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A Case of Acute Chest Pain After Acetazolamide to Treat Uncontrolled Increased Intraocular Pressure

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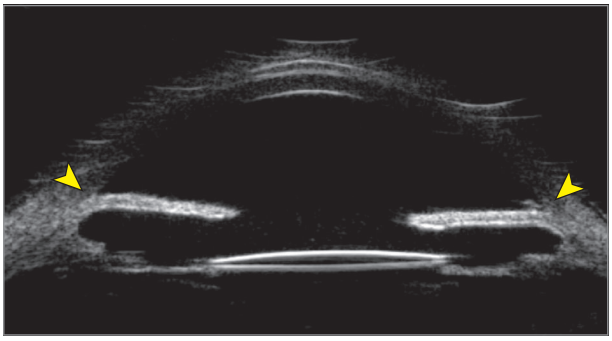

Figure 1. Ultrasound biomicroscopy of the left eye demonstrating angle closure (arrowheads).

A 61-year-old Black man with hypertension and diabetes presented with decreased vision 3 months after uneventful cataract surgery in his left eye. He had undergone laser retinopexy for retinal tears in his right eye and a scleral buckle and vitrectomy for a retinal detachment in his left eye 1 year earlier. Best-corrected visual acuity (BCVA) was 20/40 OD and counting fingers OS. Intraocular pressure (IOP) was 14 mm Hg in the right eye and 44 mm Hg in the left eye. Anterior segment examination of the right eye was unremarkable, while ophthalmoscopic examination showed vascular attenuation and treated retinal breaks. In the left eye, microcystic corneal edema and Descemet membrane folds limited examination, but no residual lenticular fragments or neovascularization of the iris or angle were seen. The patient was discharged taking timolol, dorzolamide, brimonidine, and prednisolone. Twelve days later, the cornea had cleared and IOP in the left eye improved to 32 mm Hg. Gonioscopy revealed 360° of anterior synechiae and complete angle closure, which was documented on ultrasound biomicroscopy (**Figure 1**). The patient started treatment with 500 mg of acetazolamide daily and referred to the glaucoma service for surgical evaluation.

When the patient returned for his glaucoma evaluation 3 days later, he was noted to have loss of appetite, drowsiness, nausea, and malaise. The patient's intraocular examination was stable bilaterally. During this visit, the patient developed acute chest pain and respiratory insufficiency requiring immediate transfer to the emergency department. He was found to have a hemoglobin level of 8.5 g/dL (to convert to grams per liter, multiply by 10.0) and a hematocrit level of 22.1% (to convert to a proportion of 1.0, multiply by 0.01).

WHAT WOULD YOU DO NEXT?

- **A.** Obtain blood cultures, perform vitreous tap, and inject antibiotics
- **B.** Perform laser iridotomy
- **C.** Observation
- **D.** Perform hemoglobin electrophoresis

Quiz at jamacmelookup.com

Diagnosis

Acute chest syndrome induced by acetazolamide in a patient with previously undiagnosed sickle cell disease.

What to Do Next

D. Perform hemoglobin electrophoresis

Discussion

Obtaining blood cultures, performing a vitreous tap, and injecting antibiotics (choice A) would be an appropriate choice of action in suspected endogenous endophthalmitis. However, while this patient had systemic symptoms, the examination did not demonstrate findings consistent with endophthalmitis, such as vitritis, chorioretinal infiltration, fibrin, a hypopyon, or conjunctival hyperemia. Laser iridotomy (choice B) would be useful if the patient demonstrated acute angle closure. While acute angle closure can cause nausea and vomiting, it would not be expected to cause acute chest pain or respiratory distress. Additionally, pupillary block was not found on examination. Finally, observation (choice C) is inappropriate in this systemically unstable patient.

Hemoglobin electrophoresis (choice D) is the correct answer in this patient who presented with symptoms of an acute sickling crisis (chest pain, respiratory distress) and decreased hemoglobin level after the initiation of acetazolamide, which is a carbonic anhydrase

inhibitor (CAI). The results of the hemoglobin electrophoresis test were consistent with hemoglobin SC disease.

Sickle cell disease (SCD) is a hemoglobinopathy that affects 9.3 per 100 000 people worldwide,[1] disproportionately affecting those of African descent such as this patient. The reported incidence of SCD is 1 in 365 Black individuals in the United States.[2] In patients with traumatic hyphema, a high index of suspicion is required to avoid oral CAI medications, which can induce sickling.[3,4] However, at-risk populations with glaucoma are not typically screened for SCD prior to initiation of treatment despite the potential for systemic complications. This patient harbored occult SCD that was exacerbated with the initiation of an oral CAI.

There are currently no management recommendations for populations at risk of SCD who require treatment for glaucoma. Acetazolamide increases hemoconcentration and blood viscosity, causing systemic acidosis that may induce sickling.[3] Methazolamide reduces the secretion of aqueous humor without altering the kidney secretion of bicarbonate, thus avoiding systemic acidosis and making it more appropriate for use by patients with SCD.[5] As shown in this patient who developed acute chest syndrome, consideration of hematologic screening or methazolamide use instead of acetazolamide may be appropriate in patients with a high risk of SCD. However, there is currently insufficient information, including whom to screen, to recommend this approach. More research on the medical management of glaucoma in patients at risk of SCD is warranted, including a cost-benefit analysis of routine screening prior to treatment.

Patient Outcome

Results of the workup for myocardial infarction, thrombosis, and occult blood loss were unremarkable. Hemoglobin electrophoresis testing revealed a hemoglobin S level of 49.5% [to convert to the proportion of total hemoglobin, multiply by 0.01] and hemoglobin C level of 42.3%. The patient was treated with blood transfusion and supportive care. After discharge, the patient returned to the glaucoma service with 20/400 BCVA and an IOP of 38 mm Hg in the left eye. Examination demonstrated microcystic corneal edema, neovascularization of the iris, and a 3-mm hyphema consistent with neovascular glaucoma. Widefield fluorescein angiography revealed extensive peripheral retinal ischemia that was worse in the left eye (**Figure 2**) than in the right eye, without clear neovascularization. The patient underwent intravitreal bevacizumab injection, implantation of a glaucoma drainage device, and panretinal photocoagulation. After 6 months of follow-up, the patient remained stable with a BCVA of 20/200 and IOP of 15 mm Hg in the left eye and had discontinued all glaucoma medications.

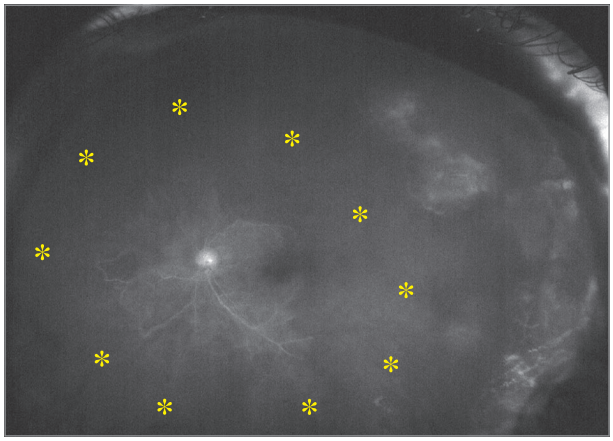

Figure 2. Ultrawidefield fluorescein angiography of the left eye demonstrating significant vascular nonperfusion (asterisks).

ARTICLE INFORMATION

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