Case Report

Clipping of a ruptured cerebral aneurysm in a toddler: a case report and review of aneurysmal treatment in children

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Background: Pediatric intracranial aneurysms account for 5% of all aneurysms and less than 10% of all aneurysms cause non-traumatic intracranial hemorrhage in children. They are most commonly secondary to trauma, infection, or genetic etiologies; however, case reports have described iatrogenic intracranial aneurysms. We describe a case of a ruptured aneurysm with an associated intracranial hematoma that was treated by surgical clipping and clot evacuation.

Case Description: The patient was a 15-month-old boy without a history of trauma or infection, who developed acute-onset nausea and subsequent neurological deterioration and status epilepticus. Imaging demonstrated a 13-mm saccular anterior cerebral artery aneurysm with accompanying large left frontal intraparenchymal hematoma and intraventricular hemorrhage. He was treated with urgent craniotomy for surgical clipping and clot evacuation. An external ventricular drain was placed to treat the hydrocephalus. The patient subsequently received a ventriculoperitoneal shunt with rehabilitation disposition. There was a family history of intracranial aneurysms and cavernous malformations; however, genetic testing was negative. One year later, he is developing his speech and has ambulated independently.

Conclusions: We highlight the importance of expeditious care in toddlers with rapidly deteriorating neurological examination results and associated intracranial findings. We demonstrate the rarity of intracranial aneurysms in a toddler and the need for further study on this topic, as there is no clear etiology for this finding in this patient.

Keywords: Cerebral aneurysm; neurosurgery; cerebrovascular; case report

Submitted Jul 25, 2024. Accepted for publication Sep 29, 2024. Published online Oct 28, 2024.

doi: 10.21037/tp-24-289

View this article at: https://dx.doi.org/10.21037/tp-24-289

Introduction

Background

Pediatric intracranial aneurysms are extraordinarily rare vascular pathologies, constituting less than 5% of all aneurysms (1,2). Of all nontraumatic pediatric hemorrhages, up to 40–90% are secondary to vascular malformations, with most arising from arteriovenous malformations (3). Most pediatric aneurysms are pseudoaneurysms, typically secondary to trauma, infection, or arterial dissection, and exhibit significantly different presentations and outcomes than adult aneurysms (4,5). Congenital aneurysms may occur due to underlying vascular wall abnormalities, such as polycystic kidney disease or connective tissue syndromes (5). The treatment of pediatric aneurysms presents significant challenges due to the complexity and acuity of these cases.

Rationale and knowledge gap

There is a limited description of sporadic distal intracranial

aneurysms in the young, and we describe a rare case of a healthy 15-month-old boy who developed acute onset emesis and status epilepticus, secondary to a ruptured distal anterior cerebral artery (ACA) aneurysm. There is also a sparse description of the use of indocyanine green (ICG) video angiography in this young population.

Objective

This case highlights the challenges and considerations in the management of rare and complex vascular pathologies in young children. We also characterized the role of the microsurgical technique given the associated mass lesion causing significant intracranial hypertension. Here, we present this case in accordance with the CARE reporting checklist (available at https://tp.amegroups.com/article/view/10.21037/tp-24-289/rc).

Case presentation

All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's mother for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

A previously healthy 15-month-old boy presented with acute onset of vomiting and mental status deterioration, with subsequent status epilepticus. Subsequently, the patient was intubated. He withdrew his left upper extremity but extended his other limbs. There were no reports of preceding trauma or signs of infection. Family history was negative for intracranial aneurysms, connective tissue disorders, and genetic kidney diseases. Initial head computed tomography (CT) revealed a large left frontal intraparenchymal hemorrhage (IPH) with diffuse intraventricular hemorrhage (IVH) with associated hydrocephalus (*Figure 1*). Subsequent CT angiography identified a large 11 mm × 13 mm aneurysmal outpouching on the left side, suspected to originate from an inferior frontal branch of the A2 segment of the ACA (*Figure 2*). The patient was graded as a Hunt Hess 5 with Fisher 4.

An emergent right frontal external ventricular drain was placed, which revealed an elevated intracranial pressure. A left frontotemporal craniotomy for hematoma evacuation and aneurysm clipping was performed. The hematoma was evacuated, and an aneurysm was seen. A temporary clip was placed across the proximal and distal ends of the aneurysm, followed by a 7-mm straight Yasargil aneurysm clip across the neck. Micro-Doppler assessment revealed no reliable flow within the aneurysm. The aneurysm was punctured using a 25-gauge needle on a syringe, confirming the absence of active bleeding and aneurysm deflation. ICG video angiography confirmed aneurysm obliteration. The bone flap placement was secured using the bioabsorbable plating system (*Figure 3*).

On postoperative day 0, CT angiography showed no evidence of a residual aneurysm. Despite clearance of the IVH, he required a ventriculoperitoneal shunt for persistent hydrocephalus (*Figures 3-5*). Transcranial Doppler was performed for 3 weeks without evidence of vasospasm, while nimodipine was administered for the same duration. An echocardiogram demonstrated a small patent foramen ovale, but no evidence of vegetation. Upon pedigree evaluation, there were some European roots on the paternal side and a maternal family history of cerebral aneurysms and cavernous malformation, which were related to an autosomal dominant connective tissue disorder with incomplete penetrance. A complete genetic panel was completed that included, but was not limited to, *ADAMTS17*, *FOXE3*, *CCM2*, *TGF*, and *COL1A1*, but was negative. In addition,

Highlight box

Key findings

- This 15-month-old boy presented with neurological extremis, and status epilepticus was found to have a ruptured large saccular intracranial aneurysm of the distal anterior cerebral artery with extensive intraparenchymal hematoma and intraventricular hemorrhage. The patient underwent emergent clot evacuation and microsurgical clipping.

What is known and what is new?

- Intracranial aneurysms in children and toddlers account for 5% of spontaneous intracerebral hemorrhages, and are most commonly false aneurysms or pseudoaneurysms secondary to trauma or infection. Most intracranial aneurysms are treated endovascularly, but the use of clipping, indocyanine green video angiography for assistance in technique, and timely care can provide optimal care for patients with a similar presentation.

What is the implication, and what should change now?

- Classic microsurgical clipping techniques are an important skill set for patients with intraparenchymal clot-associated aneurysms. The pathogenesis of intracranial aneurysms remains a great question as standard testing for connective tissue disorders and vascular malformations may be negative.

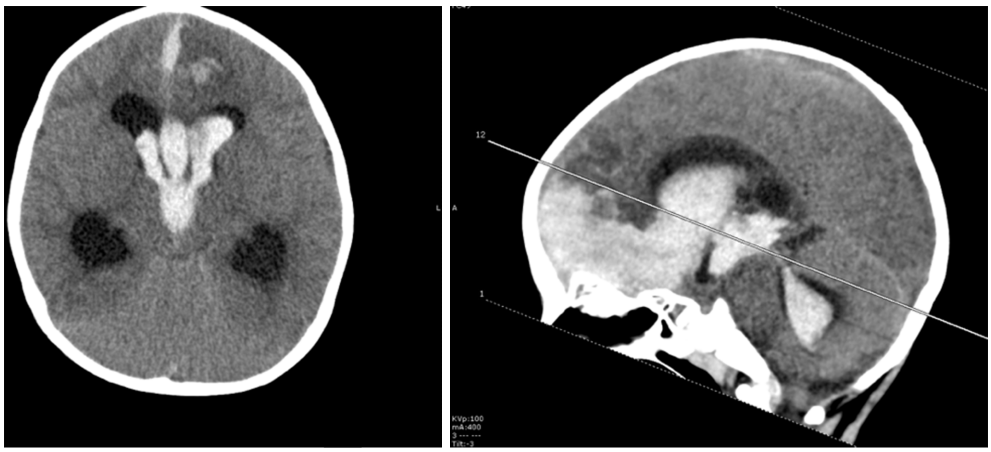

Figure 1 Computed tomography head non-contrasted axial (left) and sagittal (right) imaging demonstrates the large left frontal hematoma with diffused intraventricular hemorrhage and ventriculomegaly.

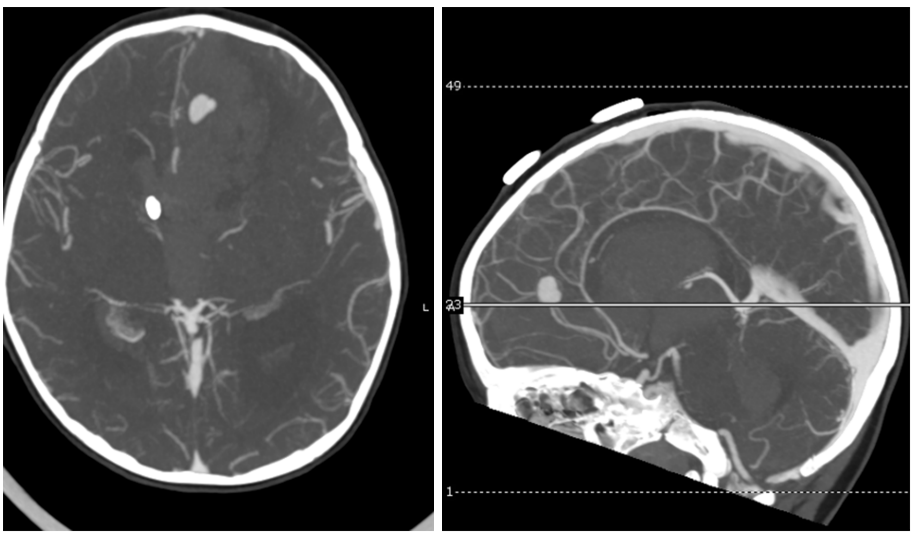

Figure 2 Computed tomography angiography of the head axial (left) and sagittal (right) demonstrated a saccular aneurysm in the inferior frontal region in close association with intraparenchymal hematoma.

there is no history of exposure to cigarette smoke (both parents are non-smokers) as this is a known risk factor for both presence and rupture of the aneurysms in adults (6,7). After 15 days, he could eat and breathe independently. At the 6-month and 12-month follow-ups, he demonstrated significant developmental progress, including appropriate speech, motor, and cognitive delay. He remains esotropic in the left eye and is kept on levetiracetam for seizure control.

Discussion

Key findings

This case of a left anterior internal frontal aneurysm in a 15-month-old boy presents a unique clinical scenario. The absence of a preceding trauma or infection, as well as an adverse family history of connective tissue disorders or genetic kidney diseases, further obscures the etiology.

Timely performance of CT angiography was critical for the early identification of this patient's aneurysm. The patient's lucent interval, with progression of focal neurological signs and subsequent posturing with seizures warranted non-contrasted CT head imaging with the addition of vessel imaging once the hemorrhage was identified.

Strengths and limitations

The patient's presentation with acute-onset emesis and status epilepticus, coupled with imaging findings of a sizeable IPH, diffuse IVH, and hydrocephalus, indicated a catastrophic event necessitating urgent intervention. Reports demonstrate endovascular management occurs in over 80% of ruptured or symptomatic cases, the decision to perform an emergent

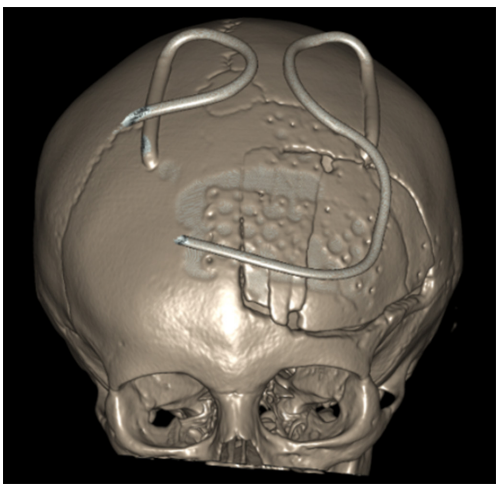

Figure 3 Postoperative computed tomography of 3-dimensional reconstruction with bilateral external ventricular drains with bioabsorbable plating system seen.

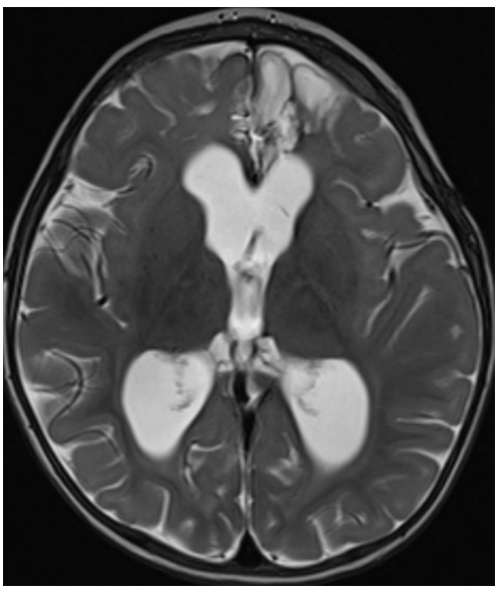

Figure 5 Magnetic resonance imaging T2 axial image showing encephalomalacia in the medial left frontal lobe with mild ventriculomegaly. The external ventricular drain was removed, and a permanent cerebrospinal fluid diversion was placed with a ventriculoperitoneal shunt.

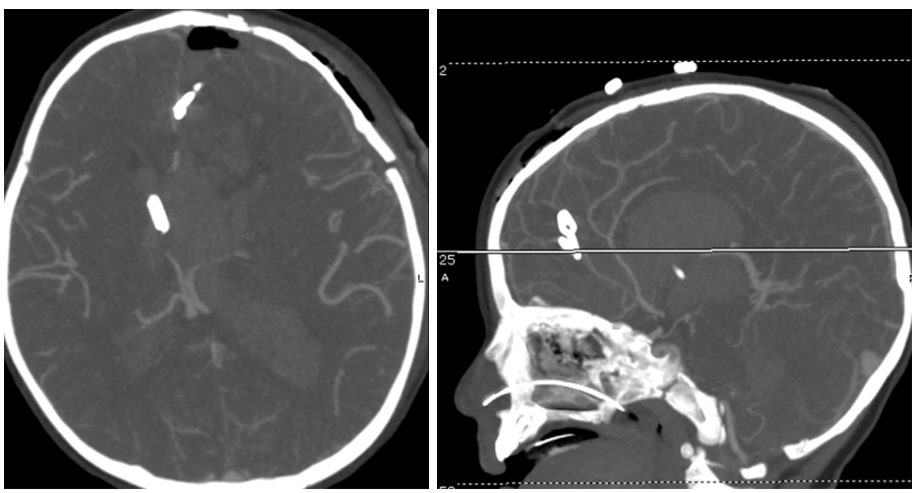

Figure 4 Postoperative computed tomography angiography of the head axial (left) and sagittal (right) with clips and no further aneurysm contrast opacification, suggestive of complete obliteration. An external ventricular drain is observed within the right frontal horn.

craniotomy and aneurysm clipping was critical in preventing further deterioration, as the intracerebral hematoma largely guided decision-making for clipping versus management with endovascular means (8-10). Although catheter angiography was not completed, ICG provided a viable alternative for intraoperative assessment. ICG use in pediatric cerebrovascular disease is not described in the literature but is well described for adult cerebrovascular intraoperative assessment, but described by some in other pediatric diseases (11). ICG critically assisted in our obliteration of the aneurysm. Completion of perioperative angiography to ensure complete aneurysm obliteration is a useful adjunct. In this case, it was deemed that cerebral angiography in the patient's age group would risk loss of limb from access related morbidity and was non-superior to postoperative non-invasive CT angiography, especially since direct visualization, micro-Doppler and needle puncture confirmed no flow into the aneurysm. Follow-up CT angiography or classical angiography will be heavily weighted in the future, given the patients' age and lifespan of the potential risk of recurrence (12). However, Xu *et al.* [2021] analyzed a cohort of pediatric aneurysms treated by microsurgical clipping and demonstrated no evidence of recurrence after a 5.5-year follow-up (13). This meticulous approach highlights the complexity of managing aneurysms in a pediatric population and provides an exciting approach for future cases. Additionally, absorbable plating systems typically used in craniofacial surgery in young patients allow for continued skull growth without the limitations of traditional titanium plating systems (14,15).

Comparison with similar published research

Among pediatric hemorrhagic stroke, a large population cohort study demonstrated that cerebral aneurysms are the underlying cause in approximately 13% of cases. The presentation of aneurysmal rupture with extensive IPH with interventricular hemorrhage is not typical; instead, it usually indicates a burden of subarachnoid hemorrhage (1). A distal ACA aneurysm in a two-month-old reported and describes the presence of IVH, and similarly IPH is more commonly presenting sign in adolescents with aneurysmal rupture (16). Unlike typical pediatric intracranial aneurysms found in the posterior circulation or major artery bifurcations, this aneurysm is in the distal ACA, either a branch of the anterior internal frontal region or orbitofrontal, making pseudoaneurysm most likely (12). However, this saccular aneurysm appeared to balloon from a parent vessel with a true neck. Additionally, there was no pertinent history or signs and symptoms of infection or trauma, which are common causes of pseudoaneurysms. The age of this patient makes this case unique as adolescent children are more likely to present with intracerebral hemorrhages for aneurysms than infants and toddlers.

Explanations of findings

The underlying family history of intracranial vascular conditions should be appreciated in this patient's history. Although genetic testing was completed and not positive, a complete genetic panel, as described, should always be completed (5,17,18). Common genetic syndromes of intracranial aneurysmal formation include autosomal dominant polycystic kidney disease with PKD1/2, connective tissue disorders such as Ehlers Danlos or Marfan. It has been described in non-syndromic patients with single nucleotide polymorphisms discovered through genome-wide studies. CKDN, SOX, TIMP-3, and ADAMTS15 have been associated with familial aneurysms. The multifactorial interplay of genetic variations makes the description of aneurysmal pathogenesis challenging, and this case adds to the importance of further genetic studies. Beyond immediate surgical success and postoperative recovery, it is crucial to consider the long-term neurodevelopmental outcomes and quality of life of pediatric patients undergoing aneurysm clipping (12). Given the critical period of brain development in children under 3 years, any surgical intervention in this age group carries the risk of impacting cognitive and motor functions. Research indicates that early life brain injuries can lead to long-term deficits in neurodevelopment and behavior. However, positive cases have been reported like this one in functional improvement following severe neurological devastation like this case (19,20). A case series of pediatric patients <18 years old with ruptured aneurysms describe a favorable modified Rankin Scale score in rupture intracranial aneurysm patients (21). Long-term follow-up with neurodevelopmental assessments is necessary to monitor and address potential deficits. After 1 year of follow-up, the patient is improving neurologically, with mild cognitive impairment and epilepsy requiring anti-epileptic medications. We believe that the factors contributing to a successful neurological outcome included timely treatment of his hydrocephalus, control of elevated intracranial pressures with hematoma evacuation, expedient occlusion of the aneurysm, and seizure control. The intraparenchymal hematoma spared the non-eloquent

cortex and the vascular territory of the diseased artery, typically sparing the premotor, supplemental motor area, and motor cortex, which were important factors in this patient's outcome.

Implications and actions needed

This case highlights the importance of timely neurosurgical evaluation and thorough microsurgical applications. Additionally, the understanding of aneurysmal pathogenesis is critical, as there is no clear etiology for this aneurysm, despite comprehensive genetic testing. A large case series of ruptured aneurysms in toddlers is lacking. A large multi-institutional study aimed to longitudinally evaluate this age group would help identify the degree of outcome in this age group.

Patient perspective from the patient's mother

This was a lifechanging event for our family, and this was one of the scariest moments of our life and weren't sure if we were going to lose our son. The compassionate care provided by the surgical team and the intensive care teams gave us incredible support. He has been improving every step of the way, still obtaining some degree of physical therapy for his rehabilitation. He is improving each and every day.

Conclusions

This case of a 15-month-old boy with a left distal ACA aneurysm underscores the rarity and complexity of pediatric aneurysms, particularly in very young patients. The successful surgical management, despite the challenges posed by the lack of intraoperative angiography, highlights the importance of prompt recognition and intervention in pediatric aneurysmal rupture. Long-term follow-up focusing on neurodevelopmental outcomes is crucial to ensure optimal recovery and quality of life for pediatric patients' post-surgery. The lack of apparent risk factors, including an underlying hereditary cause, and the rare location of this aneurysm underscores the need for further research into infantile pediatric aneurysms and management mechanisms.

Acknowledgments

Funding: None.

Footnote

Reporting Checklist: The authors have completed the CARE reporting checklist. Available at https://tp.amegroups.com/article/view/10.21037/tp-24-289/rc

Peer Review File: Available at https://tp.amegroups.com/article/view/10.21037/tp-24-289/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-24-289/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this study were in accordance with the ethical standards of the institutional and/or national research committee(s) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patient's mother for publication of this case report and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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Cite this article as: Koutsouras GW, Mensah R, Krishnamurthy S. Clipping of a ruptured cerebral aneurysm in a toddler: a case report and review of aneurysmal treatment in children. Transl Pediatr 2024;13(10):1885-1891. doi: 10.21037/tp-24-289