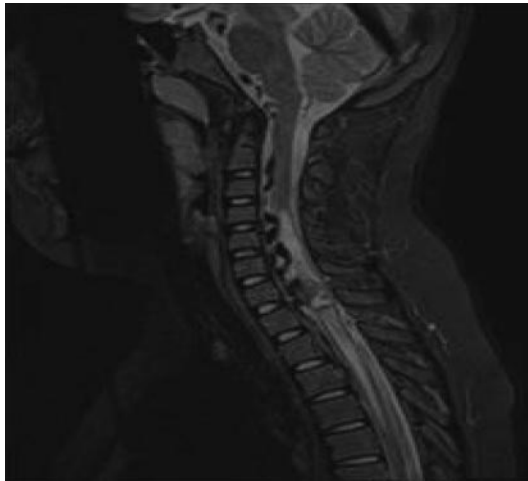


Teaching NeuroImages: Atypical Wyburn-Mason syndrome

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Figure 1 MRI



STIR fat-saturated T2-weighted sagittal MRI demonstrates cord expansion and edema (white signal from C4 to T4) secondary to spinal dural arteriovenous fistula. The serpiginous draining vein is seen anteriorly (black signal) extending cephalad.

A 10-year-old boy presented with sudden-onset left leg monoplegia. Imaging (figures 1 and 2) revealed complex vascular malformations involving the brain and spinal cord but ophthalmologic and skin examinations were normal. Wyburn-Mason syndrome is a rare phakomatosis characterized by unilateral arteriovenous malformations involving the retina, brain, and sometimes skin.^{1,2} We propose that Wyburn-Mason syndrome actually represents a continuous spectrum of disease, rather than a distinct syndrome, with some patients lacking the classic retinal and cutaneous findings. Even so, clinicians should be aware of the classic presentation and should assess any patient with a complex unilateral intracranial vascular malformation for retinal involvement.

Figure 2 Magnetic resonance angiography



Three-dimensional time-of-flight magnetic resonance angiogram demonstrates arteriovenous fistula (arrow) between abnormally dilated right posterior cerebral artery and vein of Galen.

AUTHOR CONTRIBUTIONS

Drs. Madey, Lehman, and Chaudry and S. Vaillancourt have participated sufficiently in the work to take responsibility for content. All made substantial contributions. Dr. Jay Madey was responsible for drafting the original manuscript and was directly involved with the patient care, diagnosis, and management. Dr. Rebecca Lehman was responsible for revising the manuscript for content and was directly involved with the patient and involved in the interpretation and analysis of the data. Dr. Imran Chaudry contributed to the original manuscript by submitting figures as well the interpretation of the data. Shannon L. Vaillancourt was responsible to revision and submission of the manuscript and was directly involved with the management of the patient.

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The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

Neurology®

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Neurology 2012;79:e84

DOI 10.1212/WNL.0b013e318268460f

This information is current as of September 3, 2012

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