

# Ruptured Giant Left Distal Anterior Cerebral Artery Aneurysm in a Two-Month-Old Baby

Nik Qisti Fathi<sup>a</sup> Ainul J. Syahrilfazli<sup>a</sup> Abu Bakar Azizi<sup>a</sup> Ismail Mohd Redzuan<sup>b</sup>  
Muda Sobri<sup>b</sup> Ramesh Kumar<sup>a</sup>

<sup>a</sup>Neurosurgical Unit, Department of Surgery and <sup>b</sup>Department of Radiology, University Kebangsaan Malaysia Medical Centre, Kuala Lumpur, Malaysia

## Established Facts

- Giant aneurysms in infancy are more common in the posterior circulation. Our case was of an anterior aneurysm. This article demonstrates the rarity of such a case.

## Novel Insights

- A detailed angiographic investigation is important prior to embarking on a definitive surgical treatment.

## Key Words

Intracranial aneurysm · Giant aneurysm · Paediatric case

## Abstract

**Background:** The incidence of intracranial aneurysm in childhood is rare, especially in infancy. In the literature, most of childhood intracranial aneurysms occur in the posterior and middle cerebral circulation. We report a case of a ruptured giant left distal anterior cerebral artery aneurysm in a 2-month-old baby. This report will discuss the rarity of this case as well as the diagnostic and surgical challenges in

treating this condition. **Case Report:** A 2-month-old baby girl presented to our centre with a 1-day history of multiple afebrile seizures. A CT scan of her brain showed a large frontal interhemispheric acute bleed with intraventricular extension and acute hydrocephalus. An external ventricular drain was inserted and she was nursed in the Paediatric Intensive Care Unit. Subsequent CT angiogram and 4-vessel angiogram showed a giant aneurysm originating from the A3 segment of the left anterior cerebral artery. She underwent craniotomy and clipping and excision of the giant aneurysm. **Discussion:** Giant distal anterior artery aneurysms are very rare in infants. They are more commonly associated with the

posterior and middle cerebral arteries. The aneurysms in infants tend to be larger compared to adults and they commonly present with subarachnoid haemorrhage and seizures. CT angiogram and 4-vessel cerebral angiogram are important diagnostic tools in this case. The angiogram demonstrated no distal runoff from the aneurysm and this assisted in our planning for surgical treatment of the aneurysm. A bifrontal craniotomy was done and the giant aneurysm was approached interhemispherically. The proximal and distal parent vessel was clipped and the aneurysm wall was excised. The baby recovered from surgery and did not require any cerebrospinal fluid diversion. **Conclusion:** Giant distal anterior cerebral artery aneurysms are rare in infants. Detailed angiographic investigation is important. Surgery is the treatment of choice, although there may be a role for endovascular intervention.

© 2015 S. Karger AG, Basel

## Introduction

Intracranial aneurysm in the paediatric population ( $\leq 18$  years old) is very rare, especially in infancy [1–3]. The incidence has been reported as being within the range of 0.5–4.6% [4]. The characteristics of this vascular entity differ from the adult population in many ways, including morphology, sexual preponderance, clinical presentation, and radiological features. Evidence from many reports has been consistent to show that there is a male sexual predominance [3, 4]. It is established that paediatric intracranial aneurysms are found more frequently in the anterior cerebral circulation rather than the posterior circulation [3–5].

However, giant aneurysms in infants, on the contrary, arise more frequently in the posterior circulation. A systematic review by Buis et al. [3] has revealed a significantly higher prevalence of giant aneurysms in the posterior circulation in this subgroup of paediatric patients. Therefore, it is safe to assume that cases of infants with giant aneurysms in the anterior circulation are extremely rare.

From this small group of patients with giant anterior circulation aneurysms, even a smaller fraction has aneurysms originating from the anterior cerebral artery. Huang et al. [4] and Aryan et al. [5] reported a prevalence of 0.11–0.06% of patients, respectively, with intracranial aneurysms from this particular cerebral vessel.

Despite modern day diagnostic tools and modalities of treatment, the management of paediatric intracranial aneurysms remains treacherous. Intervention in ruptured paediatric aneurysms is of paramount importance as this

foreshadows early recurrent haemorrhage in an age group not affected by either stress or hypertension [6]. At present, the intervention may either be surgical or radiological. There is no consensus for the management of paediatric aneurysms and it remains unique on a case-by-case basis. Appropriate imaging investigations are pivotal in ensuring good surgical outcome.

We report a case of a 2-month-old baby girl who presented with a ruptured giant aneurysm located on the left distal anterior cerebral artery, rendering it an extremely rare case in neurosurgery.

## Case Report

A 2-month-old baby presented to our centre with a 1-day history of four episodes of afebrile seizures. The first two episodes aborted spontaneously but the remaining episodes needed intervention with medical therapy, including the use of rectal diazepam and intravenous phenytoin. At presentation the patient had a full Glasgow Coma Score. She was moving all her limbs spontaneously despite being a bit hypertonic on closer examination. The limbs also exhibited brisk reflexes. Her vital signs were stable and she was adequately perfused. The right pupil was 5 mm and the left 4 mm – both equally reactive to light. The retinas were normal on fundoscopy examination with no significant papilloedema. However, there was bulging of the fontanelles. Clinically, there was no evidence of trauma or any signs to suggest non-accidental injuries.

Her medical history was unremarkable and none of her family members had any history of intracranial pathologies. There was no history of maternal infection. The patient is the youngest of 3 siblings. The rest of the siblings were healthy and well. They have good social support and are well taken care of. The mother works as a primary school teacher and the father is a government servant. The infant was born full term via a spontaneous vaginal delivery with a good birth weight of 2.8 kg. No birth trauma was ever recorded. She was thriving well with a weight of 5.1 kg at presentation. There was low risk of HIV infection. A paediatric cardiologist did an echocardiogram to investigate the possibility of a mycotic aneurysm. The investigation showed normal valves without any thrombus or vegetations. Essentially, it showed a normal intracardiac anatomy and function.

An initial non-contrast CT scan of her brain showed a large bifrontal intraparenchymal bleed. A round hyperdense area was seen anterior to the frontal bleed at the midline with extension into the corpus callosum and lateral, third and fourth ventricles without significant midline shift. Following this, an external ventricular drain was inserted (fig. 1) and she was subsequently nursed in the Paediatric Intensive Care Unit.

A subsequent CT angiogram showed a well-defined hyperdense lesion in the midline frontal region with avid homogenous enhancement measuring  $2.8 \times 2.8 \times 3.1$  cm (fig. 2a, b). The lesion was seen at the distal inferior sagittal sinus. However, the inferior sagittal sinus seemed to be patent with normal calibre. The middle cerebral arteries, posterior cerebral arteries and A2 segments bilaterally were also patent with normal calibre. The anterior cerebral



**Fig. 1.** An axial view of the brain on CT scan showing the giant aneurysm and an extracranial drain in situ.

arteries were not visualized. These features were suggestive of an aneurysm arising from the anterior cerebral circulation. We proceeded with a 4-vessel angiogram that showed a giant aneurysm measuring  $2.8 \times 3.1$  cm arising from the A3 segment of the left anterior cerebral artery (fig. 3a, b). The right internal carotid artery and both middle cerebral arteries were of normal calibre. No other unique vascular abnormalities such as an arteriovenous malformation were identified from the angiographic images.

A bifrontal craniotomy was performed and the giant aneurysm was approached interhemispherically. The baby was placed in supine position under general anaesthesia. A bicoronal incision was made followed by a bifrontal craniotomy that revealed an oedematous brain. The dura was opened and the superior sagittal sinus divided and ligated to allow an anterior interhemispheric approach to the aneurysm. To facilitate orientation, the right callosomarginal artery was subsequently identified. The aneurysm, measuring 3 cm in diameter, was found arising from the left callosomarginal artery and was dissected. Intraoperative Doppler was used to confirm that there was no flow in the distal vessel. The proximal and distal parent vessels were clipped to allow safe opening of the aneurysm. Clots found within were evacuated. Finally, the wall of the aneurysm was excised. No vessel reconstruction was performed; the segment was occluded and sacrificed. Intraoperatively, there were no other abnormalities of the vascular system concurring with the preoperative angiographic findings. The intraventricular clot was not evacuated and the subarachnoid haemorrhage was not washed out. This was subsequently managed via a ventricular drain until the cerebrospinal fluid cleared. No ventriculoperitoneal shunt was required.

The postoperative period was uneventful. A plain CT brain scan was done on the second day postoperatively and only showed

residual bleed with associated postoperative changes. The infant was extubated on the third postoperative day and had no neurological deficit. The extraventricular drain was eventually removed at 2 weeks after surgery. She was allowed home 19 days after surgery. Her neurological examination was essentially normal. However, she could not regain her sucking ability and was therefore sent home with a nasogastric tube with an appropriate feeding regime.

At 1 month after surgery the baby was seen again in our follow-up clinic. The histopathological report was of a ruptured aneurysm. Her neurological examination was essentially normal and she was already starting to tolerate oral feeds better. The oral intake recovery was aided by adequate swallowing physiotherapy. Subsequent follow-ups showed that she had a mild global developmental delay. On year 2 follow-up her gross motor assessment showed that she was still unable to sit and had poor head control but was able to turn her torso from supine to prone. She was attending physiotherapy twice a week. Physical examination showed a slightly increased tone in all limbs with hyperreflexia. Plantar reflexes were upgoing. At 2 years postoperatively, a reassessment magnetic resonance angiogram was done that showed no evidence of cerebral aneurysm or stenosis in the vasculature (fig. 4a). There were cystic changes that return cerebrospinal fluid intensity with brain parenchymal volume loss at the bilateral frontal regions in keeping with encephalomalacic changes due to the previous surgical intervention (fig. 4b).

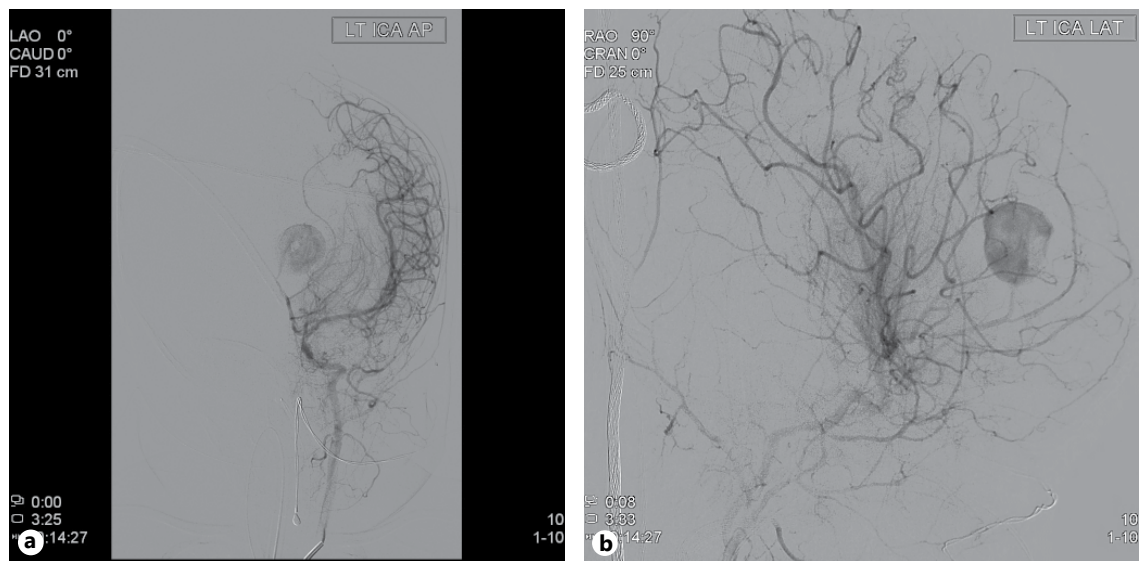
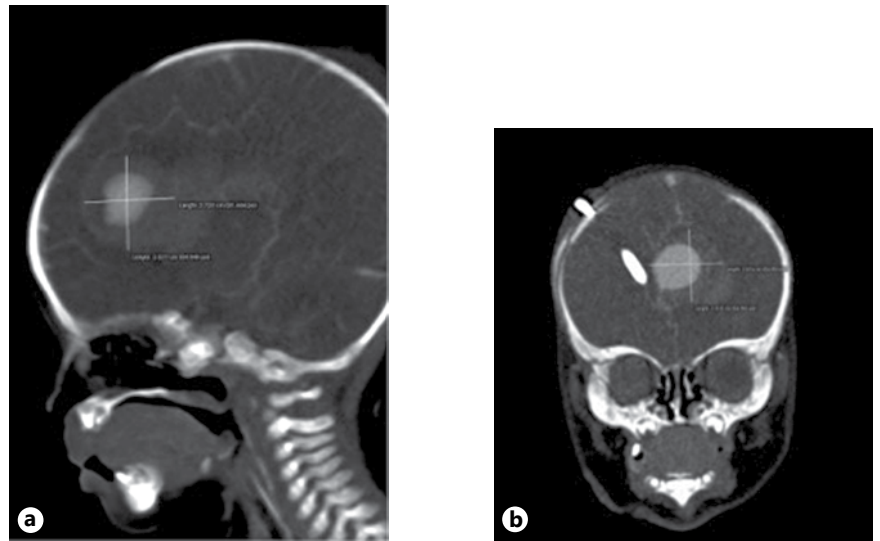
## Discussion

Intracranial aneurysms are extremely rare in infancy. Most academic neurosurgical centres are estimated to treat 1 paediatric aneurysm case per year [4]. The mean age at diagnosis of an aneurysm is  $4.9 \pm 3.5$  months. Infants presenting with an initial haemorrhage were found to be significantly younger according to a literature review by Buis et al. [3]. By and large, in patients who present with initial haemorrhage there is a subarachnoid component to it. Giant aneurysms, in particular, are significantly more often located in the posterior circulation. However, our patient presented with a ruptured giant aneurysm arising from the left anterior cerebral artery. Drowsiness is the main presenting complaint in most cases, whereas a non-ruptured aneurysm is significantly associated with a cranial nerve deficit [3].

Ferrante et al. [7] reported an incidence of 28.6% of giant aneurysms in a case series of 71 patients who were less than 5 years old. Similarly, Buis et al. [3] reported an incidence of 31% of giant aneurysms in the subgroup of infants. Statistically, the proportion of giant aneurysms in children is the same as in their adult counterparts.

As with any other treatment to disease, the treatment of intracranial aneurysms has undergone major improvements over the years, catering specifically to patients of

**Fig. 2. a** A sagittal view of a reconstructed CT angiogram showing the giant aneurysm with the associated mural thrombus measuring  $2.72 \times 2.83$  cm. **b** A coronal view of the reconstructed CT angiogram showing the giant aneurysm with the associated mural thrombus measuring  $2.83 \times 2.82$  cm.

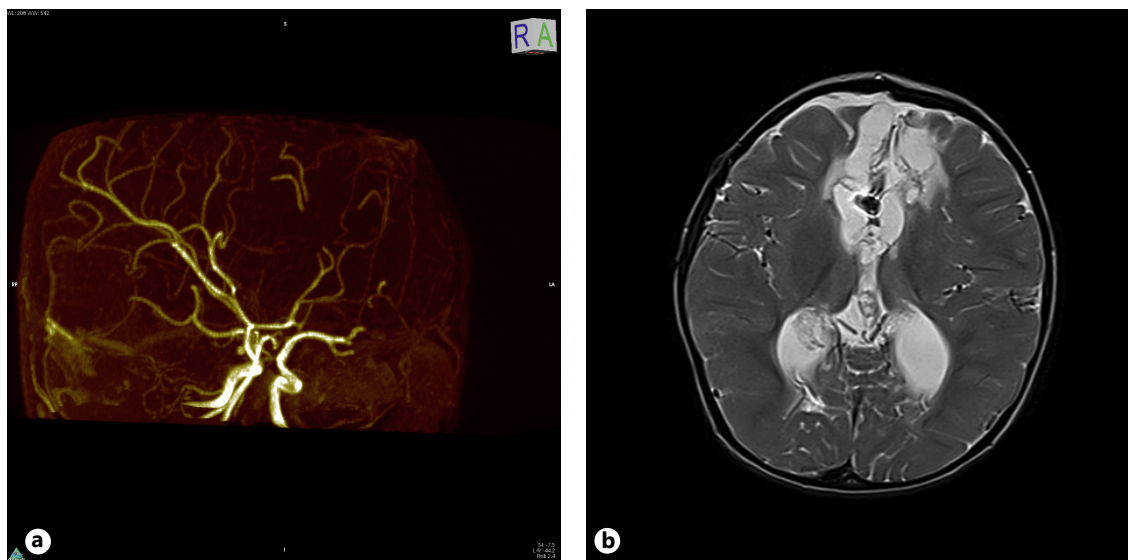


**Fig. 3. a** A left anteroposterior view of the cerebral angiogram with the blushing of contrast within the giant aneurysm. **b** A left lateral view of the cerebral angiogram with a clear view of the giant aneurysm.

different age groups and aneurysmal morphologies. The traditional conservative approach has been replaced in favour of more definitive therapies. Currently, there are advocates for endovascular intervention in ruptured aneurysms in paediatric cases. The proficiency of endovascular intervention is increasing at a fast rate but long-term data are still lacking and the issue of coil durability remains a concern. Stiefel et al. [8], in a case series of 13 paediatric patients over 12 years, showed that the results comparing surgical and endovascular intervention were equivocal.

In most cases, the aetiology of infant intracranial aneurysms remains unknown. This is consistent with our case, as the infant presented with an aneurysm that could not have risen from inflammatory, traumatic or infective causes.

Krings et al. [9] proposes that the pathogenesis of giant aneurysms differs from those of saccular or non-thrombosed saccular aneurysms. Giant intracranial aneurysm is a lesion that is regarded as a proliferative disease of the vessel wall with growth induced by extravascular activity. Recurrent bleeding from the vasa vasorum surrounding the



**Fig. 4. a** A reconstructed 3D magnetic resonance angiogram of the brain at 2 years after clipping of the aneurysm. **b** Axial view of a T2-weighted MRI demonstrating the absence of aneurysm at 2 years postoperatively. There is only a mild degree of hydrocephalus.

giant aneurysm results in an increase in aneurysmal size and in further proliferation of new membranes and vessels. This is in contrast to the previous belief that intraluminal factors, such as change in haemodynamics and weak vessel walls, lead to the increase in size. Furthermore, the thrombosed components in giant aneurysms seen on imaging studies are not intraluminal but are likely to be intramural haematomas similar to those seen on large arterial dissections [10]. Because of this histological consideration, complete surgical excision was considered the best therapy. Furthermore, the results of endovascular parent artery occlusion are not predictable and it has been shown that these aneurysms can continue to grow even after successful occlusion of the parent artery due to the persistence of vessel wall disease. In view of this, we chose to treat the aneurysm by surgical clipping followed by an excision.

The initial imaging investigations were crucial in helping us to decide on treatment. We considered performing magnetic resonance angiography for the patient but this was not possible at our centre due to technical reasons. The CT angiogram was aptly followed by a 4-vessel angiogram that showed the exact location and size of the aneurysm at the distal anterior cerebral artery. In general, these aneurysms are technically challenging to approach given the deep location and small size. In this case, however, the aneurysm was large and since there was no distal runoff, as shown by the angiogram images, the approach and surgical plan was made easier.

Aneurysm cases such as this are usually discussed in a multidisciplinary meeting between neurosurgeons and interventional neuroradiologists at our centre. An agreed treatment plan is made after consideration of multiple factors, including the aneurysmal type, the surgical approach and, more importantly, the expected outcome of treatment. The feasibility of endovascular intervention was discussed and was considered as a second-line treatment in this case. Nonetheless, it remains an option, especially in selected cases with multiple comorbidities.

Surgical intervention was justified in this case and the patient made a good recovery without requiring any cerebrospinal fluid diversion despite the large amount of intraventricular blood.

## Conclusion

Giant distal anterior cerebral artery aneurysms are rare in infants. Detailed angiographic investigation is important. In this case surgery was the treatment of choice, although there may be a role for endovascular intervention.

## Disclosure Statement

The authors have no personal financial or institutional interest in any of the drugs, materials or devices described in this article.

## References

- 1 Lansen TA, Kasoff SS, Arguelles JH: Giant pediatric aneurysm treated with ligation of the middle cerebral artery with the Drake tourniquet and extracranial-intracranial bypass. *Neurosurgery* 1989;25:81–85.
- 2 Mrak G, Paladino J, Stambolija V, Nemir J, Sekhar LN: Treatment of giant and large fusiform MCA aneurysms with excision and interposition radial artery graft in a four-year-old child. *Neurosurgery* 2013, Epub ahead of print.
- 3 Buis DR, van Ouwerkerk WJR, Takahata H, Vandertop WP: Intracranial aneurysms in children under 1 year of age: a systematic review of the literature. *Childs Nerv Syst* 2006;22:1395–1409.
- 4 Huang J, McGirt MJ, Gailloud P, Tamargo RJ: Intracranial aneurysms in the pediatric population: case series and literature review. *Surg Neurol* 2005;63:424–432.
- 5 Aryan HE, Giannotta SL, Fukushima T, Park MS, Ozgur BM, Levy ML: Aneurysms in children: review of 15 years' experience. *J Clin Neurosci* 2006;13:188–192.
- 6 Weir B: *Aneurysms affecting the nervous system*. Baltimore, Williams & Wilkins, 1987, pp 364–373.
- 7 Ferrante L, Fortuna A, Celli P, Santoro A, Fraioli B: Intracranial arterial aneurysms in early childhood. *Surg Neurol* 1988;29:39–56.
- 8 Stiefel MF, Heuer GG, Basil AK, Weigele JB, Sutton LN, Hurst RW, Storm PB: Endovascular and surgical treatment of ruptured cerebral aneurysms in pediatric patients. *Neurosurgery* 2008;63:859–865.
- 9 Krings T, Piske RL, Lasjaunias PL: Intracranial arterial aneurysm vasculopathies: targeting the outer vessel wall. *Neuroradiology* 2005;47:931–937.
- 10 Yonas H, Agamanolis D, Takoka Y, White RJ: Dissecting intracranial aneurysms. *Surg Neurol* 1977;8:407–415.