Ophthaproblem

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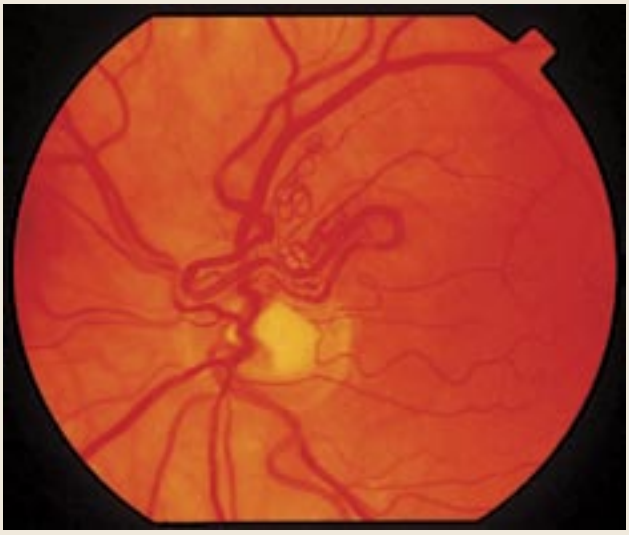

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A 67-year-old man was referred by his optometrist who had noticed an abnormal retinal vessel in the supratemporal quadrant of his left eye during a routine examination. The patient's visual acuity with correction was 6/6 bilaterally, and he had no complaints about his vision. Results of the remainder of his examination were within normal limits.

The appearance of his left fundus demonstrates which of the following?

1. Retinal neovascularization
2. Branch retinal vein occlusion
3. Retinal arteriovenous malformations (type 2)
4. Diabetic retinopathy

Answer on page 209

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3. Retinal arteriovenous malformations (type 2)

Retinal arteriovenous malformations (AVMs), first described by Magnus in 1874,[1] are rare, congenital, high-flow developmental vascular anomalies. The most important clinical sign noted was marked arterial and venous dilation associated with a tortuous pattern of vessels in a portion of the retinal circulation system. These retinal AVMs most frequently occur in the superotemporal arcade (41%), papillomacular bundle (34%), and sometimes in nasal areas of the optic disk (6%).[2] Classically, fluorescein angiography shows rapid filling of AVMs without leakage.

In 1973, Archer and co-workers summarized their experience and reviewed all published cases of congenital retinal AVMs. They divided the lesions into three groups.[1] Type 1 retinal AVM occurs when an arteriolar or abnormal capillary plexus is interposed between the major artery and vein. Type 2 retinal AVM is characterized by direct arteriovenous communication without intervening arteriolar or capillary segments. It is unusual to have intracranial AVM associated with either type 1 or 2 retinal AVM. Type 3 retinal AVM appears as widespread retinal AVMs that exist morphologically as large-calibre convoluted vessels. These lesions are frequently associated with AVM of the optic nerve, chiasm, and cerebral cortex. The association of retinal AVM with ipsilateral facial and intracranial structures is known as Wyburn-Mason syndrome.

Retinal AVMs are often seen in asymptomatic patients and are believed to be stable, unchanging lesions that pose little threat to vision. Occasionally, however, central retinal vein occlusion, vitreous hemorrhage, and retinal hemorrhage occur with these lesions.[3,4] Mansour et al[4] proposed a hypothesis to explain potential complications: AVMs could lead to decreased arterial pressure and increased venous pressure, causing the surrounding retina to be ischemic due to hypoperfusion from a steal phenomenon. In addition, retinal or vitreous hemorrhage can occur in areas of capillary ischemia or areas of increased hydrostatic pressure.

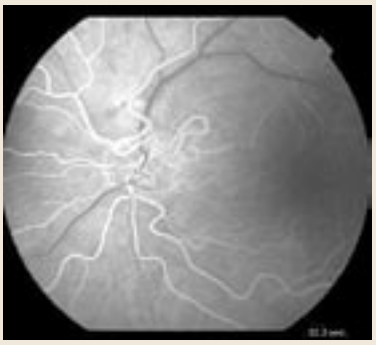

Figure 1. Early filling of retinal vein apparent on retinal angiography

Management

This patient was referred to an ophthalmologist. Examination of his dilated pupil revealed a tortuous supratemporal retinal artery and several tortuous arterioles at the disk. Retinal angiography demonstrated very early filling of the retinal vein (**Figure 1**), suggesting rapid dye inflow. No leakage was noted on later frames. Neuroimaging of the orbits and head did not reveal associated pathology.

Recommendation

Patients with tortuous retinal vessels should be referred to ophthalmologists on a semiurgent basis. Retinal arteriovenous malformation is a rare ophthalmic condition that could lead to loss of vision. All patients with retinal AVMs should have neuroimaging to rule out the possibility of orbital and intracranial AVM. If an intracranial vascular malformation is found, patients should be referred for neurosurgical evaluation.

References

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