Reminder of important clinical lesson

Recurrence of a paediatric arteriovenous malformation 9 years postcomplete excision: case report and review of literature

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Summary

Cerebral arteriovenous malformations (AVMs) are a common congenital vascular anomaly, which often present in both children and adults. Surgery is considered curative once postoperative angiography confirms the absence of vessels. We describe a 6-year-old girl, who had a Spetzler-Martin Grade II AVM resected successfully, in which a recurrent AVM was detected on routine follow-up over 9 years post excision. The aetiopathogenesis of this rare occurrence with a review of literature is discussed. Long-term postoperative follow-up in the form of MRI/MR angiogram is recommended for all fully resected AVMs in the paediatric age group, anticipating the possibility of future recurrence.

BACKGROUND

Cerebral arteriovenous malformations (AVMs) are sporadic congenital developmental vascular lesions, and are the most common congenital vascular anomaly to cause parenchymal haemorrhage in the paediatric population.^{1 2 3} AVMs occur due to a congenital failure during capillary and venous development, between the fourth and eighth week of gestation,⁴ when 'feeding' vessels may develop. During both the antenatal and postnatal period, 'functional' and 'satellite' vessels may contribute to subsequent growth.⁴ Postexcisional cerebral angiography is the gold standard to confirm the absence of residual nidus and early draining veins, which correlates with permanent total excision of the lesion, and elimination of the risk of haemorrhage.¹ However, the incidence of recurrent paediatric AVMs demonstrates that complete excision does not correlate with cure. We describe a case of a recurrent AVM 9 years postcomplete excision on cerebral angiography during follow-up.

CASE PRESENTATION

History and examination

A 6-year-old girl presented 11 years ago, with a history of headache, drowsiness, emesis, lethargy and meningism. There was a history of a fall 1 month prior to presentation.

Radiological investigation

CT of the brain revealed a subarachnoid haemorrhage with intraventricular extension. Subsequent MRI demonstrated a medial left frontal AVM, fed by one dominant branch of a hypertrophied left-sided anterior cerebral/pericallosal artery. A cerebral angiogram demonstrated a 4 cm AVM in the medial aspect of the left hemisphere, feeding from the anterior cerebral artery. There was deep and superficial venous drainage (Spetzler-Martin Grade II).

Operative and postoperative course

The patient was subsequently admitted the following year to the initial presentation and she underwent

embolisation of the two largest feeding vessels, with approximately 80% of the AVM embolised. Five days later, a bifrontal craniotomy and total excision of a left medial hemisphere AVM was performed. A postoperative angiogram revealed complete excision with filling of all major vessels. The patient was discharged 8 days later with an uneventful postoperative period.

OUTCOME AND FOLLOW-UP

A CT angiogram (CTA) was performed 9 years postexcision, which demonstrated a residual shunt with a remnant nidus filling the surgical resection cavity. Angiography revealed a recurrent nidus, fed by an enlarged left anterior cerebral artery, and meningeal branches of the left external carotid artery with both deep and superficial venous drainage (figures 1 and 2).

DISCUSSION

Cerebral AVMs may result in haemorrhage, seizures, steal and headache. Prevention of haemorrhage is the major indication for treatment. Complete exclusion of the AVM from the circulation as confirmed by postoperative angiography is deemed curative. In the paediatric population there appears to be a definite risk of recurrence in spite of apparent complete angiographic exclusion, and although rare, recurrence is more common in children than adults.⁵

Pathogenesis

Recurrent AVMs have been postulated to be associated with the dysregulation of angiogenesis by vascular endothelial growth factor, and may occur both de novo, or from angiographically occult lesions, perhaps such as in our case.^{6 7} Several large series in the literature reporting long-term surgical results in the treatment of cerebral AVMs have shown that those patients with negative postoperative angiograms (ie, ones that demonstrate no residual nidus or early draining veins) do not experience

Figure 1 Cerebral angiogram-lateral view demonstrating the recurrent arterio venous malformation (aneurysm clips can be seen from the previous surgery).

rehaemorrhage,^{8–10} which is disproven by the incidence of recurrent AVMs, especially in the paediatric population.

Interval of recurrence

AVM recurrence postangiographically proven extirpation has been reported with a rate of 1.5–5.5%.¹¹ Recurrences

Figure 2 Antero posterior view of the cerebral angiogram.

of AVMs have been associated with a higher risk of haemorrhage.¹² From published results it appears that the average age of postoperative recurrence appears between 3 and 4 years. Only two other cases note paediatric recurrence after 9¹³ and 16 years.¹¹ Table 1 summarises the literature available on recurrence of paediatric AVMs over the past three decades. The sensitivity of MR angiogram and CTA is lower than that of angiography, and therefore is not suitable for immediate postoperative follow-up, but may be suitable for interval follow-up following excision of AVM.²

Follow-up

Complete extirpation of arteriovenous malformations and negative postoperative angiograms do not correlate with cure, and patients should be followed up at suitable intervals postsurgery. There are no prospective studies conducted to assess the optimal time for follow-up of angiographically proven surgical extirpation. It is difficult to delineate the appropriate follow-up of resected AVMs, due to the rarity of late recurrence and the unknown natural history of the condition; however, it has been suggested that follow-up at 1 and 5 years via angiography, with non-invasive imaging for further follow-up at 10 years postextirpation.^{11 14}

Treatment

Management options of AVM include embolisation, sclerotherapy, surgical resection and reconstruction and gamma knife.^{15 16} We also recommend that in the initial diagnostic angiography, if there is evidence of angiogenesis surrounding the AVM, instead of surgical treatment, gamma knife can be considered as the primary modality of treatment. This suggestion is based on the probability of postexcision recurrence in the paediatric age group.

Table 1 Summary of literature available on paediatric cerebral arteriovenous malformations and recurrence

Author	Age at first presentation	Recurrence interval
Yasargil ¹⁷	10	7 years
	17	
Kondziolka <i>et al</i> ¹⁵		3 years
		3 years
Kader <i>et al</i> ¹³	5	7 years
	6	9 years
	8	6 years
	11	3 years
	13	2.25 years
Ali <i>et al</i> ¹⁸	7	3 years
Andaluz <i>et al</i> ¹²	4	5 years
Klimo <i>et al</i> ⁵	4	5 years
	7	7 months
	9	6 months
	11	7 months
	12	6 years
Park <i>et al</i> ¹⁹	12	1 year
Takagi <i>et al</i> ²⁰	4	3 months
	4	3 months
	11	3 months
Weil <i>et al</i> ¹¹	8	16 years
	6	9 years, 3 months

Learning points

- ▶ Given the relatively large risk of recurrence of arteriovenous malformations, a long-term follow-up of all postoperative patients who underwent excision of cerebral arteriovenous malformation is recommended.
- ▶ The length of follow-up is debatable; however, definitive research is needed to determine the exact pathogenesis.
- ▶ We recommend that the children should be followed up before the third postoperative until puberty by MRI/MR angiogram.

Competing interests None.

Patient consent Obtained.

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