



De novo distal middle cerebral artery aneurysm post-excision of intracerebral arteriovenous malformation in an 8-year old

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Abstract

Cerebral arteriovenous malformations (AVMs) are frequently associated with concurrent aneurysms. These aneurysms are commonly haemodynamically related to the AVM and can be classified into extranidal or intranidal in reference to the AVM nidus. An aneurysm arising from an artery that does not angiographically contribute to the blood flow to the AVM is uncommon. A distal middle cerebral artery (dMCA) aneurysm itself is also a rare presentation, especially in paediatric population. We present a rare case of dMCA aneurysm that was noted after successful surgical management of a ruptured AVM in an 8-year-old child and its management.

Background

About 10–30% of patients with cerebral arteriovenous malformation (AVM) have an associated artery aneurysm. The majority of these aneurysms are flow-related to the malformation. These aneurysms can be classified into extranidal or intranidal in reference to the AVM nidus Rammos et al *Am J Neuroradiol* 37:1966–1971, [1]. An aneurysm arising from a different artery that does not angiographically contribute to the blood flow associated with the AVM is less common and would generally be regarded as unrelated to the AVM. Distal cerebral artery aneurysm itself is also a rare presentation, comprising of 1–7% of all middle cerebral artery aneurysm. In children, mycotic infection and dissection are the two most common causes for aneurysm in this location. Unlike in adults, berry aneurysms are uncommon in children. We describe a young patient who was found to have distal middle cerebral artery (dMCA) aneurysm in follow-up DSA (Digital Subtraction Angiogram) after the initial successful surgical treatment for a cerebral frontal AVM. In this particular case, endovascular repair is thought to be the best strategy to treat the aneurysm. However, there still remains a lack of consensus of the best management strategy (surgery or endovascular) in treating flow-related aneurysms in general. This is usually based on an individual case scenario and the treatment is tailored depending on various factors including the expertise of the treating team.

Keywords Cerebral arteriovenous malformation · Distal middle cerebral artery aneurysm

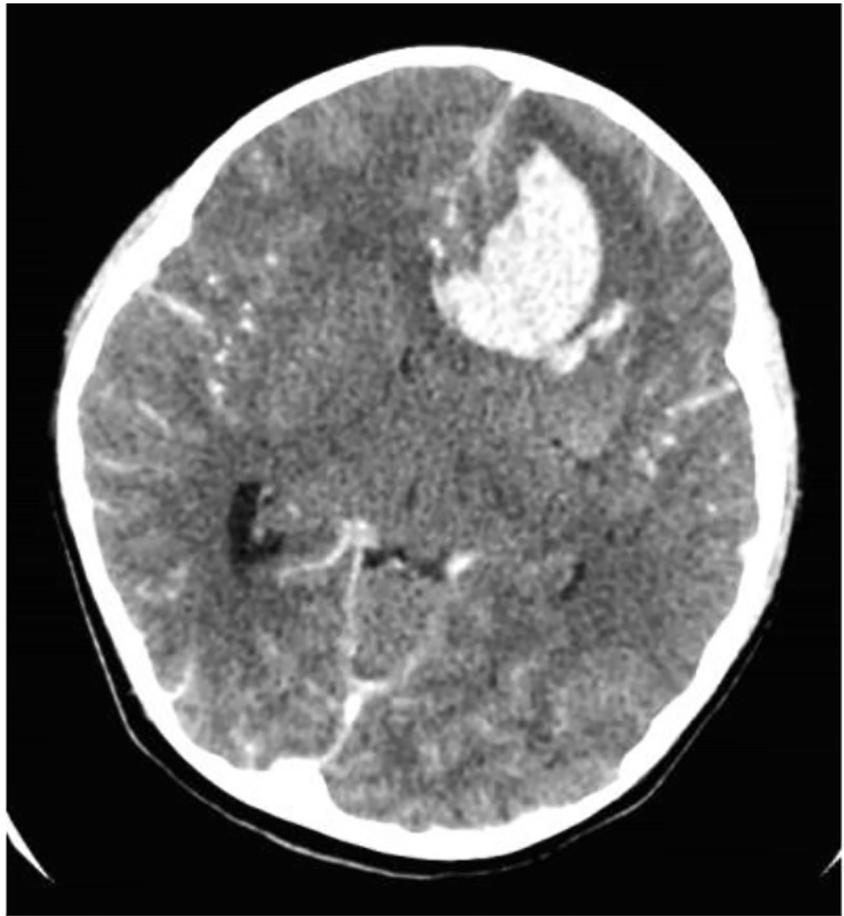
Case presentation

An 8-year-old girl with background of normal antenatal and postnatal history presented to the local hospital with 2-week history of worsening headache and vomiting. On

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Fig. 1 CT head on admission revealed acute intraparenchymal haemorrhage in the left frontal lobe with mass effect



examination, her Glasgow Coma Scale (GCS) was 7/15 and she had right sided weakness of 1/5 in her upper and lower limbs. Initial CT head revealed a left frontal large intracranial haemorrhage with 1-cm midline shift (Fig. 1). She underwent emergency craniotomy and evacuation of the haematoma. CT head post-evacuation showed residual left frontal haematoma but decreased mass effect (Fig. 2). She was transferred to the quaternary centre for further management. An early magnetic resonance imaging (MRI) of head and computed tomography angiography (CTA) showed a small medial frontal AVM, supporting the fact that the initial haematoma was a result of the ruptured AVM (Fig. 3). This was subsequently confirmed by a digital subtraction angiography (DSA) that revealed a residual left paramedian frontal pole micro AVM, lying at the medial margin of

the haematoma cavity. There was an ill-defined diffuse nidus measuring approximately 1 cm lying just to the left of the midline adjacent to the anterior cerebral artery (ACA) bifurcation into the pericallosal and callosomarginal arteries. In addition, numerous short perforating type tidal feeders from the ACA were noted and did not look suitable for endovascular treatment. The deep vein runs back along the ependymal surface to drain into internal cerebral veins and onto the vein of Galen and two small superficial draining veins running antero-superiorly to the superior sagittal sinus were demonstrated. Her case was discussed at the neurovascular multidisciplinary meeting. The decision was to treat the AVM by surgical excision based on the grade of the AVM and features that do not favour endovascular treatment. Stereotactic surgery is not a

Fig. 2 CT head post-evacuation showed residual left frontal haematoma but decreased mass effect and no evidence of new acute bleed



good option in this case because of the delay for AVM obliteration and reluctance to expose radiation to the developing nervous system. She subsequently underwent a neuro-navigation-guided complete excision of the residual AVM.

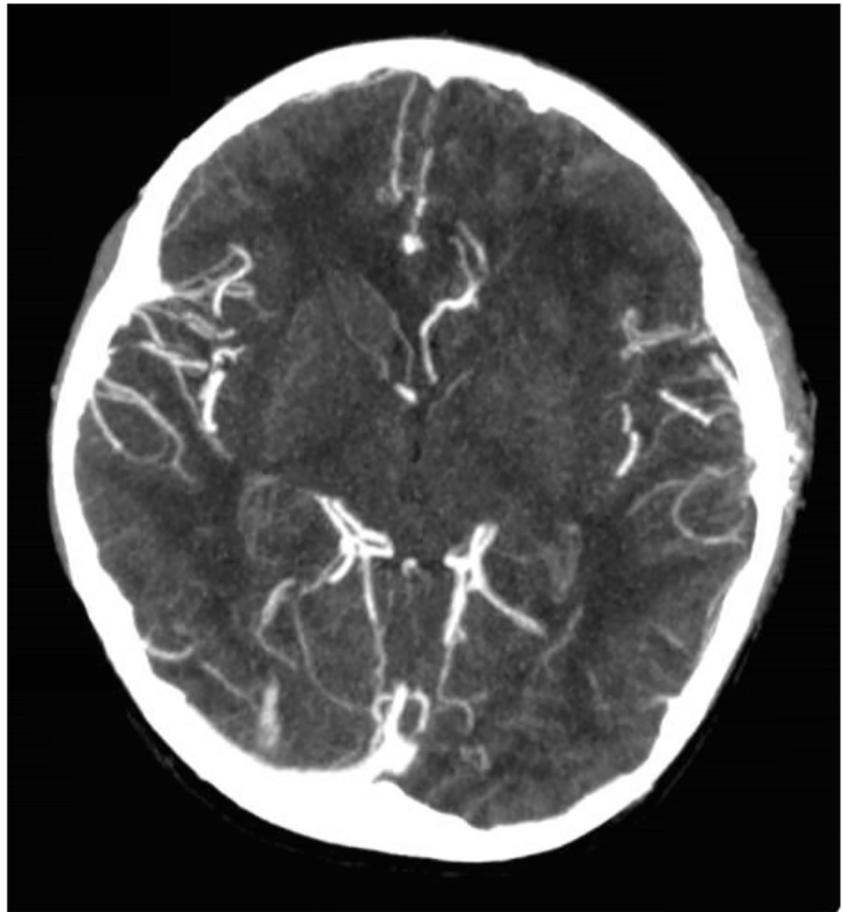
Post-operatively, she made good clinical progress and was discharged home with no neurological deficits. Post-operative cerebral angiogram showed that the previously resected AVM was no longer evident (Fig. 4). She had further follow-up cerebral angiogram in 5 months that revealed resolution of the AVM and a new 5-mm dissecting fusiform aneurysm arising from a distal frontal branch of the left middle cerebral artery was noted (Fig. 5a, b). The new dissecting aneurysm was successfully embolised by the radioneurologist in the same setting. Follow-up MRA in 3 months later showed no new aneurysm and the embolised aneurysm showed no intranidal flow (Fig. 6). She remains well with no residual disability.

Investigations

Discussion

A proportion of patients with AVM have concurrent intracranial aneurysm. This is postulated to be related to haemodynamic factors based on the observation that most aneurysm are located on proximal arteries haemodynamically connected to the AVM nidus [1]. However, in our case, the aneurysm was identified from an artery without direct blood flow to the AVM. This could be due to an iatrogenic cause secondary to the initial surgery. The dissecting fusiform formation of the aneurysm supports the iatrogenic cause. Other causes that might be contributing to the aneurysm formation include genetic predisposition or other host-specific factors (Table 1). Although most studies suggest the presence of aneurysm increased risk of haemorrhage in cerebral

Fig. 3 CT angiography revealed a medial left frontal AVM



AVM, it remains unclear whether presence of a coexisting flow unrelated aneurysm increases risk of haemorrhage from ruptured brain AVM. While several factors that could potentially contribute to increased risk of bleeding, a small AVM size (<3 cm) and deep venous drainage were found in this patient albeit the evidence from the literature is inconsistent and inconclusive.

There is paucity in the literature on dMCA aneurysm with this clinical background and regarding its management. A retrospective study of patients who underwent surgical repair of ruptured intracranial aneurysm found only 9 patients with ruptured distal MCA aneurysm [2]. Nan Lv et al. reported 8 cases of dMCA aneurysm in a 10-year retrospective study, with an incidence rate of 4.1% [3]. A 5-year retrospective study identified 35 distal artery aneurysm in paediatric patients (<18 years old) [4]. Aneurysms are rare in children and when they

do occur, they are likely dissecting. Most MCA aneurysms in adults arise from the main bifurcation (M1-M2 junction) due to haemodynamic stress factors [1]. On the other hand, dMCA aneurysm is often caused by other aetiologies, including infectious emboli or vasculitis. dMCA aneurysms can be classified into four segments—M2 (insular segment), M2-M3 junction, M3 (opercular segment) and M4 (cortical segment) [5].

Improvement in modern diagnostic techniques has increased the identification of distal aneurysm. It is debatable whether an asymptomatic small distal aneurysm requires active surgical treatment as these aneurysms may potentially remain stable and the risk of rupture is uncertain [6].

It is also arguable whether microsurgical clipping or endovascular treatment is more effective in treating dMCA aneurysm. A case report presented a case successfully treated by microsurgical clipping [7]. Another

Fig. 4 Post-operative cerebral angiogram showed complete excision of the AVM

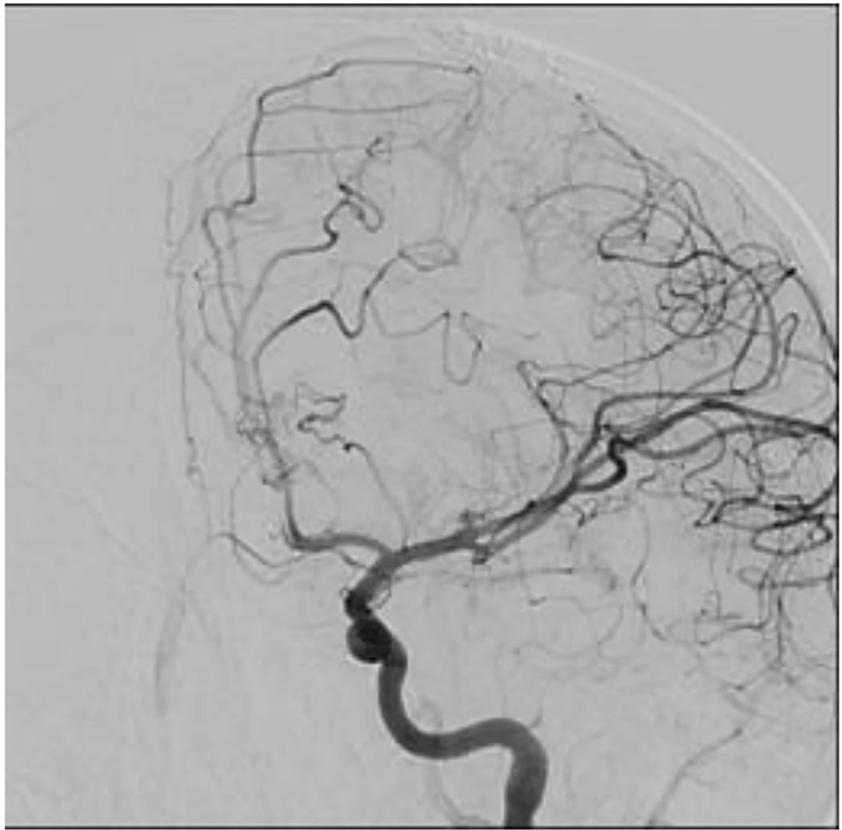
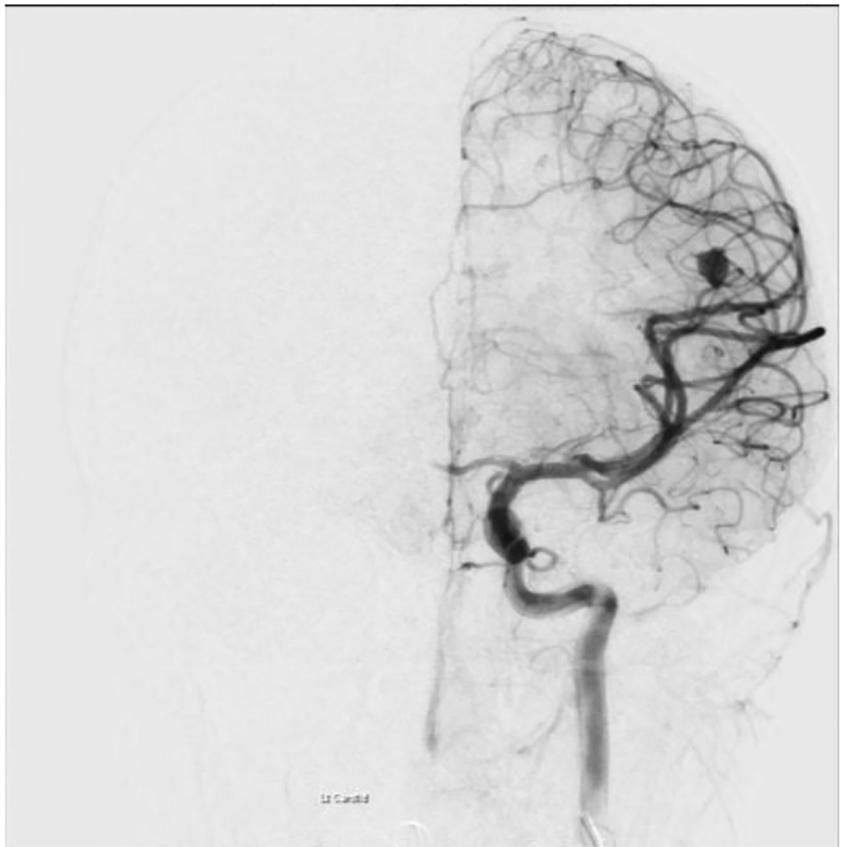


Fig. 5 Follow-up cerebral angiogram revealed a new 5-mm dissecting fusiform aneurysm arising from a distal frontal branch of the left middle cerebral artery



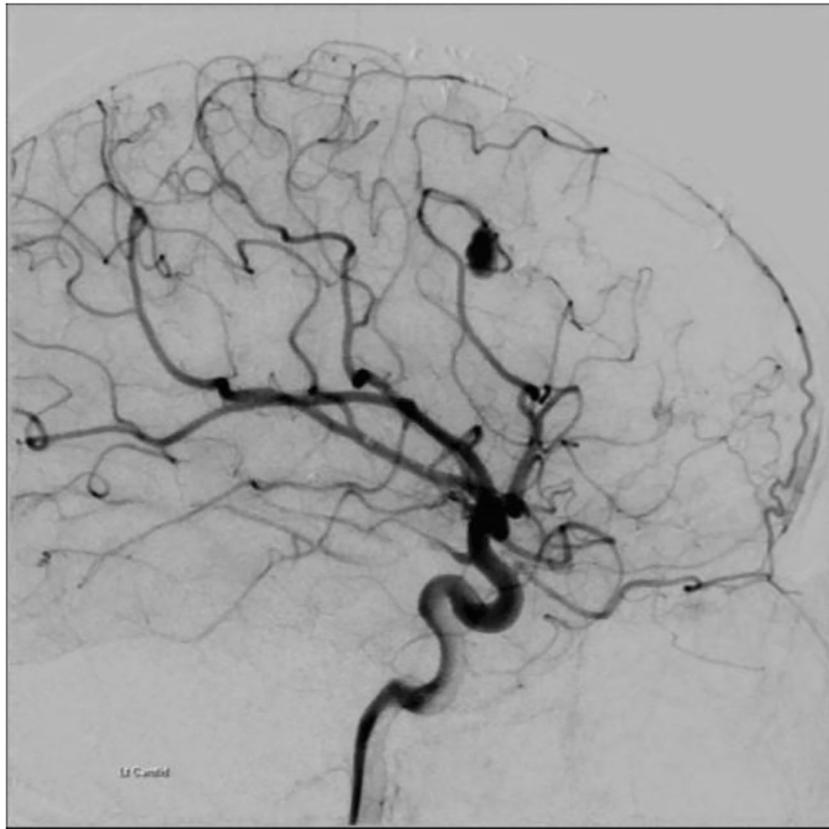


Fig. 5 continued.

case series suggested endovascular treatment is a safe and effective method for distal middle cerebral artery aneurysms [3]. The distal location of dMCA aneurysm may pose significant surgical challenge especially when the aneurysm is small and neuro-navigation may be needed as an adjunct. It is also technically difficult to perform aneurysm clipping and evacuation of haematoma at the same time in small aneurysm [3]. The disadvantage of endovascular repair is higher risk of recurrence despite smaller procedural risk.

Lastly, this case highlights the importance of long-term follow-up imaging in patients with treated cerebral AVM to ensure early identification of new aneurysm or recurrence of AVM. Literature review showed a wide range of recurrence interval from 3 months to 16 years after initial successful treatment of AVM [8]. To date, there are no universal accepted guidelines on long-term follow-up of these patients. In paediatric population, follow-up is important as there is a higher recurrence

rate compared with adults [8]. Cost effectiveness, patient safety and likelihood of recurrence should be taken into consideration.

Conclusion

Cerebral arteriovenous malformations (AVM) are known to be associated with aneurysms. A de novo distal middle cerebral artery aneurysm following successful treatment of intracranial AVM is a clinical rarity. Endovascular management remains the treatment of choice and these patients need to be treated in a quaternary care centre needing multi-disciplinary input. These patients do need a long term radiological follow up.

Fig. 6 MRA showed no new vascular abnormality



Learning points

- Early intervention of ruptured cerebral AVM determines the prognosis.
- Presences of concurrent aneurysm in cerebral AVM pose an increased risk of haemorrhage.
- Treatment of distal cerebral aneurysm poses technical difficulties.
- Importance of long-term follow-up imaging in treated cerebral AVM.

Table 1 Possible aetiopathogenesis of distal MCA aneurysm

Possible aetiopathogenesis of distal MCA aneurysm
Idiopathic
Genetic predisposition
Iatrogenic—secondary to the initial surgery or subsequent surgery (retraction induced damage)
Altered flow dynamics post-surgical resection of AVM
Pseudo-aneurysm

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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