Wyburn-Mason Syndrome and Management of Cerebral AVMs during Labor

Abstract Type: Case Report/Case Series
Patrick W. Clark, M.D.; Jamie Murphy, M.D.
The Johns Hopkins University

Introduction: Wyburn-Mason syndrome (WMS) is a congenital neurocutaneous disorder comprised of ipsilateral cerebral arteriovenous malformations (CAVMs), vascular abnormalities of the visual pathway, and lesions in the oronasopharyngeal area. WMS in a parturient raises specific anesthesia related concerns.

Case Report: Patient was a 36 y/o female G1P0 at 38wks gestation with a history of a left sided retinal AVM extending from the retina to the superior midbrain. In 1983 she was diagnosed with Wyburn-Mason syndrome and in 1994 suffered an intracranial hemorrhage requiring prolonged intubation, resulting right sided hemiparesis, facial droop, and short term memory loss. Since, she has had decreased vision in the left eye and was found to have central retinal vein occlusion. Upon presentation, she denied changes in her neurological exam. Given the increased risk of CAVM rupture with acute hypertension, it was determined that a C-section would be performed under epidural anesthesia with awake arterial line placement. A combined spinal-epidural technique was rejected because of the potentialdelay in detecting epidural malfunction, the risk of post-dural puncture headache confusing the neurologic exam, and the risk of CSF leak. An MRI of the spine was obtained to rule out AVM. The patient received a L3-4 epidural. Systolic BP was tightly regulated to 90-120mmHg. She delivered a healthy infant with Apgars of 9/9. She had an uncomplicated recovery.

Discussion: The overall risk of AVM hemorrhage during pregnancy is controversial. Some reports claim pregnancy carries an 87% risk of hemorrhage whereas a study of 451 patients with known AVMs, the occurrence of hemorrhage was 3.5% during the year after a patient's last menstrual period. However, after the first CAVM hemorrhage, there is a higher risk for a subsequent hemorrhage during pregnancy. Also, bleeding from an AVM during pregnancy has a higher maternal mortality (28%) than in the non-gravid patient. The placement of an arterial line prior to epidural placement or induction of anesthesia is recommended. The use of an epidural is preferred over GA since it provides good peri- and post-op analgesia, the avoidance of cardiovascular stresses, and the ability to monitor the neurological status of an awake patient.

References: