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AORTIC RUNOFF AS A SIGN OF INTRACRANIAL ARTERIOVENOUS MALFORMATION

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Intracranial arteriovenous malformations rarely cause pulmonary hypertension and congestive heart failure in the newborn. Their diagnosis is, however, challenging because cardiomegaly may suggest an intracardiac structural lesion.

Introduction: Congenital arteriovenous malformations (AVM s) are structural abnormalities resulting from arrest in normal morphogenetic processes leading to absence of normal intervening capillary bed between artery and veins so blood rushes from the artery type vessel to vein type vessel without being slowed down by capillaries, which may give rise to hemodynamic symptoms. The hemodynamic signs and symptoms produced by systemic AVMs are determined by their location (brain, liver, thorax, extremities ...) size, and the patient's age (as hydrops fetalis during fetal life, neonatal congestive heart failure, or beyond infancy as a hyperkinetic circulatory state) Small cranial AVMs may cause no symptom until they rupture causing signs of cerebral or subarachnoid hemorrhage. Other manifestations would be neurological problems such as hydrocephalus, seizure, headache, and numbness in one part of body paralysis or loss of speech, memory or vision. Arteriovenous malformation of the vein of Galen in infancy is a rare cause of neonatal heart failure. Symptoms of high-output congestive heart failure due to unique cardiovascular hemodynamics of neonate (patency of ductus arteriosus and foramen ovale, elevated pulmonary vascular resistance, and relative hypertrophy and diminished compliance of right ventricle) may lead to miss diagnosis of intracardiac defect.

Method: In this case report we introduce two neonates which were referred to our hospital (as tertiary cardiac referral center) due to cardiac failure and were though to suffer from congenital heart defect. In fact cardiac symptoms in these cases were secondary to cranial AVM .Echocardiography helped us to reach the correct diagnosis. Our aims in reporting these two cases are contributing to medical knowledge, clinical practice, precise using diagnostic instruments (echocardiography), disease management, and follow-up. Also we believe on its educational value.

Case description: A two-day-old male and eleven-day-old female newborns with intracranial arteriovenous malformations and misdiagnosis of congenital heart disease are presented here.

Discussion: Infants with severe congestive heart failure caused by an intracranial AVM are critically ill, and prompt diagnosis is essential. Two-dimensional ultrasonography of the heart and brain provides a rapid, efficient method for the detection of intracranial AVMs. Alongside the complex adjustments that occur on conversion to extrauterine circulation, the newborn infant with a large intracranial AVM has an additional circulatory burden. Shortly after birth, when the pulmonary vascular resistances are still elevated, the decrease in the total systemic vascular resistance caused by the presence of a large AVM promotes right-to-left ductal shunting. The large venous return to the right atrium from the AVM augments the right-to-left atrial shunting. An increased blood flow to a low-resistance fistula leads to the dilatation of the ascending aorta and carotid arteries, and an increased venous return from the AVM causes the dilatation of the superior vena cava, right atrium, and right ventricle. Doppler examination of the descending aorta shows evidence of a retrograde diastolic flow

Conclusion: Cranial AVMs cause volume overloading and cyanosis due to a persistent fetal circulation. Most of cranial AVM infants are initially deemed to have congenital heart disease. Careful echocardiography, by demonstrating a normal intracardiac anatomy via two-dimensional mode and the "steal" of blood in the cranial region by pulsed and color Doppler modes, helped us reach the correct diagnosis promptly. It should be emphasized that cranial auscultation, albeit an integral component of physical examination in children, more often than not tends to be ignored.

Biography

Maryam Moradian has completed her Residency in pediatrics, Tehran University of Medical Sciences (Children's Medical Center) and Mashhad University of Medical Sciences. She was the Director of pediatrics echocardiography lab, Rajaie Cardiovascular Medical and Research Center from 2011-2013. She has published more than 14 papers in reputed journals.

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