

Ruptured Cerebral Aneurysm in the Early Stage of Life – a Congenital Origin?

C. Pollo¹
 K. Meagher-Villmure²
 M.-A. Bernath³
 O. Vernet¹
 L. Regli¹

Abstract

We report the case of a large fusiform aneurysm of the anterior cerebral artery in a 3-month-old girl presenting with generalized seizures and complicated by a cardiorespiratory arrest needing resuscitation. The native cerebral CT scan revealed a diffuse subarachnoid hemorrhage (Fischer III grade); CT angiography showed an aneurysm of the distal A1 segment of the anterior cerebral artery (ACA). Successful treatment was achieved through surgical trapping of the aneurysm and sacrifice of the distal A1 segment. A histological study of the aneurysmal wall revealed the absence of elastic fibers in an otherwise fibromuscular media and showed no signs of previous hemorrhage. Intracranial aneurysms of the ACA are very rare in the early stage of life and their pathogenesis is not clear. This case is of interest as the location and shape of the aneurysm, the absence of relevant familial and medical history, and the histopathological findings raise the hypothesis of a congenital origin. It further describes the value of CT angiography for studying aneurysms in the very young.

Key words

Anterior cerebral artery aneurysm · intracranial aneurysm · newborn · histopathology · congenital abnormalities

Introduction

Intracranial aneurysms in the pediatric population under the age of 5 years are extremely rare. In contrast to adults, they occur more frequently at the carotid artery bifurcation, the distal por-

tions of the middle cerebral artery, and the posterior circulation [6,9]. The pathogenesis of aneurysmal formation remains unclear and may result from both congenital and acquired factors [3,4,7,8].

We report the case of a ruptured fusiform aneurysm of the anterior cerebral artery (ACA) in a 3-month-old baby and describe the structural abnormalities observed in the arterial wall. We address the clinical, morphological, and histopathological findings that support the hypothesis of a possible congenital defect in the arterial wall leading to the aneurysm development.

Case Report

A 3-month-old girl, with no familial history of cerebrovascular disease, born full-term without birth trauma or congenital illness, presented with generalized seizures followed by a cardiorespiratory arrest needing resuscitation. The anterior fontanelle was bulging and there was no evidence of meningismus.

The native cerebral CT scan (Fig. 1a) showed a diffuse subarachnoid hemorrhage (Fischer III grade) and ventricular dilatation; contrast enhancement revealed a large aneurysm of the anterior communicating artery (AcomA) complex measuring 7 × 9 × 11 mm (Fig. 1b). A CT angiography confirmed the diagnosis of an AcomA complex aneurysm projecting upwards and slightly posterior (Fig. 1c).

A right frontal ventriculostomy was performed and the aneurysm was approached through a right pterional craniotomy. The

Affiliation

¹ Department of Neurosurgery, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland

² Department of Pediatric Pathology, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland

³ Department of Anesthesiology, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland

Correspondence

Luca Regli, M.D. · Neurosurgical Department · Centre Hospitalier Universitaire Vaudois · Av. du Bugnon · 1011 Lausanne · Switzerland · E-mail: luca.regli@chuv.hospvd.ch

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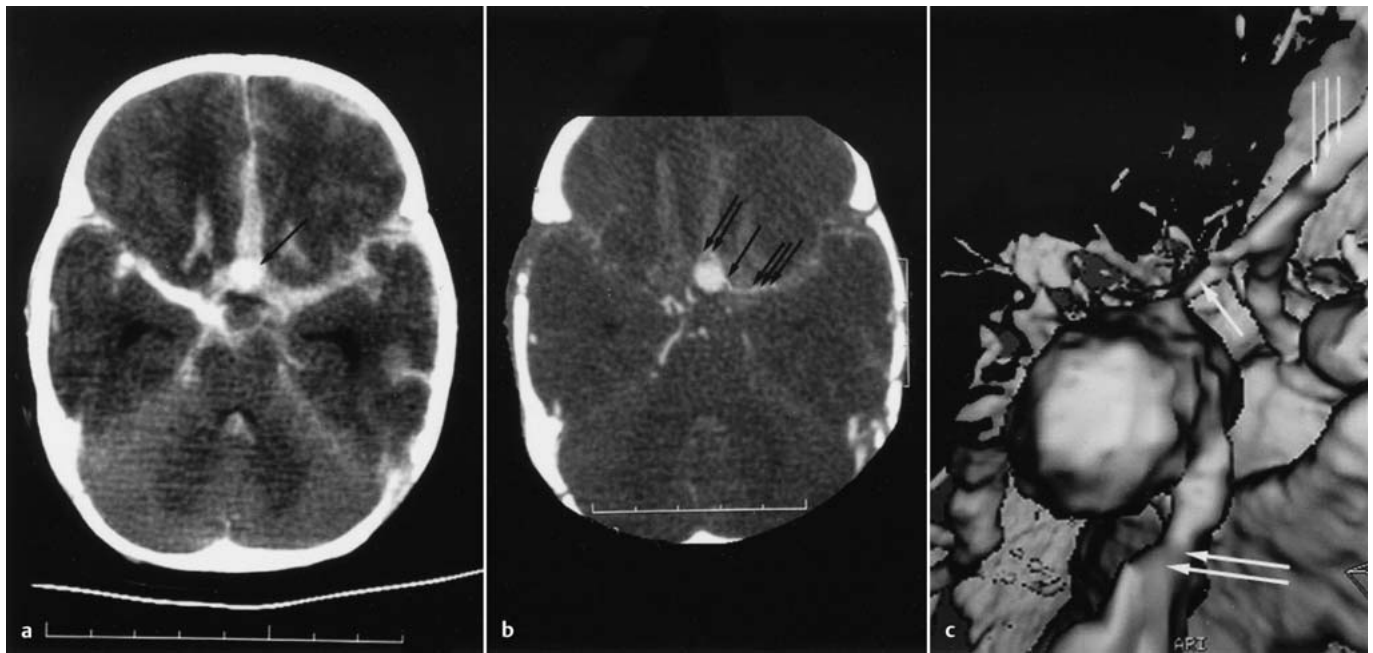


Fig. 1 a to c **a** Contrast CT showing subarachnoid hemorrhage and the aneurysm (arrow) located on the anterior communicating artery complex. **b** Source image of the CT angiography showing the relation between the aneurysm and the left A1 segment (single arrow), both A2 segments of the anterior cerebral artery (double arrow) and

the left middle cerebral artery (triple arrow). **c** Superior, anterior, and right view of the 3D CT angiogram showing the projection of the aneurysm, left A1 segment (single arrow), and both A2 segments of the anterior cerebral artery (double arrow). Triple arrow: left middle cerebral artery.

aneurysm was found to originate from the distal left A1 segment which was incorporated into the aneurysmal wall, without any identifiable neck. The aneurysm was trapped between 2 clips, the first proximal to the aneurysm on the A1 segment, the second distal to the aneurysm, just proximal to A1-A2 transition preserving the AcomA. Both A2 segments filled from the right A1 segment through the AcomA.

The aneurysm was resected and histological study showed a portion of the vessel wall made of a band of smooth muscle fibers partly dissociated by collagen fibers and acute hemorrhagic necrotic debris with polymorphs, fibrin, and red blood cells. No elastic fibers could be visualized in the wall of the vessel and no feature of previous hemorrhage either (Fig. 2).

A postoperative cerebral perfusion CT scan performed in the anterior circulation revealed normal blood flow, symmetrical blood volume, and mean transit time values confirming adequate collateral flow and suggesting no evidence of vasospasm.

The postoperative clinical course was uneventful. Weaning of the external ventricular drain resulted in progressive hydrocephalus treated by ventriculoperitoneal shunting

At 2 years follow-up, neurological and neuro-developmental examinations were normal.

Discussion

Pediatric aneurysms are known to represent 0.17 to 4.6% of all intracranial aneurysms. Among the pediatric population, early

childhood aneurysms (age 5 years or less) represent only 0.6 – 1.9% of the cases and are more frequent during the first 2 years of life. A male preponderance of 2 – 3 : 1 has been documented [6].

Most authors agree that the internal carotid artery bifurcation (ICA) is the main location of intracranial aneurysms in infants (36 – 50%) [6]. Tekkök et al. reviewed 54 cases of aneurysms presenting in the first year of life and observed that the middle cerebral artery (MCA), especially its distal branches, was involved in 45.5% of cases whereas ICA bifurcation aneurysms were encountered in 9.1% of cases [9]. Posterior circulation aneurysms were also more frequently observed in children (up to 31.4%) than in adults (10 – 15%) [6,9]. ACA aneurysms were reported in only 7.2% of cases and more than 75% of children under 5 years had large (> 10 mm) or giant (> 25 mm) lesions [9].

Angiography remains the gold standard in the diagnosis and pre-surgical evaluation of intracranial aneurysms. Although it may be performed with acceptable risks in experienced neurovascular centers, increased risks have been observed in very young children [9]. Interestingly, in this case, CT angiography showed useful angioanatomy for the preoperative evaluation of the aneurysm, avoiding the need for angiography. This corroborates our results obtained with CT angiography compared to digital subtraction angiography in adult patients investigated for aneurysms [11] and supports the idea that CT angiography may be adequate for investigating aneurysms in the very young child.

Management of aneurysms in children is usually difficult because of their large size, wall fragility, and absence of identifiable base and should be discussed by a multidisciplinary team.

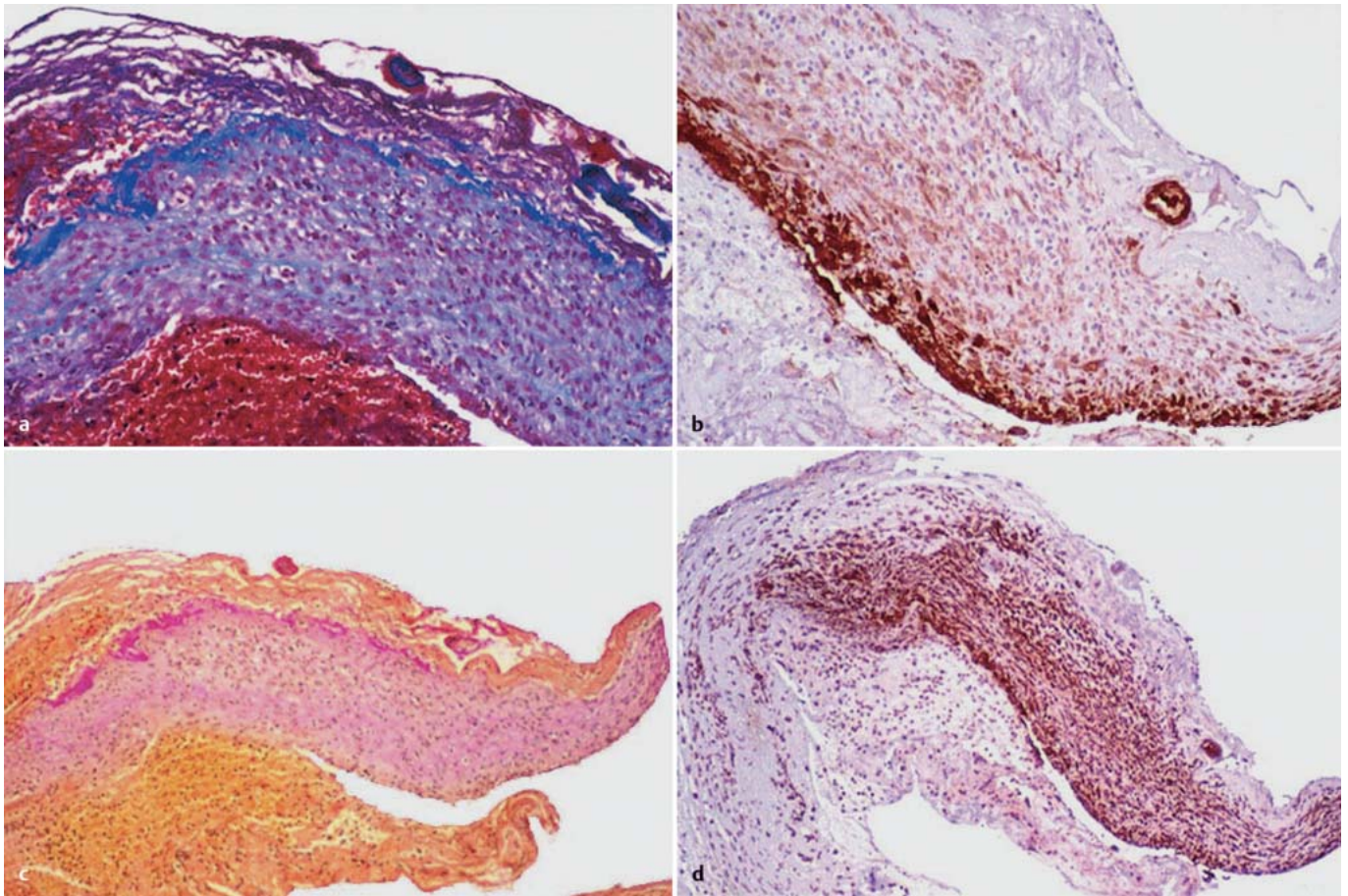


Fig. 2a to d **a** Fibrous tissue (blue) and smooth muscle fibers (purple) within the vessel wall by Masson's trichrome stain, $\times 250$. **b** Immunohistochemistry with SMA antibodies showing the positive smooth muscle fibers within the vessel wall, $\times 250$. **c** No elastic fibers

demonstrated with VanGieson Elastic stain, fibrous tissue in pink, $\times 250$. **d** Immunohistochemistry with Vimentin showing the positive collagen fibers within the vessel wall, $\times 250$.

Although endovascular treatment has been reported in older infants [1], it still needs to be validated on larger series, especially in early childhood, as the long-term stability is of particular importance when dealing with a pediatric population. In our patient, the lack of an aneurysmal neck and the vessel fragility prevented direct clip-reconstruction of the parental artery.

By trapping the aneurysm and occlusion of the proximal ACA (A1 segment), the hemodynamic stress on the AComA will increase. The incidence of de novo aneurysm formation after vessel occlusion, such as the carotid artery, was reported to be up to 4% [10]. Therefore, long-term follow-up, preferentially using non-invasive imaging to detect the possible development of an aneurysm in the AcomA region, should be done in these patients.

Several hypotheses regarding the mechanisms of aneurysm formation in young children have been reported: hemodynamic stress, trauma, degenerative changes, and congenital defects of the vessel wall.

Some embryologic studies suggest that a *hemodynamic stress* may lead to the development of cerebral aneurysms. Embryologically, the posterior circulation is more prominent than the anterior. This could explain the higher incidence of posterior aneurysms in children than in adults. In early childhood, the anterior

circulation will become more important with more prominent growth than the posterior circulation. This could be the explanation for aneurysms on the MCA, which develops earlier than other cerebral vessels and supplies more blood flow to the developing cerebral hemispheres, accounting for the more frequent occurrence of aneurysms on the MCA in young children [7].

A *birth trauma* may damage the arterial wall and may explain aneurysms developing in the area of dural folds [5]. Lam et al. documented one case of pericallosal aneurysm from shaken baby syndrome and postulated the same etiology [2]. *Degenerative changes* in the arterial wall have been described to result from bacterial damage [7]. This could also explain the preferential distal location of aneurysms in early childhood. The hypothesis of a *congenital defect* in the arterial wall is supported by the fact that some inherited connective tissue disorders (Ehlers-Danlos type IV, Marfan syndrome, neurofibromatosis type I, polycystic kidney disease) have been associated with a higher incidence of cerebral aneurysms [3, 4]. Furthermore, a microscopic study carried out in a neonate with aneurysms showing fragmentation or absence of the internal elastic lamina and smooth muscle within the aneurysmal wall and no inflammatory process also supports this [8].

The histological findings from our case report demonstrate an incomplete vascular wall partly made of a band of smooth muscle fibers and collagen and further favor the hypothesis of a congenital defect of the arterial wall. The usual internal and external elastic fibers were completely absent in an otherwise fibromuscular media. Although these findings may be encountered in an old scarring process resulting from a perinatal or *in utero* vessel injury, the following observations are in favor of a developmental abnormality of the vascular wall:

1. Unlike acquired aneurysms of traumatic or infectious origin, the aneurysm was located proximal on the vascular tree (A1 segment of ACA);
2. there were neither clinical nor radiological features of shaken baby syndrome and there was no history of birth trauma or previous bacterial infectious disease;
3. hemodynamic stress is unlikely because the aneurysm was not located on a dominant A1 segment nor on a bifurcation.

Ruptured cerebral aneurysms are very rare in the early stage of life. Like in the adult populations, CT angiography may be sufficient for the presurgical work-up of individual cases or for emergency situations. In the absence of dysplastic vascular systemic syndromes clinical, morphological, and histological findings support that such aneurysms may result from a focal congenital vascular wall defect.

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