Findings that shed new light on the possible pathogenesis of a disease or an adverse effect

An early venous abnormality: a potential cause of arteriovenous malformation recurrence

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Summary

In adults, complete surgical resection of arteriovenous malformations (AVMs) is generally curative. Recurrence of AVMs is extremely rare and most often delayed over many years. The authors report the case of a man in his 20s with rapid AVM recurrence and dual blood supply from the dura and intracerebral vessels. Early recurrence of the AVM allowed documentation of the early events associated with this recurrence. This was evidenced by the first appearance of an early vein without any signs of abnormal vasculature, suggesting that abnormality of the venous drainage system might be an inciting event in the recurrence and perhaps genesis of AVMs.

BACKGROUND

Recurrence of arteriovenous malformations (AVMs) is rare and delayed over several years.^{1 2} Here, recurrence of an AVM was documented at a very early stage and was first evidenced by a venous abnormality. This suggests that pathological venous drainage is a potential aetiology for recurrence.

CASE PRESENTATION

The patient is a man in his 20s who presented with dysphasia, seizure and headaches. A CT scan at admission showed an intracerebral haemorrhage. MRI revealed a left frontal parietal AVM. The dysphasia slowly improved while the patient was in hospital. A diagnostic angiogram showed a 3.3×4.1×2.1 cm AVM in the left frontal parietal region (figure 1A,B). Anterior and middle cerebral arteries supplied the AVM. External carotid artery angiograms showed no dural-based efferent communication with the AVM. There were superficial and deep draining veins but no evidence for venous stenosis, efferent arterial aneurysm or intranidal aneurysm.

Staged embolisation of the AVM with ONYX was performed. This accomplished near-complete embolisation of the nidus. This was followed by partial operative excision of the AVM, which was complicated by intraoperative haemorrhage. A month later, the AVM was completely excised and verified by an intraoperative angiogram that showed no abnormal vasculature (figure 1C,D). However, an angiogram performed on postoperative day 1 revealed an early vein suggestive of a small arteriovenous shunt within the excision bed. This arose from the left pericallosal artery with superficial venous drainage into the superior sagittal sinus during the late arterial and early capillary phase, with no clear AVM nidus (figure 1E,F).

An angiogram 5 months after surgical excision demonstrated new, small, serpiginous vessels in the late arterial

phase superiorly in the left frontoparietal lobe. A follow-up angiogram 7 months later demonstrated growth of the AVM with several small nidi spread over a 5 cm diameter area (figure 1G,H). New arterial supply arose from dural branches of the left middle meningeal artery in addition to the left anterior cerebral artery. The primary venous drainage was to the superior sagittal sinus with secondary drainage into a now-enlarged inferior sagittal sinus.

TREATMENT

The recurrent AVM was successfully embolised through the middle meningeal artery branch. The patient subsequently underwent single-dose focused radiotherapy for the residual AVM. There have been no recurrent episodes of haemorrhage.

DISCUSSION

Arteriovenous malformations are generally believed to be congenital lesions that result from the malformation of blood vessels during development, though de novo AVM development has been reported.^{3 4} They are characterised by a tangle of abnormal blood vessels with malformed structure and altered physiology resulting in the shunting of blood under high pressure and at high velocity from the arterial tree to the venous circulation. The most common clinical manifestation is intracranial haemorrhage. When left untreated, most AVMs also increase in size even though a rare lesion may regress.⁵ The mechanisms for the genesis and pathological increase in AVM size are not entirely clear.

The growth of AVMs is presumed to result from the action of multiple cell-derived and extracellular factors,⁶⁻⁹ likely owing to differential gene expression in AVM tissues.¹⁰ Prominent among these are the vasogenic factors.¹⁰⁻¹³

Figure 1 (A) Frontal and (B) lateral views of diagnostic angiogram showing a left frontal parietal arteriovenous malformation supplied by the left anterior and middle cerebral arteries and superficial venous drainage. No arterial side aneurysms or venous stenoses are seen. (C) Frontal and (D) lateral views of a left internal carotid artery postoperative angiogram demonstrate no abnormal vasculature immediately after surgical resection. (E) Frontal and (F) lateral views of the angiogram on postoperative day 1 show a small early vein within the excision bed. (G) Frontal view of a left internal carotid artery angiogram 5 months after surgical excision demonstrates new, small, serpiginous vessels in the late arterial phase superiorly in the left frontoparietal lobe. (H) Follow-up 7 months after resection of the left external carotid artery angiogram demonstrates new arterial supply from dural branches of the left middle meningeal artery.

Altered expression of angiopoietin-2 and vascular endothelial growth factor is detected in the deep venous drainage systems of AVMs.¹⁴ The involvement of venous dysfunction, such as stenosis and chronic hypertension, is also suggested in AVM genesis and growth.⁹ This may lead to local ischaemia/hypoxia with subsequent activation of hypoxia-inducible factor and the cascade of angiogenesis, a phenomenon that may ironically be induced by current AVM treatments.¹⁴ AVM recurrence and growth usually evolve over a number of years, as observed in patients with AVMs who have been followed angiographically over extended periods. Rapid recurrence is distinctly rare.

In the case reported, the initial postoperative angiogram revealed an early draining vein without a distinct AVM nidus. This however rapidly developed into a cluster of AVM vessels in less than a year. Angiographic observation of the status of the AVM over such a short period has allowed documentation of the events leading to its recurrence.

The initial visualisation of an early draining vein may suggest a 'venous' event as being the inciting factor as reported. Whether this was sufficient in inducing vascular growth or represents a hidden compartment model of recurrence remains to be resolved.¹⁵ In addition to the speed at which the AVM reformed, the derivation of subsequent feeding vessels from the dura was also highly unexpected, considering that the patient had undergone operative procedures that would have separated the dura from the cerebral cortex.

In conclusion, this report of the rapid recurrence of an AVM from a residual early draining vein suggests that such a seemingly innocuous finding of an early postoperative vein in the absence of a discrete AVM nidus might be the first and earliest sign of recurrence. This finding must therefore be followed carefully with serial angiograms to detect any potential for recurrence.

Learning points

- ▶ Complete arteriovenous malformation (AVM) excision shown by one angiogram is not adequate to demonstrate a cure. Serial angiograms are essential to ensure no recurrence of the AVM.
- ▶ A venous abnormality in the bed of a previously completely excised AVM must be followed up with additional angiographic studies.
- ▶ Long-term follow-up with angiography is important to ensure no late recurrence.

Competing interests None.

Patient consent Obtained.

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Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Weingarten D, Pakbaz S, Cho A, U HS. An early venous abnormality: a potential cause of arteriovenous malformation recurrence. *BMJ Case Reports* 2012;10.1136/bcr.01.2012.5646, Published XXX

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