

## A late diagnosis of vein of Galen aneurysm in newborn

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Dear Editor;

A term boy weighing 3900 gr, whose apgar score was 7-9 (in 1 min and 5 min, respectively) was delivered through caesarean section after an unproblematic pregnancy by a 38-year old healthy mother. Hydrocephaly was reported on obstetric ultrasonography follow up. On his physical examination (PE) done in the delivery room, mongoloid facial appearance, intercostal, subcostal retractions, nasal flaring, 4 cm palpable liver, 3/6 pansystolic murmur at all cardiac foci, hyperdynamic heart beat (156 bpm), 66/33 mmHg arterial blood pressure (mean 35 mmHg) and palpable full pulse were detected. Telecardiography and echocardiography were performed as right heart failure findings were detected. Echocardiography revealed a small ventricular septal defect (VSD), a small atrial septal defect (ASD), and a large patent ductus arteriosus (PDA) which is identified through angiography (Figure 1b). Right cardiac structures of the patient were quite large. The patient was re-evaluated as that large right cardiac structures could not be explained with an isolated ASD. On transfrontal ultrasonography (USG) done due to presence of hydrocephaly in prenatal USG, right lateral ventricle diameter was detected as 11 mm, left lateral ventricle diameter was 5 mm and a cystic lesion measuring 36.8x35.2 mm with regular contours was detected neighboring the right ventricle posterior horn (Figure 1a). An arterio-venous malformation was considered due to continuous cranial murmur on physical examination. The patient was diagnosed with

Galen vein aneurysm malformation according to the result of computed tomography angiography obtained for verification of the diagnosis (Figure 1c, 1d). Chromosome analysis was done due to mongoloid facial appearance and it was reported as 47 XY. Two cycles of ibuprofen treatment (10 mg/kg on the first day, continued with 5 mg/kg daily) was administered to achieve PDA closure

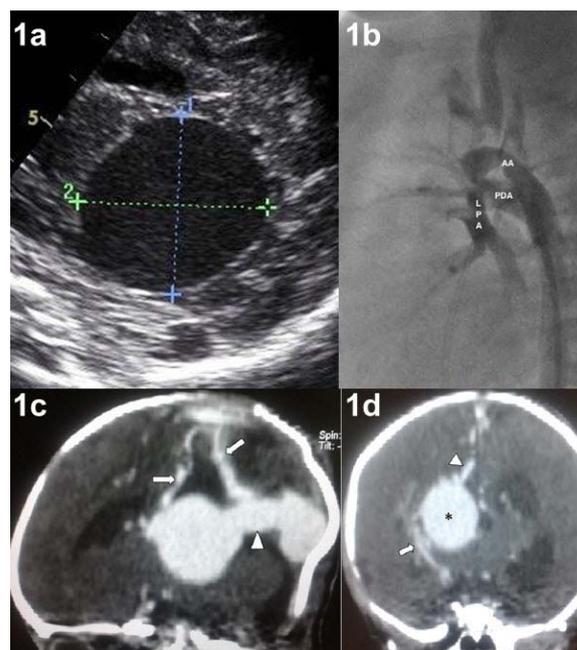


Fig. 1a. Transfrontal ultrasonography demonstrates vein of Galen malformation like a cystic malformation. 1b. Large patent ductus arteriosus is shown through angiography. 1c. Sagittal multiplanar reconstruction (MPR) image of contrast-enhanced CT demonstrates vein of Galen malformation with venous drainage into the straight sinus (arrow head) and superior sagittal sinus (arrows). 1d. Coronal contrast-enhanced CT demonstrates vein of Galen malformation (asterix) with a feeding artery from anterior cerebral artery (arrow head) and middle cerebral artery (arrow). PDA: patent ductus arteriosus, LPA: left pulmonary artery, AA: Arcus aorta.

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which is hemodynamically important for endovascular approach that provides successful results in treatment of galen vein aneurysm malformation (GVAM). Surgical ligation was done as PDA was not closed. The patient whose general condition deteriorated was lost due to multi-organ failure. We wanted to emphasize the importance of transfontanelle auscultation which is simple but diagnostic and a part of physical examination in GVAM which is a rare cause of right heart failure and which may be overlooked.

Galen vein aneurysm malformation was first described by Steinhel in 1895 (1). Galen vein aneurysm is a rare congenital intracranial malformation and although its incidence is 1:25000, it consists 1% of all vascular malformations (2). It commonly presents with high output right heart failure, pulmonary hypertension, intrauterine growth retardation, subarachnoid hemorrhage or intraventricular hemorrhage, hydrocephaly and convulsions during the neonatal period (1). Right heart failure and hydrocephaly were detected in our case. GVAM may develop at 6th-11th weeks of gestation and may be diagnosed with intrauterine USG, however, diagnosis is usually made after birth (1). Although hydrocephaly was detected during intrauterine follow up, inability to detect GVAM may be due to the physician being inattentive or inexperienced. Presence of cardiac abnormalities such as PDA, ASD and VSD accompanying GVAM may lead to early development of heart failure and delay the diagnosis of GVAM. The reason for the delay in the diagnosis until postnatal third day was the

presence of hemodynamically significant PDA accompanied by ASD and VSD. Therefore GVAM should be kept in mind in unexplained right heart failure (3, 4). While mortality rate was 100% before, survival rate relatively increased with endovascular treatment approaches (5). However GVAM is still an arteriovenous malformation with high mortality. GVAM should be kept in mind in unexplained right heart failure in the newborn.

Head auscultation which is usually a neglected part of physical examination may be diagnostic in these cases. Guidelines formed as a result of studies done with large series are required in order to manage GVAM cases and reduce mortality rates.

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