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Vein of Galen Aneurysm as a Possible Cause of Congestive Heart Failure in the Neonatal Period: About 2 New Cases

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Abstract

Aneurysm of the vein of Galen is a pseudo-aneurysmal dilation of the ampulla of Galen with one or more arteriovenous fistulas, discovered ante or postnatally. We report two cases for which the diagnoses of aneurysm of the vein of Galen were made in the neonatal period at 5 and 17 days of life in front of clinical signs of heart failure and an intracranial murmur. Cardiac ultrasound remains a tool that can guide the diagnosis objectifying the ascending diastolic flow at the level of the arch of the aorta. The transfontanellar ultrasound remained the examination of choice in our two patients for the confirmation of the diagnosis. In our two patients, no specific treatment could be carried out and their evolution was unfavorable with death of the two patients. The aneurysm of the vein of Galen is a rare disease whose prognosis is often unfavorable in our context of exercise. Auscultation of the skull must be systematic in the face of any severe heart failure that cannot be explained by the data of the cardiovascular examination.

Subject Areas

Oncology

Keywords

Galen Vein Aneurysm, Neonatal, Senegal

1. Introduction

A vein of Galen aneurysm is a rare, complex intracranial arteriovenous malfor-

mation associating pseudo-aneurysmal dilation of the ampulla of Galen with one or more arteriovenous fistulas [1]. In the neonatal period, it usually manifests in the first week of life and most often, with congestive heart failure [2]. In the absence of aggravating factors, the disease can manifest later in childhood or even in adulthood [3]. We report two severe cases with neonatal revelation.

However, neonatal Galen vein aneurysm is very rare, and diagnosis is not to be forgotten.

2. Observation 1

It was a newborn male, the third of three siblings, the first two of whom were alive and well. There was no notion of consanguinity between the parents. The pregnancy was poorly monitored; no blood tests or ultrasound examinations had been carried out. The delivery took place at term vaginally on a vertex presentation without any incident. The newborn was eutrophic and extra-uterine adaptation was satisfactory with an Apgar score of 8/10 at the 1st minute then 10/10 at the 5th minute. He was admitted to our department at 6 days of life for a refusal to suckle, incessant crying and frank mucocutaneous jaundice without discoloration of the stools and a fever. The entrance examination showed a temperature of 38.8˚ Celsius, a heart rate of 156 beats/minute and a respiratory rate of 78 cycles/minute. Weight was 2500 g, height 50 cm and head circumference 35 cm. Physical examination revealed mucocutaneous pallor, superficial polypnea without signs of struggle, regular tachycardia with a 3/6 mesocardiac murmur. The femoral pulses were well perceived and the pulmonary fields were free. Biological examinations showed predominantly neutrophilic leukocytosis, anemia with a hemoglobin level of 11.7 g/lnormochrome, normocytic, a CRP of 96 mg/l and mixed hyperbilirubinemia (Total bilirubinemia = 137 µmol/direct bilirubinemia = 73.5 µmol/I). The frontal chest X-ray showed global cardiomegaly with a cardio-thoracic index of 0.7. Echocardiography revealed a stenosis on the left pulmonary artery with moderate pulmonary arterial hypertension. In front of this table we evoked an early neonatal infection associated with congenital heart disease.

Antibiotic therapy combining cefotaxime for 10 days and gentamicin for 48 hours was initiated. The evolution was marked by stable apyrexia, normalization of leukocyte and C-reactive protein levels. A blood transfusion also normalized the hemoglobin level by reducing the level to 14.8 g/l. However, there was a persistence of mucocutaneous jaundice. At 17 days of postnatal life and 11 days of hospitalization, we observed the appearance of cardiac decompensation with the onset of respiratory distress with tachypnea at 74 cycles/minute, crackling rales in the fields lungs, regular tachycardia at 164 beats/minute, hepatomegaly and turgidity of the jugular veins. The electrocardiogram showed sinus tachycardia 154 beats/minute, signs of atrial and right ventricular hypertrophy and negative waves in the anterior and inferior part of subepicardial ischemia. pulmonary arterial hypertension and ascending diastolic flow at the level of the arch of the

aorta with dilatation of the brachiocephalic trunk. Faced with this major heart failure poorly correlated with the importance of the underlying heart defect, the hypothesis of an extracardiac cause is raised.

Physical examination revealed, on auscultation of the skull, a continuous murmur audible at the level of the anterior and posterior fontanelles pointing towards an intracranial vascular malformation. Auscultation of the hepatic area revealed no abnormalities. The transfontanellar ultrasound then revealed a liquid formation taking the color on the Doppler, 36.7 × 19 × 22.9 mm behind the third ventricle which it pushes forward in favor of an aneurysm of the associated Galen's ampulla to early hydrocephalus (Figure 1).

Cranial Doppler ultrasound showed fluid formation taking color on Doppler behind the third ventricle, which it pushes forward in favor of an aneurysm of the ampulla of Galen (Echo-Doppler figure).

Ultrasound examination of the liver revealed echo hepatomegaly with normal structure and slight dilation of the suprahepatic veins.

Biologically, there was a slight decrease in bilirubinemia with total bilirubin at 87 µmol/l and direct bilirubin at 47.8 µmol/l, hepatic cytolysis with ALT level at 324 UI/L and AST level at 196 UI/l, a drop in the prothrombin level with a TP at 42% and an INR at 2.34. Renal function was normal. The evolution was unfavorable with death of the patient at 20 days of life in an array of cardiac decompensation refractory to medical treatment.

3. Observation 2

This was a newborn female, the fourth of four siblings, the first three of whom were alive and well. There was no notion of parental consanguinity. She was born to a 29-year-old mother, the pregnancy was followed with four prenatal consultations, the blood tests carried out were unremarkable. A late ultrasound was performed showing an evolving intrauterine monofetal pregnancy. The delivery took place at 36 weeks vaginally in a cephalic presentation. There were no risk factors for maternal-fetal infection, the amniotic fluid was clear. The newborn did not cry at birth. He was resuscitated for 10 minutes, with suction and balloon ventilation. Her Apgar score was 5/10 at the tenth minute, her birth

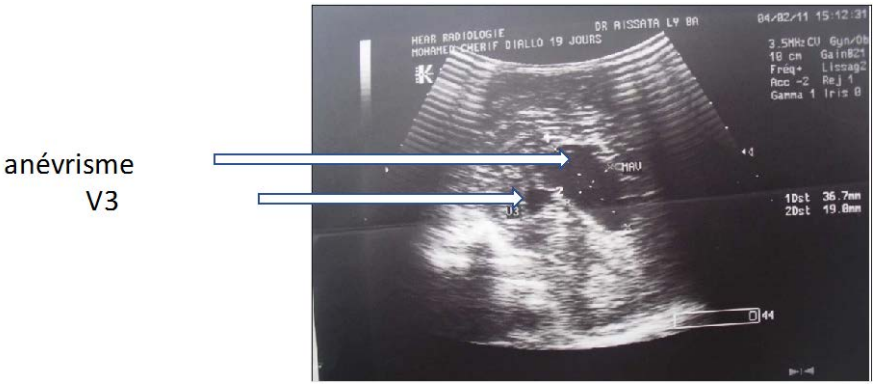

Figure 1. Ultrasonic image.

weight was 3000 g. He was admitted to the pediatric department of the Pikine hospital and his examination noted as constants: temperature at 35.3˚ Celsius, respiratory rate at 36 cycles per minute, heart rate at 136 beats per minute, preductal saturation at 54% and postductal at 47%, a dextro at 1.02 g/l. He had presented respiratory distress with a silverman score of 3/10, hypoxemia with peripheral cyanosis, neurological distress with dull archaic reflexes. Cardiac auscultation was normal, femoral pulses were well perceived and pulmonary fields were free. The biological examinations showed normal leukocytes at 7140/Ul, a normal hemoglobin level at 14.7 g/lnormochrome, normocytic, and platelets at 192,000/Ul A CRP less than 6 mg/l and The frontal chest X-ray showed a slight global cardiomegaly with a cardio-thoracic index of 0.68.

Faced with this picture, we mentioned neonatal asphyxia complicated by SARNAT 2 anoxic-ischemic encephalopathy. He had benefited from nasal CPAP oxygenation improving his saturation. On the 5th day of hospitalization, a generalized edematous syndrome set in; it is referred to the CHNEAR. His examination on arrival revealed a heart failure syndrome, regular tachycardia with a mesocardiac murmur of 4/6. On auscultation of the skull, a continuous murmur audible at the level of the anterior and posterior fontanelles.

Cardiac Doppler ultrasound had objectified an ascending diastolic flow at the level of the arch of the aorta with dilation of the brachiocephalic trunk, significant dilation of the heart chambers with global hypercontractility and severe impairment of cardiac function. No structural cardiac malformation found.

Transfontanellar ultrasound revealed a well-defined, anechoic oval formation measuring 41.2 × 18 × 25.9 mm, located on the midline behind the 3rd ventricle, above the cerebellum, within which the Doppler highlighted shows a flow. The lateral ventricles and the 3rd ventricle were slightly dilated (Figure 2).

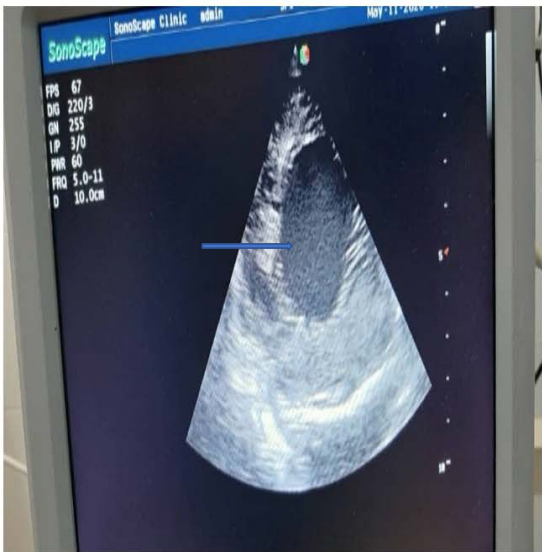

Figure 2. A well-defined, anechoic oval formation of 41.2 × 18 × 25.9 mm, located on the midline behind the 3rd ventricle, above the cerebellum, within which the Doppler highlighted a flow. The lateral ventricles and the 3rd ventricle were slightly dilated.

On the biological level we noted an alteration of the renal function with uricemia at 1.7 g/l, transaminases at more than 8 times the normal.

Therapeutically, he was intubated and sedated under the fentalin-midazolam protocol and put on dobutamine at 15 gamma/kg/minute, furosemide 2 mg/kg. The evolution was unfavorable with death of the patient on the 7th day of life in a picture of cardiac decompensation with refractory cardiogenic shock to medical treatment.

4. Discussion

Galen's vein aneurysm is a rare cerebral vascular malformation. Its prevalence is estimated at 2.5 cases per 100,000 live births [4]. It accounts for 1% of intracranial vascular malformations and 30% of cases diagnosed in pediatrics [5]. It is characterized by the abnormal persistence of the median porencephalic vein of Markowski which, supplied by the embryonic primitive meningeal arteries, becomes the blood collector of this malformation. The aneurysm results from the absence of differentiation in the mature arteriovenous system, between the 7th and 12th week of gestation, of a primitive, median embryonic vein which drains the primitive choroid plexuses [6] [7].

The development of obstetric ultrasound coupled with Doppler has made it possible to considerably improve antenatal diagnosis. The malformation is most often evoked during the third trimester of pregnancy in front of an intracerebral image of the vascular type associated or not with a dilation of the cardiac chambers [8]. The antenatal diagnosis which would have allowed us to anticipate the management from birth before the onset of complications could not be made in our two patients, despite the third trimester ultrasound performed in the second observation.

Galen's aneurysm can manifest in the neonatal period or later in childhood [3]. In the neonatal period, it most often manifests in the first week of life as congestive heart failure, noted in our second patient. On the other hand, in our first patient, the clinical manifestations of cardiac dysfunction were late, during the third week of life. Cherif *et al.* [9] reported a similar neonatal case with late revelation in the third week of life. This evolutionary mode would be favored either by an aggravating factor, or by a variability of expression of the malformation. The aggravating factor may be an intercurrent infection or a malformation of cardiac or cerebral origin [9]. We found the infection and the heart defect in our first patient. Neonatal asphyxia would be the aggravating factor in the second patient. Magnetic resonance imaging, when it is possible, often makes it possible to specify the vascular network involved in the arteriovenous anastomosis, and also to determine its cerebral impact [10] [11]. The seriousness of the clinical picture of our patients did not allow us to transfer them to an imaging department to perform this examination.

Well-tolerated forms often have a better prognosis with the possibility of treatment based on transarterial embolization which can be performed at the age

of 5 months [2] whereas the presence of aggravating factors, such as case in our observations, can make the prognosis more reserved. The analysis according to the Bicêtre score of the impact of the aneurysmal malformation and of the aggravating factors shows that the multiorgan failure was such in our patients that it was beyond any therapeutic perspective [2].

5. Conclusion

The aneurysm of the vein of Galen is a rare disease whose prognosis is often unfavorable in our context of exercise. It must be evoked in the presence of severe heart failure which cannot be explained by the data of the cardiovascular exploration. Its perinatal management can be improved by antenatal screening by an experienced and well-trained sonographer.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

Conflicts of Interest

The authors declare no conflicts of interest.

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