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CASE REPORT

Galen vein aneurysm: case report

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Abstract

An aneurysm malformation of the Galen vein is a rare anomaly, accounting for less than 1% of all congenital cerebral vascular malformations, but representing 30% of these in the pediatric age. It is a cerebral arteriovenous malformation, prenatal development, which begins before ten weeks of gestation, and results from the formation of arteriovenous fistulas between the choroidal circulation and the median prosencephalic vein, an embryonic vessel precursor of the Galen vein, which becomes dilated. We present a rare case of this malformation and its evolution after treatment with endovascular embolization.

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INTRODUCTION

Vein of Galen aneurysmal malformations (VGAM) account for less than one percent of all congenital cerebral vascular malformations. Nevertheless, 30% of the cases of this rare anomaly affect pediatric populations.[1,2] It is a cerebral arteriovenous malformation that develops prenatally before the tenth week of gestation and results in the formation of arteriovenous shunts between choroidal circulation and the median prosencephalic vein of Markowski, a precursor to the vein of Galen that becomes dilated in cases of VGAM.[1,3,5]

The severity of VGAM varies and clinical manifestations may differ depending on patient age at the time of disease presentation.[2,5] Signs of disease in prenatal life include heart failure and hydrocephalus caused by venous congestion or oligohydramnios, a consequence of poor renal blood flow secondary to heart failure.[1,10] In neonatal life, the most frequent clinical manifestation is high-output heart failure. Other rare presentations include necrotizing enterocolitis secondary to bowel hypoperfusion.[2,7]

CASE REPORT

A newborn previously diagnosed with fetal malformations based on ultrasound examination had VGAM with dilated venous system and cardiomegaly suggestive of heart failure. The ductus had an increased pulsatility index (PI).

A male infant with a gestational age of 30 weeks and a day in cephalic presentation weighing 3000 grams was born after a cesarean section. The amniotic fluid was clear and the membranes ruptured during the cesarean section performed at the Hospital Materno Infantil de Brasília, Brazil. The newborn did not have a nuchal cord. He was placed on sterile drapes and cried loudly at birth. His cord was clamped with some delay. The newborn was taken to a preheated cot, dried, had his mouth and upper airways suctioned and his heart rate (HR) measured (<100 bpm). He was breathing irregularly and responded well to positive-pressure ventilation (PPV) with a mask (FiO2: 30%). The newborn developed moderate respiratory distress. His pulse oximeter read oxygen saturations ranging from 75% to 82%. The fraction of inspired oxygen (FiO2) was increased to 60% and then gradually decreased to 25%. Physical examination was performed and anthropometric measures were taken. His Apgar scores at 1 and 5 minutes were 6 and 8, respectively. Stool and urine were present. HIV and VDRL tests performed on admission were non-reactive.

The newborn was in respiratory distress and was referred to the Neonatal Intensive Care Unit (NICU). He was placed on nasal CPAP. Chest X-rays showed an enlarged heart and normal lungs. He was removed from nasal CPAP 20 hours after birth and equipped with an infant hood for CPAP therapy. Transfontanellar ultrasound performed on his second day of life found a cystic structure in the area of the vein of Galen. The Doppler mode showed a vascular lesion with turbulent flow (Figure 1).

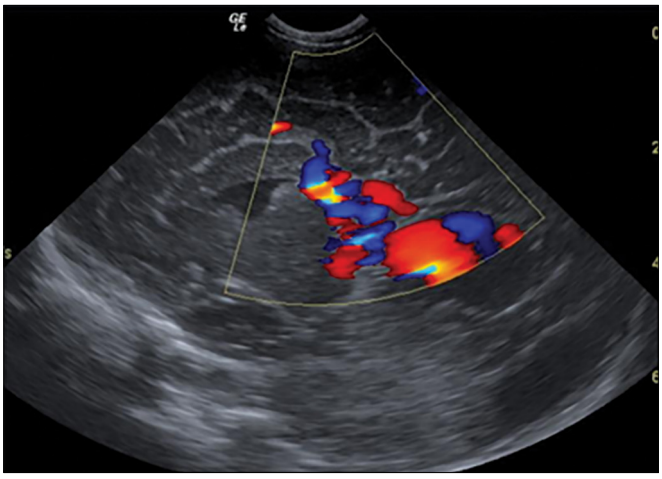


Figure 1. The Doppler mode showed turbulent flow, indicating the vascular nature of the lesion.

An echocardiogram performed on the third day of life of the infant showed signs of severe pulmonary hypertension. He was started on furosemide. His respiratory distress worsened, peripheral perfusion deteriorated, and he had a hyperkinetic apex. He was placed on a mechanical ventilator (MV), the dose of furosemide was increased and combined with noradrenaline (replaced with dobutamine after day 6). An echocardiogram performed on day 7 showed signs of pulmonary hypertension and good ventricular function. He was kept on the MV [FiO2: 25%; delta-p: 14; inspiratory time: 0.38; PEEP 6.0; respiratory rate (RR): 45]. The patient was on dobutamine, furosemide, spironolactone, and sedoanalgesia (fentanyl and Midazolam). On day 8, the newborn was transferred to Instituto Hospital de Base – DF (IHB-DF) to undergo an embolization of the vein of Galen aneurysm at the hospital's catheterization lab. The newborn had a cardiac arrest probably due to hypoventilation. The Pediatric Intensive Care Unit (PICU) team was called about ten minutes later for support. Chest compressions were performed and three minutes later his heart showed signs of electrical activity, with a palpable central and a thin peripheral pulse. The infant's skin was mottled and he was hypothermic and bradycardic, with mydriatic pupils unresponsive to light. He was on arrest for 13 minutes. He was given seven bolus injections of adrenalin and two bolus injections of atropine. Blood gas test after the return of electrical activity: pH: 6.71; PaO2: 28.4; PaCO2: 157.8; Bicarbonate: 19.6; base excess (BE): -16.9; oxygen saturation (O_2 sat): 31.5%; Lactate: 6; Na^+: 134; K^+ 3,6; Hematocrit (HTC): 29.2%. The anesthesia cart did not generate effective lung volume and was changed for bag-valve mask (BVM) ventilation. The HR improved to about 125 bpm and perfusion. Blood gas after 15 minutes on BVM read as follows: pH: 6.99; PaO2: 170.8; PaCO2: 52.2; Bicarbonate: 12.4; BE: -18.3; O_2 sat: 98.7% Lactate: 8.2; Glucose: 149; Na^+: 141; K^+: 3.31; HTC: 29.1%.

On the first day after surgery the patient was hemodynamically stable on adrenalin 0.6 mcg/kg/min and dobutamine

8 mcg/kg/min, with proper blood pressure (BP) levels and tachycardia. He had two bouts of fever (38ºC) and tachycardia, which prompted a decrease in the temperature of the incubator and the administration of dipyrone. The infant seized and was given a loading dose of phenobarbital; he was kept on a maintenance dose and did not seize again. His urine output was still increased. He was on furosemide, with jaundice but not at a level in which he would require phototherapy. The infant did not move his left leg.

On the second day after partial embolization (30%) of the aneurysm the patient's condition was still severe, on MV with moderate settings: inspiratory pressure (IP): 16; PEEP: 6; FiO2: 40%; the patient was hemodynamically stable and on adrenalin (0.5mcg/kg/min) and dobutamine (5 mcg/kg/min). The settings were decreased in the early afternoon and the patients tolerated the changes well. He did not seize again. Fever bouts (37,8ºC) were observed in the last checks. The patient had been on cefepime and vancomycin since the first day after endovascular surgery. The patient tolerated the start of diet well, without signs of bloating or vomiting. No complications were reported. A chest X-ray found the following: significant cardiomegaly with mild bilateral perihilar infiltrate and without areas of condensation or atelectasis.

The infant's hemodynamic parameters deteriorated, requiring an increase in the dose of epinephrine to 0.7 mcg/kg/min; he also had episodes of desaturation. Fentanyl was increased to 3 mcg/kg/min, but the infant still had bradycardia (77bpm) and desaturation for approximately 5min. Chest compressions were performed and the infant was given adrenalin and atropine, and the dose of adrenalin was increased to 1mcg/kg/min; his respiratory parameters were as follows: IP: 24; PEEP: 1cmH_2O; RR: 45; FiO2: 100%. Inspiratory time: 0.4.

The infant went into cardiac arrest, was resuscitated (chest compressions, 8.4% sodium bicarbonate, adrenalin, atropine), and had the adrenalin dose increased to 1mcg/kg/min. Mechanical ventilation parameters were increased and the FiO2 was set at 100%. Resuscitation measures were unsuccessful and the patient died at the age of one month and five days.

DISCUSSION

Cases of the disease are diagnosed mostly after birth[5], although changes in obstetric ultrasound examination can be observed at 14 weeks pregnant. Cystic cerebral dilation may be seen in association with ventriculomegaly and oligohydramnios[1] (Figure 2). Newborn transfontanellar Doppler ultrasound is a valuable, widely available, non-invasive diagnostic tool that does not require sedation.[5,9]

Most cases (94%) are diagnosed after birth possibly from signs of heart failure observed in affected neonates,[11] as in our case. Managing heart failure in patients with the condition is challenging. However, diuretics and volume restriction to decrease preload are the pillars of therapy.[12] The patient described in this report was on diuretics and vasoactive drugs to compensate for heart failure.

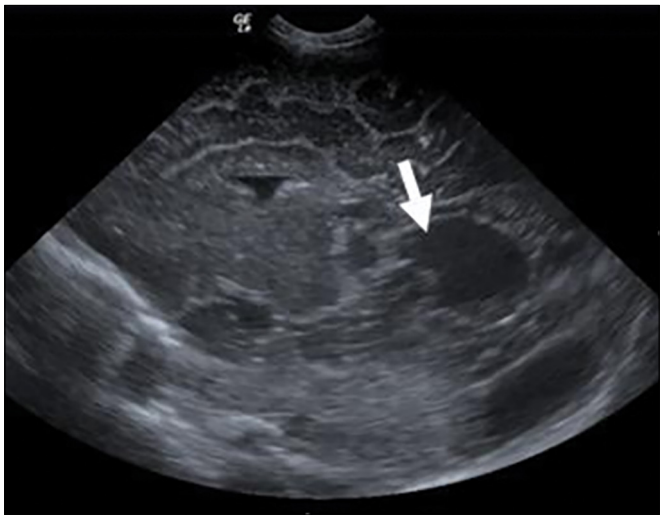


Figure 2. Cerebral cystic dilation seen in transfontanellar ultrasound examination.

Lasjaunias conducted the largest transarterial embolization study on record (216 patients with VGAM) and reported that 10.6% of the patients died despite embolization or as an outcome of the procedure. Almost three quarters (74%) of the survivors had normal global development, 15.6% had moderate cognitive impairment, and 10.4% had severe cognitive impairment. Age seems to be an important prognostic factor. The death rate in the group of neonates was 52% versus 10.6% of the cohort as a whole.[12] The patient described in our report died, thus corroborating the study cited above in terms of the high death rates seen in newborns.

The newborn described in our case underwent transarterial embolization of anterior circulation. This has been considered the best option to treat the malformation he presented, leaving open surgery for cases with poorer prognosis or, in rare occasions, to supplement the endovascular procedure.[13] Total embolization of the malformation is not required, since partial obliteration - a third to half of the malformation as performed in our patient (30% embolization) - should be enough to achieve clinical improvement.[14] Embolization coils and microballoons may be used to decrease flow in the fistula, thus allowing a safer use of Onyx® or cyanoacrylates.

Our patient went into respiratory arrest during the endovascular procedure and was resuscitated by the PICU team, with electrical activity returning after 13 minutes of arrest. His condition had become even more severe. The patient died at the age of one month and five days.

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