A 9-year-old girl presented with sudden loss of consciousness and CT scan of the brain showed right thalamic hemorrhage with ventricular extension (figure 1). Carotid and vertebral angiographic study revealed a right thalamic arteriovenous malformation (AVM) extending into the right orbit. Ophthalmoscopy demonstrated extensive AVM involving the retina of the right eye (figure 2). A similar eye examination performed 1 year ago had revealed congenital retinal vascular anomalies, but no further investigation was arranged. Unilateral AVM involving the retina, brain, and sometimes skin constitutes Wyburn-Mason syndrome. Retinal AVM may signify concomitant intracranial AVM, which warrants detailed neurologic assessment.

Figure 1. CT scan of the brain shows intracerebral hemorrhage over right thalamic area with ventricular extension and hydrocephalus.

Figure 2. Ophthalmoscopy demonstrates extensive arteriovenous anastomoses involving the entire retina of the right eye. The anomalous vessels are extremely tortuous and congested. The venous stasis caused scattered retinal hemorrhage. The visual acuity of this eye was 20/70.

Wyburn-Mason syndrome

Wai-Man Chan, MRCP, FRCS, Nelson K.F. Yip, FRCS, Dennis S.C. Lam, FRCS, FRCOphth, Kowloon, Hong Kong

A 9-year-old girl presented with sudden loss of consciousness and CT scan of the brain showed right thalamic hemorrhage with ventricular extension (figure 1). Carotid and vertebral angiographic study revealed a right thalamic arteriovenous malformation (AVM) extending into the right orbit. Ophthalmoscopy demonstrated extensive AVM involving the retina of the right eye (figure 2). A similar eye examination performed 1 year ago had revealed congenital retinal vascular anomalies, but no further investigation was arranged. Unilateral AVM involving the retina, brain, and sometimes skin constitutes Wyburn-Mason syndrome. Retinal AVM may signify concomitant intracranial AVM, which warrants detailed neurologic assessment.


Address correspondence and reprint requests to Dr. Wai-Man Chan, Associate Professor, Department of Ophthalmology & Visual Sciences, The Chinese University of Hong Kong, Hong Kong Eye Hospital, 147K Argyle Street, Kowloon, Hong Kong; e-mail: cwm6373@netvigator.com
Wyburn-Mason syndrome
Wai-Man Chan, Nelson K.F. Yip and Dennis S.C. Lam
Neurology 2004;62;99
DOI 10.1212/01.WNL.0000099187.15025.97

This information is current as of January 12, 2004