Ultrasound Prenatal Diagnosis and Emergency Interventional Radiologic Therapy of Galen Aneurysmal Malformation in a Newborn

Roxana Elena BOHILTEA\textsuperscript{a,b}, Natalia TURCAN\textsuperscript{a,b}, Cristian MIHALEA\textsuperscript{c,d}, Bogdan DOROBAT\textsuperscript{b}, Eliza Elena CINTEZA\textsuperscript{a,e}, Adriana DAN\textsuperscript{b}, Magdalena MIHA\textsuperscript{b}, Adela DIMITRIADE\textsuperscript{b}, Cristian BOROS\textsuperscript{b}, Mircea DUMITRU\textsuperscript{b}, Ionut GOBEJ\textsuperscript{f}, Octavian MUNTEANU\textsuperscript{a,b}, Monica Mihaela CIRSTOIU\textsuperscript{a,b}

\textsuperscript{a} “Carol Davila” University of Medicine and Pharmacy, Bucharest, Romania
\textsuperscript{b} University Emergency Hospital, Bucharest, Romania
\textsuperscript{c} Department of Interventional Neuroradiology, NEURI, Hopital Bicetre, APHP, Paris Sud Universite´ - Paris, France
\textsuperscript{d} Department of Neurosurgery, University of Medicine and Pharmacy “Victor Babes,“ Timisoara, Romania
\textsuperscript{e} “Maria Sklodowska Curie” Emergency Children’s Hospital, Bucharest, Romania
\textsuperscript{f} Colentina Clinic Hospital, Bucharest, Romania

ABSTRACT

Located under the cerebral hemispheres and draining the anterior and central regions of the brain into the sinus of the posterior cerebral fossa, the vein of Galen aneurysmal malformation is considered to be a rare cause of hydrocephaly. The presence of this condition in the neonatal period typically includes intractable heart failure and a poor prognosis. We report a case of aneurysm of the vein of Galen diagnosed prenatally at 28 weeks of gestation, with the delivery at term by caesarean section of a female infant. Sonographically, the vein of Galen appeared in the mid-sagittal plane, large, supratentorial, non-pulsatile; on color Doppler, the structure filled with bright color, reflecting a turbulent venous flow. A low grade of ventriculomegaly was present during the evolution of pregnancy; regarding the cardiovascular function, an intrauterine right cardiac insufficiency overlapped a tricuspid regurgitation and right atrial dilatation. A multidisciplinary committee decided a neonatal embolization of the aneurysm as an emergency requirement due to increased pulmonary hypertension developed in the next 24 hours after birth.

After the embolization of the two main drainage vessels, the cardiac dysfunction persists. Two days later the evolution became unfavorable, leading to the necessity of the second embolization, which resulted in a 48 hours’ coma and death, due to a cerebral hemorrhage secondary to thrombosis and fissure of the embolized aneurysm. The prognosis for the neonate with malformation of the Galen vein depends upon the severity of the cardiovascular status. Embolization represents actually the treatment of choice with the best results of these cases, but the mortality remains as high as 50 percent even in the most specialized centers of the world. As far as we know this is the only case of Galen aneurysmal malformation in Romania which benefitted of embolization by interventional treatment in neonatal period.

Keywords: Galen aneurysm, interventional neuroradiology, embolization

Address for correspondence:
Eliza Cinteza
Postal address: “Maria Sklodowska Curie” Emergency Children’s Hospital, Constantin Brancoveanu Bte, nr 20, sect 4, Bucharest, Romania
E-mail: elizacinteza@yahoo.com, Bucharest, Romania

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INTRODUCTION

The aneurysm of the vein of Galen lines among intracranial arteriovenous malformations, with an incidence of 1% of all fetal arteriovenous vascular abnormalities. The association with the persistence of the embryonic median prosencephalic vein of Murkowski is not uncommon (1). This is a rare choroidal arteriovenous malformation that begins to develop in the early embryonic stages and has a declared mortality rate of about 50% (2). The symptomatology is rarely present before third trimester of pregnancy, therefore the diagnosis before 24 weeks of gestation is unusual. Very rare cases early diagnosed are mentioned in literature: one case reported by Lee et al. (3) of vein of Galen aneurysm was diagnosed at 23 weeks of gestation and another case of detection of an aneurysm of the vein of Galen following signs of cardiac overload was diagnosed in a 22-week old fetus (4). Increased morbidity and mortality related to this condition is importantly correlated to the associated fetal or neonatal impaired heart function. Severe heart failure is developed early in the fetal life and is produced by a large systemic shunt. This causes cardiomegaly, heart failure and pulmonary hypertension (5). Clinical manifestations of Galen aneurysmal malformation depend on the volume of arteriovenous shunting. Two forms are distinguished: the severe form with congestive heart failure with a rapid in utero progression and a high mortality, and a mild childhood form (6). According to Litvak et al (7), Galen aneurysmal malformation can be subdivided into a true form (if arteriovenous shunting is occurring directly into the vein of Galen) and secondary form (arteriovenous shunt is located near or far from the vein of Galen). The signs of cardiac overload or fetal hydrops may be first detected on a routine ultrasound detailed fetal examination and implicitly lead to detection of various vascular malformations. On ultrasound, an intracerebral cystic structure or a hypoechoic intracranial structure, usually, represents aneurysmal malformation. Doppler technique elucidates the diagnosis by identifying the structure as a vascular malformation. In 1983, Hirsch et al, declared that the vein of Galen aneurysm is the first malformation where the application of Doppler ultrasound has confirmed the prenatal diagnosis (8). The poor prognosis of vein of Galen aneurysmal malformations is due to the quick development of multisystem organ failure, the hypoperfusion of intact brain parenchyma, the “steal” phenomenon, especially when the brain injury is already present. There are two main methods of treatment suggested: direct surgical removal and endovascular embolization. Since the introduction of the endovascular method the mortality decreased importantly and varies from 4% to 40% (9). By endovascular embolization the arteriovenous shunt is occluded and the pathological arteries and aneurysmal veins are disconnected from the circulation.

CASE REPORT

We present the case of a female newborn, extracted through caesarian section at 38 weeks of gestation, for intrauterine signs of cardiac failure due to Galen aneurysmal malformation.

A 21-year-old woman, gravida 1, para1, was admitted in our department at 28 weeks of gestation for a routine pregnancy control and a third trimester ultrasound. The fetus had been diagnosed on ultrasound (US) with a cerebral arteriovenous malformation, confirmed by Doppler 3D ultrasound technique (Figure 1a-c, 2) and magnetic resonance imaging (MRI) (Figure 3-4). Sonographically, the vein of Galen appeared in the mid-sagittal plane, anechoic, large, ovalar shaped, supratentorial, non-pulsatile, continued to the posterior by the dilated straight sinus; on

FIGURE 1 a-c. Ultrasound 3D HD live. Flow image of aneurysm of Galen
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Color Doppler, the structure filled with bright color, reflecting a turbulent venous flow. A particular “8 shape” aspect of Willis polygon has been noticed (Figure 2). A mild ventriculomegaly was present during the evolution of pregnancy; regarding the cardiovascular function, an intrauterine right cardiac insufficiency overlapped a tricuspid regurgitation and right atrial dilatation. The pregnancy had an uneventful course until 38 weeks of gestation when in context of a labor debut the newborn was extracted through caesarean section. The mother had a quick recovery without any post operatory complications. At birth baby had: weight 3050 g, length 50 cm, head circumference 32.5 cm, chest circumference 31 cm, Apgar score of 7 at 1 minute and 8 at 5 minutes. Clinical examination after birth revealed difficult postnatal adaptation, with respiratory distress syndrome; tachypnea, expiratory grunting, oxygen-dependency, signs of cardiac failure (edema, tachycardia, loud sound II, cardiac murmur, positive Harzer sign, cardiomegaly, hepatomegaly), and murmur at anterior fontanel, mild hypotonic.

Chest X-ray revealed an increased cardiothoracic index and globally decreased pulmonary transparency. Head US showed, in coronal plane, a large anechoic circular image in the midline, interventricular, with a loud Doppler signal, turbulent flow, suggesting a vascular anomaly. Echocardiography evaluation confirmed the enlargement of heart chambers, especially on the right side, important tricuspid regurgitation, severe pulmonary hypertension (systolic pressure in main pulmonary artery up to 108 mmHg), and large patent ductus arteriosus, with bidirectional shunting.

Immediately after birth, the newborn was admitted in the neonatal intensive care unit (NICU), placed in an incubator, received hood oxygen therapy (FiO₂ 40%) for 24h, cardiac inotropic support with Milrinone, diuretic therapy with Furosemide, prophylactic antibiotic therapy (Ampicillin and Gentamycin), endovenous infusion of electrolytes, parenteral nutrition. Due to unfavorable course of the respiratory status, with increased oxygen need, the baby was intubated at 24 hours of age and put on mechanical ventilation. Because of the persistent increased pulmonary pressure, above the systemic mean arterial pressure, the shunting through the patent ductus

![FIGURE 2. Ultrasound Color Doppler atypical image of Willis polygon](image1)

![FIGURE 3. MRI image in sagital plane of aneurysmal dilatation and straight sinus dilatation color Doppler, the structure filled with bright color, reflecting a turbulent venous flow. A particular “8 shape” aspect of Willis polygon has been noticed (Figure 2). A mild ventriculomegaly was present during the evolution of pregnancy; regarding the cardiovascular function, an intrauterine right cardiac insufficiency overlapped a tricuspid regurgitation and right atrial dilatation. The pregnancy had an uneventful course until 38 weeks of gestation when in context of a labor debut the newborn was extracted through caesarean section. The mother had a quick recovery without any post operatory complications. At birth baby had: weight 3050 g, length 50 cm, head circumference 32.5 cm, chest circumference 31 cm, Apgar score of 7 at 1 minute and 8 at 5 minutes. Clinical examination after birth revealed difficult postnatal adaptation, with respiratory distress syndrome; tachypnea, expiratory grunting, oxygen-dependency, signs of cardiac failure (edema, tachycardia, loud sound II, cardiac murmur, positive Harzer sign, cardiomegaly, hepatomegaly), and murmur at anterior fontanel, mild hypotonic.

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![FIGURE 4. MRI image in transverse plane of aneurysmal dilatation and straight sinus dilatation](image2)
arteriosus became unidirectional, right to left, which was considered beneficial for decreasing the pulmonary pressure, therefore treatment with Prostin (prostaglandine E1) for maintaining the ductus was initiated.

Because in this case, as in most of the arterial-venous malformations, the cause of the cardiac failure is the increased flow to the heart (increased preload), the only efficient treatment should be aimed to close by embolization the anomalous vessels. Although generally recommended age for intervention is 3 to 6 months, in this case the intervention was decided to be done in emergency, due to the gravity of the cardiac disease. The embolization procedure was made in two steps, first at 3 days of life, second
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at 7 days of life, by catheterization of femoral artery, under general anesthesia, in the angiography laboratory of the Emergency University Hospital Bucharest. The purpose of the endovascular treatment was to close at least 60-70% of the direct arterial shunts involved mainly from the posterior circulation (left and right branches of posterior cerebral arteries and left and right superior cerebellar arteries were involved) of this mural type VGAM (Figure 5). During the first procedure, our neuro-interventional team succeed to close 45-50% from the total flow (Figure 6) by obstructing two direct shunts, one from right superior cerebellar artery using 2 detachable coils and a mixture of 80% Glubran and 20% Lipiodol, and the second one from left posterior cerebral artery with a mixture of 60% Glubran and 40% Lipiodol. The most challenging technical aspect was to close the first shunt due to the huge aspiration effect and to avoid the pulmonary artery direct embolization; we were obliged to use coils and also a higher Glubran concentration. The second endovascular treatment was imposed by clinical presentation and had taken place just 7 days after the first one, even the best option, if it should be possible from cardiac point of view, is to have at least 3 months interval (Figure 7). The micro-catheterisation was this time more difficult due to important tortuosity of the selected two branches (Figure 8) one from right internal carotid artery and one from posterior circulation. In the end of the procedure almost 80% of the flow was reduced (Figure 9) and no contrast extravasation, respective active bleeding was observed during this procedure. At the trans fontanel echography done 2 hours post procedure a high suspicion of intra-ventricular haemorrhage was evocate and finally confirmed by CT scan.

Our opinion is that this delayed rupture was initiated due to acute thrombosis of the vein of Galen aneurysm itself, an important quantity of proteolytic enzymes being realised and leading to acute wall rupture. This mechanism is well knotted and described in the delayed giant intracerebral aneurysms rupture post endovascular treatment and represents a terrifying complication in our field.

In the next 3 hours an external ventricular drainage was put in emergency by neurosurgery team in the hope that this intra-ventricular haemorrhage could be tolerated.

The second intervention was considered necessary because of the unsatisfactory response of the cardiac function after the first embolization (systolic pulmonary pressure measured echocardiographically had become 60mmHg, but with signs of decreased biventricular systolic function). Indeed, after the second procedure a significant improvement in cardiorespiratory state of the baby was noticed: the murmur at the anterior fontanel disappeared, the heart rate decreased, Harzer sign disappeared, the heart size decreased on chest X ray and echocardiography, systolic pressure in main pulmonary artery decreased to 39 mmHg with normalized biventricular systolic function.

Despite improvement in hemodynamics, the clinical course of the baby was unfavorable from neurological point of view: decreased tone, reactivity, reflexes, evolving to coma, due to cerebral hemorrhage (intraventricular and periventricular) with secondary hydrocephaly. There were debates regarding the opportunity of anticoagulant therapy (thrombosis of the cerebral sinuses is often a complication of embolization), but since the newborn had prolonged coagulation time, the treatment was not initiated. External ventricular drainage was instituted which evacuated approximately 50 ml of cerebrospinal fluid, with hematic content, during 10 days. Despite complex therapeutic measures the baby died at 17 days of life. □
DISCUSSION AND CONCLUSION

Along with the continuous medical development, the pathologies related to arteriovenous malformation, including vein of Galen aneurysm was repetitively described and upgraded. Formed by the confluence of the two internal cerebral veins among to the basal vein of Rosenthal, vein of Galen aneurysm in 1989 was classified into three different types: choroidal, mural and dilatation of vein of Galen secondary to increased blood inflow as a consequence of the arteriovenous malformation (9). Only the last type is considered to be actually the vein of Galen aneurysm. The first two types are characterized by the dilatation of the embryonal median vein of prosencephalon. Our case can be included in the third type; the aneurysm in this case had an important size due to the blood drainage supplied by the arteriovenous malformation.

Regarding the manifestations of this condition, they depend on the age of patient and on the proportion of the indirect signs (cardiomegaly, pulmonary hypertension, fetal hydrops) when diagnosed antenatally. The proportion of the antenatal diagnosis is declared to be about 40% (10). Imaging methods as angiography, ultrasonography, computed tomography (CT) and magnetic resonance imaging are essential for a clear and certificated diagnosis. In the neonatal period, the aneurysm of the vein of Galen is diagnosed with the help of pulsed Doppler ultrasonography and color flow mapping. In our ultrasound case, a dilated sagittal sinus was observed and a midline supratentorial cystic structure which on colors Doppler presented a turbulent vascular flow, respectively a pronounced multicolored mosaic pattern; in this conditions the diagnosis was clarified at 28 weeks of gestation. In the neonatal period, clinical manifestations may be represented by macrocephaly, loud intracranial bruit and dilatation of orbital veins, seizures, hydrocephalus (caused by the obstruction of the aqueduct of Sylvius by the dilated aneurysm) and subarachnoid hemorrhage (10). In our cases the signs of cardiac failure were developed after 32 weeks of gestation, after the arteriovenous malformation was diagnosed; in that moment, a mild tricuspid insufficiency was present among to a mild right cardiomegaly. In childhood, the patient may present with headache, cognitive dysfunction or a focal neurological deficit caused by the “steal” phenomenon, with the diversion of the blood to the aneurysm and poor circulation into the parenchyma, resulting brain infarcts and perivenricular white matter lesions (11).

One of the greatest complications that could appear in a gigantic dilated vein of Galen is the thrombosis of the aneurysm; two main pathophysiological ways are involved in the spontaneous thrombosis of vein of Galen: slow blood flow and impediment to the venous outflow (12). This complication is encountered rarely, latest case was described by Konovalov in 2002 (13) and is confirmed by CT and MRI when varying degree of calcification are seen.

The aneurysm of the vein of Galen is considered to arise between 6 and 11 weeks of gestation as a result to an insult to the cerebral vasculature. This appears to be more frequent on male fetuses (14). In our case, a female infant was born. Clinically the symptomatology was dominated by signs of heart failure. While in the most cases medical stabilization of heart failure suffices while preparing for interventional procedure, in our case it was impossible.

The treatment of choice is represented by vascular embolization. There were reported good after-procedural results and good neurological outcome (15). Acute cardiac failure, such as in our case necessitates an emergency mode intervention, that is preferred to be performed early in order to prevent irreversible cardiac failure and brain injuries. The success of the embolization is dominated by the necessity of knowing accurately the vascular anatomy of the vein of Galen aneurysm. The MRI is preferred pre-interventional due to the fact that it can “freeze” fetal movements (16).

The aneurysm of the vein of Galen is a rare and severe arteriovenous malformation, with an overall rate of survival of 58% (17). It is considered that above the major impact on the cerebral parameters and heart function, the only measurement that can predict a better or poorer diagnosis is the volume of the aneurysm. As far as we know this is the only case of Galen aneurysmal malformation in Romania which benefited of embolization by interventional treatment in neonatal period.

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