dilated fundus examination revealed a bilateral sectorial macular star that was more extended in the left eye, with retinal veins slightly dilated and tortuous, attenuated arterioles, some juxta and peripapillary nerve fiber layer infarcts, and optic disc edema with marked papillary telangiectasia; in addition, some faint, small, tan-yellow dots were observed at the level of the retinal pigment epithelium in the posterior pole, some of them already grayish and with a hypopigmented halo (Figure 1). Optical coherence tomography displayed elevated optic discs and hyperreflective material in the outer plexiform layer temporal to the optic disc in both eyes and a macular neurosensory detachment in the left eye. Results of visual field testing were normal in the right eye and

showed a central scotoma in the left eye.

Figure 1.




Fundus image of the right eye (A) and left eye (B) showing slight dilation of retinal veins (magenta arrowheads), intraretinal lipid deposits in the papillomacular bundle and the inferonasal macular quadrant forming an incomplete macular star (yellow arrowheads), optic disc edema and epipapillary telangiectasia (blue arrowheads), nerve fiber layer infarcts or cotton-wool nodules (green arrowheads) over the inferior margin of the optic disc in the right eye and juxta and parapapillary in the left eye, and paramacular temporal light-pigmented subretinal dots at the level of the retinal pigment epithelium (white arrowheads) in both eyes, with some already scarred (pigmented dot surrounded by a pale halo; gold arrowhead).

**What Would You Do Next?**

1. Prescribe systemic corticosteroids
2. Order serologic testing for infectious neuroretinitis
3. Rule out brain tumor

4. Check blood pressure

Discussion

Diagnosis

Hypertensive retinopathy

What to Do Next

D. Check blood pressure

Discussion

Option D is the correct approach. The child underwent immediate hospitalization and systemic evaluation. Clinical examination revealed severe systemic hypertension (225/160 mm Hg). Laboratory test results showed leukocytosis, elevated erythrocyte sedimentation rate (35 mm), hypercholesterolemia, hypokalemia, proteinuria, and microalbuminuria.

Echocardiography showed left ventricle hypertrophy, and kidney doppler ultrasonography revealed advanced bilateral kidney atrophy with parenchymal scarring that was presumed to be the consequence of undertreated relapsing urinary tract infections. Results of magnetic resonance imaging of the brain and orbits were normal.

The most common masquerader for neuroretinitis is malignant hypertension.[1] Systemic hypertension in children is rare, potentially underrecognized,[2] and its prevalence ranges from 2% to 5%.[3] Mostly asymptomatic, pediatric hypertension may manifest by symptoms originating from target organ disease, and most cases present secondary hypertension from kidney disorders, especially kidney parenchymal disease.[4] Hypertensive retinopathy occurs in 8% to 18% of children with severe hypertension, figures substantially lower

than those observed in adults with hypertension, with the highest prevalence in those with kidney and renovascular disease.[5]

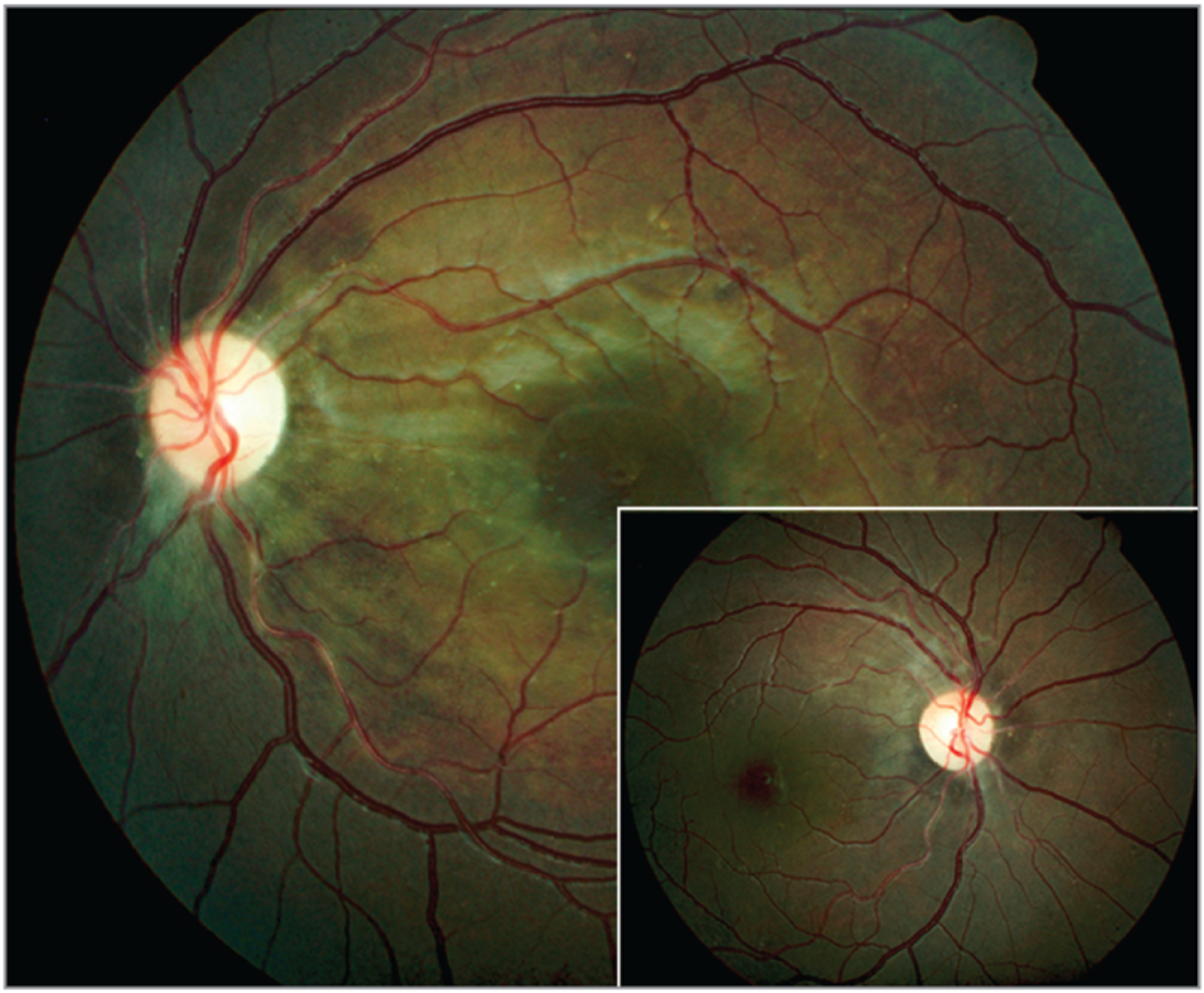
Malignant hypertension is an acute, severe rise of more than 180 mm Hg in systolic pressure and/or more than 120 mm Hg in diastolic pressure, associated with severe hypertensive retinopathy and papilledema. It constitutes a medical emergency, where severe acute increase in blood pressure results in end-organ damage.[6] The bilateral retinal disease, the presence of nerve fiber layer infarcts and Elschnig spots (focal signs of choroidal ischemia at the level of the retinal pigment epithelium), and the accompanying neurologic symptoms (severe headache and lethargy) suggested a severe systemic disease and were the presenting signs of malignant hypertension, which is a life-threatening condition in this patient secondary to kidney parenchymal disease from undertreated relapsing urinary tract infections since early infancy.

Option A is incorrect. It is not advisable to prescribe corticosteroids before ruling out an infectious condition. Also, corticosteroids are contraindicated in the context of malignant hypertension. For option B, in the presence of the triad unilateral vision loss, macular star, and optic disc swelling in a child, infectious neuroretinitis must be ruled out, with cat scratch disease (CSD) its most frequent cause. However, neuroretinitis is a rare complication of cat scratch disease, occurring in only 1% to 2% of cases, and it generally presents unilaterally and self-limited.[1] In this patient, results of serum tests for infective causes were negative. Besides, retinal and optic nerve disease were bilateral, and the child presented with neurologic signs. Option C is incorrect. Although this patient had neurologic symptoms with bilateral optic disc edema, other features, like macular lipid deposits, nerve fiber layer infarcts, and Elschnig spots, are not clinical signs that accompany papilledema from intracranial hypertension.

## Patient Outcome

Treatment with amlodipine, 5 mg, every 12 hours, daily losartan, 50 mg, and a hyposodic diet reduced blood pressure drastically and slowly normalized the retinal disease. Visual acuity progressively improved in the left eye, as did the macular star, optic disc edema, and nerve fiber layer infarcts in both eyes. One year later, blood pressure was generally within normal values under treatment but with intermittent episodes of high values. Best-corrected visual acuity was 20/20 OD and 20/25 OS, and optic discs were nonedematous but moderately pale in both eyes (Figure 2). Optical coherence tomography performed during follow-up showed a diffuse thinning of the neurosensory retina and the peripapillary nerve fiber layer.

Figure 2.



Fundus image of the left eye (panel) and the right eye (insert) at 1-year follow-up showing a pronounced sclerotic appearance of arteries, a complete resolution of both optic discs' edema and the macular stars, a moderate optic disc pallor, and no evidence of nerve fiber layer infarcts.

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Article Information

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