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0196-0644/\$-see front matter

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doi:10.1016/j.annemergmed.2011.01.016



**Figure 1.** Right-eye proptosis.



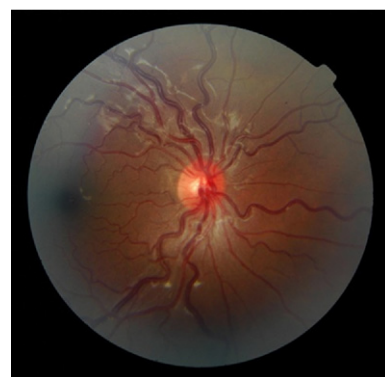
**Figure 2.** Episcleral injection and upper lid swelling.

[Ann Emerg Med. 2011;58:239.]

A 6-year-old girl presented to the emergency department with several months of progressive right-eye redness and swelling. There was no history of fever or respiratory symptoms. She denied pain or blurred vision but did report dizziness and intermittent diplopia. History was notable for nonpenetrating trauma to the right eye from a fishing pole 8 months ago. The result of a noncontrast brain magnetic resonance imaging (MRI) performed 3 months earlier for ongoing headaches was negative. Physical examination on presentation revealed proptosis of the right eye, with lid swelling and diffuse scleral injection (Figures 1 and 2). Extraocular movements were intact and visual acuity was normal. A bruit could be auscultated over the right globe. Computed tomography (CT) of the orbits was performed (Figure 3), followed by dilated fundusoscopic examination (Figure 4).



**Figure 3.** CT of the orbits, showing a dilated tortuous superior ophthalmic vein and engorgement of the right cavernous sinus.



**Figure 4.** Fundusoscopic examination of the right eye, showing dilated retinal vessels and a hyperemic disc. Used with permission of Susan C. Lipsett, MD, Department of Medicine, Children's Hospital Boston, Boston, MA.

*For the diagnosis and teaching points, see page 256.*

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### DIAGNOSIS:

*Carotid-cavernous fistula (direct type).* A carotid-cavernous fistula is an abnormal connection between the carotid artery and the cavernous sinus. A direct shunt between the artery and the cavernous sinus commonly results from traumatic disruption of the intracavernous carotid artery sheath. The resulting arterial flow can lead to rapid progression of symptoms and potential visual compromise.<sup>1</sup> Indirect shunts result from spontaneous rupture of small dural arteries, leading to low flow connections between branches of either the internal or external carotid artery and the cavernous sinus.<sup>2</sup> Carotid-cavernous fistulas are rare in the pediatric population.

Presenting symptoms can include progressive eye pain and swelling, pulsatile tinnitus,<sup>3,4</sup> headache, and diplopia. Examination findings may include unilateral proptosis, episcleral injection and chemosis, and lid swelling (Figures 1 and 2). Ocular motility may be limited, visual acuity decreased, and ocular pressure increased.<sup>1</sup> Auscultation of the globe may reveal a bruit. Rarely, intracranial hemorrhage or life-threatening epistaxis can occur.<sup>3</sup> Fundoscopic examination may reveal engorged retinal vessels (Figure 4), retinal hemorrhage, or optic disc swelling. CT and MRI will reveal dilation of the superior ophthalmic vein (Figure 3). The criterion standard is cerebral angiography.<sup>1</sup>

Direct fistulas are usually treated with embolization. Indirect fistulas may resolve spontaneously; therefore, a conservative approach with close monitoring is often favored.<sup>1,5</sup>

Our patient underwent diagnostic angiography and coil embolization of the fistula, with subsequent resolution of the proptosis and vascular engorgement.

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