Check for updates

Giant Aneurysm Arising from Anomalous Branch of the Middle Cerebral Artery in a Pediatric Patient: Case Report and Review of the Literature

Kai Rui Wan¹, Ramez W. Kirollos¹⁻³, Hwei Yee Lee⁴, David C.Y. Low¹⁻³, Lee Ping Ng¹, Wan Tew Seow¹⁻³, Sharon Y.Y. Low¹⁻³

Key words

- Intracranial aneurysm
- Middle cerebral artery

Abbreviations and Acronyms

IA: Intracranial aneurysm ICA: Internal carotid artery MCA: Middle cerebral artery MRA: Magnetic resonance angiography

From the ¹Neurosurgical Service, KK Women's and Children's Hospital, ²Department of Neurosurgery, National Neuroscience Institute, ³SingHealth Duke-NUS Neuroscience Academic Clinical Program, and ⁴Department of Pathology, Tan Tock Seng Hospital, Singapore

To whom correspondence should be addressed: Sharon Y. Y. Low, M.B.B.S.

[E-mail: sharon.low.y.y@singhealth.com.sg]

Citation: World Neurosurg. (2019) 128:165-168. https://doi.org/10.1016/j.wneu.2019.05.012

Journal homepage: www.journals.elsevier.com/worldneurosurgery

Available online: www.sciencedirect.com

1878-8750/\$ - see front matter \circledast 2019 Elsevier Inc. All rights reserved.

INTRODUCTION

Pediatric intracranial aneurysms (IAs) are rare occurrences with a reported prevalence of 0.5%-4.6%.1-3 They have a myriad of presentations such as spontaneous subarachnoid hemorrhage,⁴ headaches. seizures, symptoms because of local mass effect, or in some instances, as incidental findings on imaging.⁵⁻⁷ In children, middle cerebral artery (MCA) anomalies have been observed to be associated with the development of IAs.^{8,9} Pediatric IAs also trend toward a higher incidence of giant aneurysms.^{10,11} Spontaneous thrombosis has been reported in 8%-17% of this subset of aneurysms; however, subsequent spontaneous recanalization has also been known to occur.¹²

We report a pediatric patient who presented with seizures secondary to a giant partially thrombosed right MCA aneurysm. Interestingly, this unruptured aneurysm was found to originate from an anomalous M2 BACKGROUND: Pediatric intracranial aneurysms are extremely rare. In this age group, cerebral vascular anomalies have been associated with the development of intracranial aneurysms.

CASE DESCRIPTION: We present a case of a previously well 11-year-old boy who presented with seizures secondary to a giant, unruptured, and partially thrombosed right middle cerebral artery (MCA) aneurysm. Extensive workup for underlying infective and autoimmune etiology was negative. Of interest, this vascular lesion was found to originate from an anomalous M2 branch, which ran an aberrant parallel course within the Sylvian fissure to the main and distally bifurcating MCA. The patient underwent successful surgical clipping and excision of the giant aneurysm.

CONCLUSIONS: Because of the infrequency of the diagnosis, clinical presentation, and its unique neurovascular anatomy, the management of this case is discussed in corroboration with current literature. In addition, highlighting this unusual case in an individual adds to the growing body of literature for better disease understanding, especially in the pediatric population.

branch, which had an aberrant parallel course within the Sylvian fissure to the main and distally bifurcating MCA. Because of the infrequency of the disease and its unique vascular anatomy, this case is discussed in corroboration with the current literature.

CASE REPORT

A previously well 11-year-old boy was admitted for seizures and a 5-day history of headache prior to his presentation. His initial Glasgow Coma Scale score was 8 out of 15 (E1 V2 M5), but was otherwise hemodynamically stable. He had no recent trauma, infective events, or other significant medical history. Computed tomography scan of his brain demonstrated a lobulated lesion (2.5 \times 2.5 \times 2.5 cm) within the right sylvian fissure. There was no evidence of infarct, hemorrhage, midline shift, or hydrocephalus (Figure 1). His blood cultures, inflammatory markers, and transthoracic echocardiography of the heart were unremarkable. A diagnostic cerebral angiography demonstrated a large fusiform aneurysm $(1.7 \times 3.1 \times 2.5 \text{ cm})$ with no discernible neck arising from the right MCA's M₂ branch of the inferior division (Figure 2).

The patient subsequently underwent clipping of the aneurysm via a right pterional approach. Intraoperatively, we observed a dilated branch arising from the M₂ and coursing distally and parallel to the main MCA trunk within the Sylvian cistern, forming a smooth fusiform aneurysm that was embedded into the insula and medial temporal lobe. There were no branches or perforators arising from the proximal segment. As expected, intraaneurysmal partial thrombosis was evident. Temporary aneurysm clips were placed on the feeding and draining arteries. The aneurysm sac was opened, and it was internally decompressed prior to en bloc excision of the aneurysm sac wall (Figure 3). This maneuver allowed circumferential dissection to avoid injury to vascular branches and perforators arising from or adjacent to the aneurysm. After the excision of the aneurysm, the KAI RUI WAN ET AL.



Figure 1. Representative nonenhanced computed tomography brain scan images of the patient at first presentation in axial (**A**) and coronal (**B**) views. A well-defined round $2.5 \times 2.5 \times 2.5 \times 2.5$ cm lesion is seen in the right temporal region and appears to be located in the right Sylvian fissure. The lesion has a hyperdense anteromedial component with the posterolateral component isodense to the brain. Of note, no evidence of subarachnoid hemorrhage is seen.

distal outflow which provided some degree of temporal cortical blood supply was confirmed using indocyanine green administration.

Postoperatively, his clinical course was uneventful, and he was discharged on postoperative day 4 with no discernible neurologic deficits. Histologic examination of the sac wall demonstrated inflammation, necrosis, and an organizing thrombus (Figure 4). No organisms were cultured from the sac wall or the aneurysm thrombus. The patient remains well on outpatient follow-up.

DISCUSSION

Giant IAs are rare in the pediatric population.⁶ If present, they are associated with high risks of mortality and morbidity, given the increased propensity to rupture.^{6,13} Broadly speaking, most (45%) pediatric IAs are the result of idiopathic causes.^{12,14} Trauma is the next most



Figure 2. Representative images from the patient's diagnostic cerebral angiogram showing the anteroposterior view of the right internal carotid artery (ICA) run (**A**) and lateral view of the right ICA run (**B**). There is a large superior-laterally directly aneurysm ($1.7 \times 3.1 \times 2.5$ cm) with no discernible neck seen arising from the right middle cerebral artery (MCA) M2 inferior division. Delayed washout of contrast in the aneurysm and filling of the MCA M2 inferior division, distal to the aneurysm is noted. There is no active contrast extravasation or other vascular anomaly detected. *Red arrows* point to where we presumed is the origin of the aneurysm.

common cause,^{7,11} often relating to sites where a major conducting artery crosses a dural fold or sites where a vessel is fixated between 2 points near the skull base. Following that, mycotic aneurysms account for approximately 5% of pediatric IAs. Children at risk are usually immunocompromised and/or have congenital cardiac malformations complicated by endocarditis.¹⁴ In addition, they may have a genetic abnormality that cause developmental defects in the arterial tissue.^{2,7} Examples include Marfan syndrome, Ehlers-Danlos syndrome, and autosomal dominant polycystic kidney disease.15 Other less frequent associations of pediatric IAs include cerebral arteriovenous malformation, tumors, radiation injury, or previous treatment with intrathecal colloidal gold.5,7

Typically, most saccular IAs are located in the anterior circulation and at the internal carotid artery (ICA) bifurcation, followed by the MCA bifurcation, or at the segment distal to the first major bifurcation.^{4,11,12,15} However, for fusiform aneurysms, the MCA is often the most common site for their origin.^{16,17} In the absence of atherosclerosis, predisposing factors for the latter include conditions that cause structural anomalies of the intracranial vessels. These can be either developmental, as seen in our patient, or acquired.^{8,16,18,19}

Comparatively, MCA anomalies are less commonly reported than other major intracranial arteries.²⁰ If present, these are often associated with the development of IAs,^{8,9} observed in our as patient. Embryologically, the MCA is formed by a process of fusion and regression of multiple plexiform arterial twigs, which are early collaterals to the cranial branch of the ICA, which eventually continues as the anterior cerebral artery.²⁰ Anomalous MCA vessels can be fenestrated, accessory, or duplicate vessels.^{8,21} In contrast, our patient's fusiform aneurysm was uniquely located on a dilated anomalous M2 branch. which was initially thought to be a temporal branch of the M1. Based on preoperative imaging, we were under the impression that it possibly contributed to the vascular supply of the inferior temporal lobe. However, intraoperatively, this anomalous M2 branch was noted to run an aberrant parallel course within the Sylvian fissure to the main and distal bifurcating MCA trunk.



Figure 3. (A) View via a right pterional craniotomy to show delineation of the right internal carotid artery—middle cerebral artery anatomy in relation to the optic nerve (cranial nerve II). (B) Position of the aneurysm sac in relation to the inferior M2 and its anomalous M2 branch. (C) Temporary aneurysm clips at both ends of the anomalous M2 branch with decompressed aneurysm sac that was adjacent to the temporal lobe. CN, cranial nerve; ICA, internal carotid artery; MCA, middle cerebral artery.

By far, most of the reported aneurysms associated with anomalous branches tend to occur where the latter originate from the main vessel.²² The hypothesis in our case is that the aneurysm's parent vessel is a developmentally anomalous branch given its size, despite its relatively limited territory of blood supply; and that the main MCA trunk was patent. The configuration of the fusiform aneurysm without any branches identified at its base is presumably related to hemodynamic factors, or alternatively, a histologically unidentified structural abnormality of the vessel wall. From a surgical perspective, we were fortunate with the location of this aneurysm. The patient's fusiform aneurysm was positioned at the right inferior branch of an accessory M2 segment, which appeared to supply a small region in the inferior temporal lobe. Putting it all together, the patient's aneurysm location provided favorable vascular anatomy for its complete surgical excision. This enabled the following aims to be met: 1) reduction of mass effect, 2) elimination of the risk of future rupture, and 3) total excision and sacrifice of the vessel without postoperative ischemic sequelae.

In present times, the discussion of microvascular clipping is incomplete without the mention of endovascular options. The ultimate goal of neurovascular intervention is to reduce the risk of aneurysmal hemorrhage, reduce mass effect, and preserve cerebral vasculature.6,13 Microvascular clipping and endovascular treatment techniques both need to be assessed in relation to the patient's vascular anatomy, the child's condition, and the experience of the multidisciplinary team to determine an optimal management strategy.4,12 Microsurgical options include that of clip reconstruction, trapping, or bypass with proximal occlusion. Surgical obliteration of the aneurysm sac has been associated with a lower recurrence rate and subsequent de novo aneurysmogenesis. However, giant IAs can present technical challenges with open surgical treatment, and may require clip reconstruction, wrapping, trapping, or excision with or without bypass.⁴ In contrast, endovascular treatments for similar cases include options of primary coiling with or without stent, proximal occlusion or trapping, liquid embolic agents such as ONYX-HD 500 (Micro Therapeutics, Inc., Irvine, California, USA), or a



Figure 4. (A) Low power view of the aneurysm wall. There is an intraluminal laminated thrombus. No well-formed granulomas are seen (hematoxylin and eosin stain; magnification, $\times 20$). (B) Slide depicting necrosis and inflammation in a separate region of the aneurysm wall (hematoxylin and eosin stain; magnification, $\times 20$).

flow diversion device. However, the necessity of long-term antiplatelet therapy for a flow diversion device, and its Food and Drug Administration—approved use for ICA aneurysms only, limits its utility in patients with ruptured aneurysms.⁶ Given the size of the aneurysm in our patient's case, the choice of endovascular treatment will still pose significant mass effect on the adjacent brain parenchyma.

As reflected in our case, pediatric IAs often result in good outcomes,^{7,15} if they are managed mindfully. Nonetheless, this cohort of patients remains at risk of developing de novo or enlarging aneurysms and therefore require long-term surveillance.7,23 In the pediatric cohort, magnetic resonance angiography (MRA) is usually the preferred choice for surveillance imaging to reduce exposure to excess ionizing radiation. However, MRA has notable limitations. These include decreased sensitivity in visualizing aneurysms with slow flow, difficulty in envisaging aneurysms <3 mm, and a longer image acquisition time which may necessitate the use of anesthesia,¹⁵ especially in young children. Until the time comes for a more advanced and logistically convenient modality, MRA is still a feasible choice for such patients in the meantime.

CONCLUSIONS

Pediatric IAs are rare and represent an independent pathophysiologic entity from their adult counterparts.⁷ They are a heterogenous group in terms of etiology, clinical manifestations, aneurysm characteristics, and patient outcomes. Concurrently, the long life expectancy of children coupled with the complexity of these aneurysms present unique challenges to the treating doctors. We advocate in-depth molecular studies, multidisciplinary efforts, and international collaborations to best manage this disease for affected patients.

REFERENCES

- I. 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 [discussion: 32-33].
- Gemmete JJ, Toma AK, Davagnanam I, Robertson F, Brew S. Pediatric cerebral aneurysms. Neuroimaging Clin N Am. 2013;23:771-779.
- Pruvot AS, Curey S, Derrey S, Castel H, Proust F. Giant intracranial aneurysms in the paediatric population: suggested management and a review of the literature. Neurochirurgie. 2016;62:20-24.
- 4. Jian Z, Yang Z, Smerin D, Xiong X. Intracranial giant aneurysms in children and adolescents misdiagnosed as intracranial tumors before operation: 2 cases report. Int J Clin Exp Med. 2018;11:6268-6275.
- Ghali MGZ, Srinivasan VM, Cherian J, et al. Pediatric intracranial aneurysms: considerations and recommendations for follow-up imaging. World Neurosurg. 2018;109:418-431.
- Sekhar LN, Tariq F, Mai JC, et al. Unyielding progress: treatment paradigms for giant aneurysms. Clin Neurosurg. 2012;59:6-21.
- Sorteberg A, Dahlberg D. Intracranial nontraumatic aneurysms in children and adolescents. Curr Pediatr Rev. 2013;9:343-352.
- LaBorde DV, Mason AM, Riley J, Dion JE, Barrow DL. Aneurysm of a duplicate middle cerebral artery. World Neurosurg. 2012;77:201.e1-201.e4.
- Menshawi K, Mohr JP, Gutierrez J. A functional perspective on the embryology and anatomy of the cerebral blood supply. J Stroke. 2015;17:144-158.
- Requejo F, Ceciliano A, Cardenas R, Villasante F, Jaimovich R, Zuccaro G. Cerebral aneurysms in children: are we talking about a single pathological entity? Childs Nerv Syst. 2010;26:1329-1335.

- II. Deora H, Rao K, Somanna S, Srinivas D, Shukla DP, Bhat DI. Surgically managed pediatric intracranial aneurysms: how different are they from adult intracranial aneurysms? Pediatr Neurosurg. 2017;52:313-317.
- Goncalves VM, Cristino N, Cunha ESM. Spontaneous thrombosis in giant aneurysm of the anterior communicating artery complex in pediatric age: fiveyear follow-up. Case Rep Vasc Med. 2014;2014:236041.
- Almefty K, Spetzler RF. Management of giant internal carotid artery aneurysms. World Neurosurg. 2014;82:40-42.
- Aeron G, Abruzzo TA, Jones BV. Clinical and imaging features of intracranial arterial aneurysms in the pediatric population. Radiographics. 2012;32:667-681.
- Pinto FC, Valiengo L, Santos PP, Matushita H, Plese JP. Intracranial arterial aneurysms in childhood: case report. Arq Neuropsiquiatr. 2006;64:676-680.
- Day AL, Gaposchkin CG, Yu CJ, Rivet DJ, Dacey RG Jr. Spontaneous fusiform middle cerebral artery aneurysms: characteristics and a proposed mechanism of formation. J Neurosurg. 2003;99:228-240.
- Park SH, Yim MB, Lee CY, Kim E, Son EI. Intracranial fusiform aneurysms: it's pathogenesis, clinical characteristics and managements. J Korean Neurosurg Soc. 2008;44:116-123.
- Kong DS, Kim MH, Park WY, et al. The progression of gliomas is associated with cancer stem cell phenotype. Oncol Rep. 2008;19:639-643.
- Ceylan S, Karakus A, Duru S, Baykal S, Ilbay K. Reconstruction of the middle cerebral artery after excision of a giant fusiform aneurysm. *Neurosurg Rev.* 1998;21:189-193.
- 20. Uchiyama N. Anomalies of the middle cerebral artery. Neurol Med Chir (Tokyo). 2017;57:261-266.
- 21. Yamamoto H, Marubayashi T, Soejima T, Matsuoka S, Matsukado Y, Ushio Y. Accessory middle cerebral artery and duplication of middle cerebral artery-terminology, incidence, vascular etiology, and developmental significance. Neurol Med Chir (Tokyo). 1992;32:262-267.
- 22. Ren H, Ma L, Wei M, Li J, Yu M, Yin L. Duplicated middle cerebral artery origin with an aneurysm. *Medicine (Baltimore).* 2018;97:e9947.
- Koroknay-Pal P, Laakso A, Lehto H, et al. Longterm excess mortality in pediatric patients with cerebral aneurysms. Stroke. 2012;43:2091-2096.

Conflict of interest statement: The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Received 7 April 2019; accepted 2 May 2019

Citation: World Neurosurg. (2019) 128:165-168. https://doi.org/10.1016/j.wneu.2019.05.012

Journal homepage: www.journals.elsevier.com/worldneurosurgery

Available online: www.sciencedirect.com

1878-8750/\$ - see front matter © 2019 Elsevier Inc. All rights reserved.