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MRI Presentation of an Aneurysmal Malformation of the Vein of Galen Revealed by Proptosis and Epileptic Seizure in an 18-Month-Old Child: A Case Report

Anouar Benhima[1*], K. Kassi[1], B. Zouita[1], D. Basraoui[1], H. Jalal[1]

[1]Mother and Child Radiology Department of the CHU Mohammed VI, Marrakech Morocco

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***Corresponding author:** Anouar Benhima
Mother and Child Radiology Department of the CHU Mohammed VI, Marrakech Morocco

Abstract **Case Report**

Vein of Galen aneurysm is a rare and complex congenital, vascular malformation of the median line with severe prognosis. It is commonly associated with refractory heart failure. This disorder usually occurs during the neonatal period and in infants. It is characterized by age- dependent clinical polymorphism. Diagnosis is based on cerebral angiography which plays a role in diagnostic and therapeutic decisions. Now, the treatment of choice is based on endovascular embolization which has improved survival rates. We here report a case of Galen aneurysmal malformation in a small child aged 18 months revealed by seizures and bilatéral proptosis.

Keywords: Vein of Galen aneurysm, congenital affection, arteriovenous malformation.

INTRODUCTION

Aneurysm of the ampulla of Galen or aneurysm of the great vein of Galen (AGVG) is a rare, non-hereditary congenital midline vascular malformation located in the pineal region. This embryonic malformation, which occurs between the 6th and 11th week of pregnancy, consists of a series of fistulas leading to an abnormally persistent and dilated median vein of Markowski which is the embryological precursor of the vein of Galen. It is the most frequently diagnosed cerebral vascular malformation in the ante- and neonatal period with a male predominance [1]. The clinical manifestations depend on the age of discovery, and the diagnosis is usually made antenatally on obstetric ultrasound, or during the neonatal period in a picture of congestive heart failure, or postnatally. Clinical presentation in adults is exceptional. The prognosis of aneurysms of the ampulla of Galen is very poor, particularly in the case of neonatal revelation, where death or serious neurological sequelae may occur [2]. With the advent of neuro-interventional radiology techniques, the prospects of successful treatment of these lesions, previously disappointing, are now promoted. Indeed, endovascular embolization has supplanted curative surgery from its difficulties of approach associated with a high mortality rate, and currently represents the treatment of choice for this condition [3].

CASE REPORT

This is an 18-month-old child, who was treated for generalized tonic-clonic seizures and bilateral proptosis. He was born from a full term pregnancy without any notion of peri- or post-natal complication. The clinical examination of the child was apyretic, slightly tachycardic and dyspneic. He presented a bilateral proptosis with macrocrania. A cerebral MRI was requested for better characterization. The MRI revealed a large aneurysmal dilatation of the ampulla of Galen extending to the right sinus, torcular sinus, superior sagittal sinus, transverse sinus, and petrous sinus bilaterally, which was responsible for an active upstream bi-ventricular hydrocephalus (Figure 1). This aneurysmal dilatation is associated with the presence of several drainage veins of the various intraparenchymal and cortical venous tributaries, notably the internal cerebral veins, the basilar veins of Rosenthal, the internal temporal veins and the cavernous sinus (Figure 2).

In front of this MRI aspect, the diagnosis of an aneurysmal malformation of the ampulla of Galen was retained and was referred to the interventional radiology unit to propose treatment by endovascular embolization.

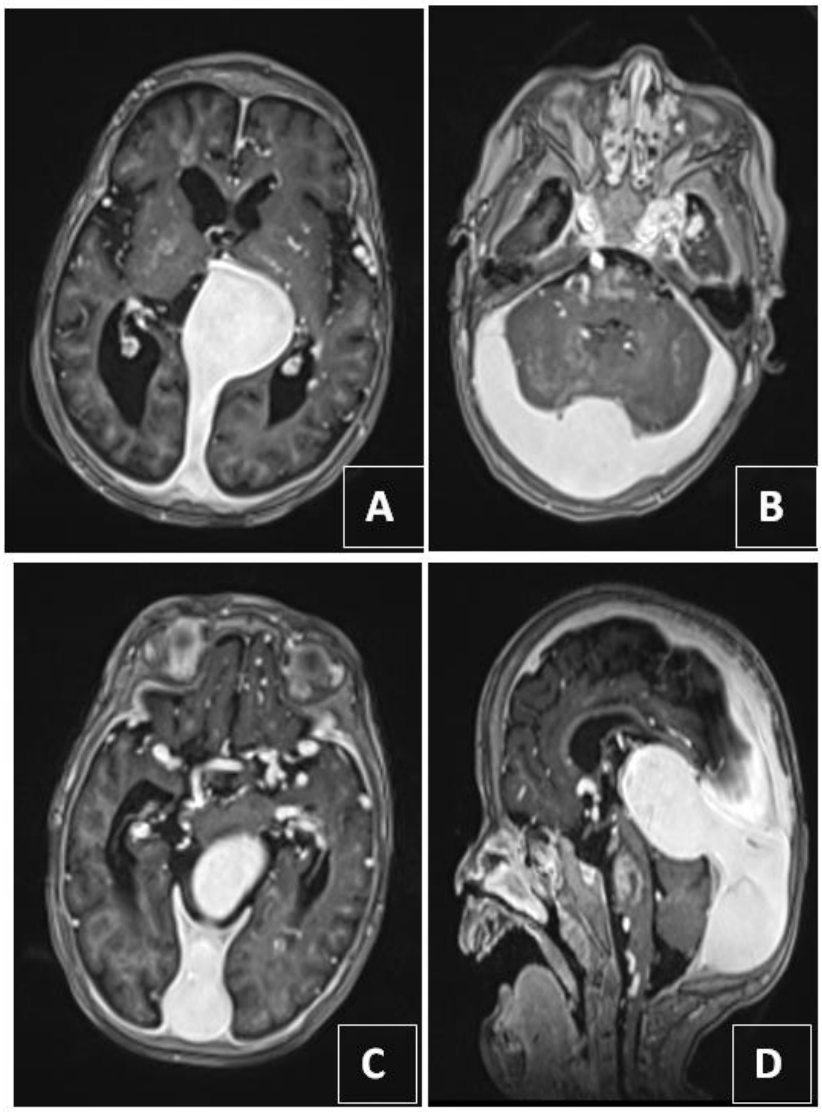

Figure 1: MRI after injection of gadolinium product in T1 axial (A, B, C) and sagittal (D) sections showing vascular-type enhancement of the aneurysmal dilatation of the ampulla of Galen extended to the right sinus, the torcular, the superior sagittal sinus, and the transverse sinuses bilaterally. It is associated with dilatation of several cortical drainage veins, internal cerebral veins, basilar veins of Rosenthal, internal temporal veins and cavernous sinus (yellow arrow)

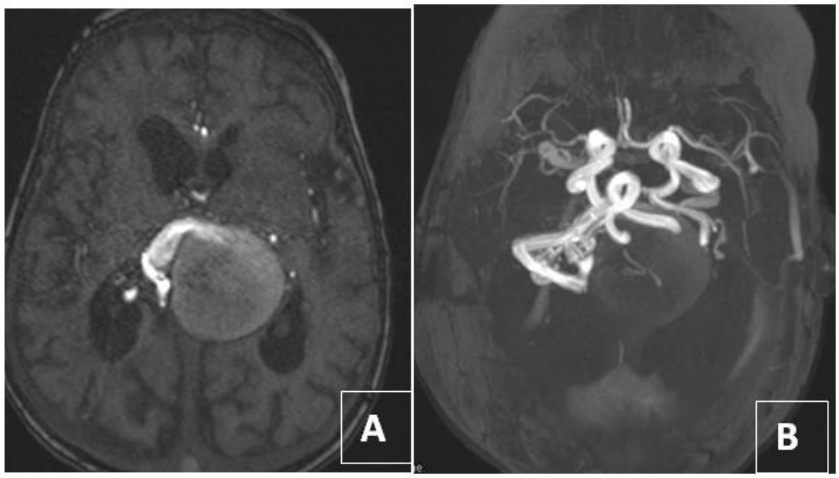

Figure 2: 3D angio sequence in axial section (A): Arteriovenous shunt between the vertebro-basilar system (right posterior cerebral) and the aneurysm of the ampulla of Galen.3D angio sequence (B): Arteriovenous fistula between the vertebro-basilar system (right posterior cerebral) and the aneurysm of the ampulla of Galen

DISCUSSION

The malformation of the vein of Galen was first described by Jager in 1937 [4]. It is a complex arteriovenous fistula caused by multiple abnormal communications between the venous system of Galen and the cerebral arteries. It is a rare congenital condition that represents less than 1% of intracranial arteriovenous malformations [5, 6]. Although rare, it is one of the most frequently diagnosed arteriovenous malformations in children before and after birth. It mainly affects newborns and small children: two thirds of cases are diagnosed before the age of 28 months [2, 3, 7]. The clinical symptomatology of the ampulla of Galen aneurysm is very varied depending on the age of onset. Three periods can be clearly distinguished, namely the neonatal period (from birth to 28 days), the first age (from 1 to 28 months) and the second age and adult (beyond 28 months) [1]. These clinical manifestations are mainly divided into those related to high output heart failure and those involving neurological symptoms due to venous congestion and abnormal CSF flow. Their severity and tolerability are variable and depend on the angioarchitecture of the malformation and the age of the child.

In the neonatal period, the diagnosis is most often made during systematic antenatal ultrasound scans, with clinical symptoms dominated by heart failure of varying severity and occurrence. This cardiac insufficiency is associated with pulmonary hypertension, and can be complicated by renal damage, hepatic insufficiency with a maximum of Hepatic insufficiency with, at the most, a multi-visceral failure picture with a catastrophic, even fatal prognosis. Beyond the age of 2 months, clinical symptoms are dominated by macrocrania and hydrocephalus secondary to compression of the aqueduct of Sylvius by the malformation and to cerebrospinal fluid resorption disorders. Seizures have been noted in 20% of patients and meningeal hemorrhage due to rupture of the aneurysm sac itself, or more frequently dilated veins at a distance from the aneurysm, has been found in 2% of cases [8]. In older children and adults the symptomatology is much more varied, and may include growth retardation, cranial vault hypertrophy, mental retardation, the presence of psychiatric signs or intracerebral calcifications related to chronic venous ischemia. In the face of this variable clinical presentation, imaging is essential to make a positive diagnosis. Currently, the diagnosis of aneurysm of the great vein of Galen is made more and more often in the antenatal period. In the antenatal period, the diagnosis is made in the second or more often in the third trimester of pregnancy during routine ultrasound monitoring. The ultrasound scan shows a strictly anechoic extra parenchymal image of the midline located behind the third ventricle. The so-called racket shape corresponds to venous dilatation draining through a tubular structure corresponding to the falcoriel sinus. Color Doppler confirms the vascular nature of the lesion by demonstrating the flow of the aneurysm in the form of bidirectional turbulent flows. The cardiac impact is also important to evaluate by looking for dilatation of the right cavities, tricuspid regurgitation and dilatation of the jugular veins [9].

MRI is a very useful complementary examination, it helps to differentiate an aneurysmal malformation from an intra-parenchymal arteriovenous malformation draining into the vein of Galien, and allows to specify the parenchymal lesions which can be of leucomalacia or hemorrhagic type. In the neonatal period, transfontanellar ultrasonography is currently a standard examination performed during the first year of life. Coupled with Doppler ultrasound, it allows the study of the deep cerebral vessels, it finds the same semiological elements as in the ante-natal period. On CT scan, arteriovenous malformations are spontaneously discretely hyperdense and sometimes present calcifications within them. After injection, a very intense vascular-type contrast is observed. The malformative vessels appear as tubular and curvilinear opacities. The deep venous drainage of the malformation is often clearly identifiable as a tubular opacity draining into a dilated ampulla of Galen. MRI is the most sensitive examination for the detection of parenchymal lesions and should be performed systematically. MRI allows to better specify the anatomical relationships of the malformation, especially with the deep cerebral structures. The aneurysmal dilatation, arteries and veins are empty of signal on T1 and T2 sequences. Typically medial, the aneurysm is sometimes lateralized retro thalamic, taking a tubular or curvilinear shape, well visible in relation to the brain parenchyma and cerebrospinal fluid, sequelae of old hemorrhages may also be visible as hyper signals [9]. The diffusion sequence shows the lesions of acute cerebral ischemia. At this time of neonatal diagnosis, arteriography is not necessary if no immediate treatment of the malformation is envisaged.

After the neonatal period, even if ultrasound remains interesting to perform as long as the anterior fontanel remains open, finding the signs already mentioned, MRI is the most indicated imaging method allowing a more contributory parenchymal study and to evaluate the exact nature of the lesion. The description of the malformation is identical to the neonatal data. Arteriography is performed around 5-6 months of age when endovascular treatment of the malformation begins. It is diagnostic and therapeutic; it is the reference examination to establish with precision the angioarchitecture of the malformation, to appreciate the flows which circulate in it and to evaluate the quality of the cerebral venous drainage routes. Chest radiography and echocardiography show the cardiopulmonary repercussions of the condition by demonstrating cardiomegaly associated with dilatation of the right cavities and the pulmonary artery [10]. The differential diagnoses of aneurysm of the great vein of Galen are

essentially with the liquid lesions of the midline among others of the cyst of the pineal gland, colloid cyst, arachnoid cyst, subependymal pseudocysts [11]. Another surgical alternative with disappointing results, endovascular embolization methods currently represent the treatment of choice with a satisfactory success rate but requires a team of experienced neuroradiologists. The goal of the treatment is occlusion of the shunts of the malformation. Its effectiveness depends largely on the size of the malformations and the complications developed [5]. The prognosis of the condition depends on the severity of the heart failure but also on the extent of the cerebral ischemia. Early diagnosis is fundamental to the prognosis [12].

CONCLUSION

Aneurysm of the ampulla of Galen is a rare congenital vascular malformation. It is defined by the association of one or more arteriovenous fistulas running into the ampulla of Galen or one of its immediate venous afferents. This condition mainly affects newborns and small children. Its clinical manifestations depend on the age of discovery. The progress of obstetrical ultrasound and the development of Doppler techniques have made it possible to improve diagnostic possibilities diagnostic possibilities from the antenatal period. Cerebral angiography is the key examination from its diagnostic and therapeutic role. Endovascular embolization is currently the treatment of choice for this pathology with a survival rate that is constantly evolving according to the teams.

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