Case Report

Recurrent Cerebral Arteriovenous Malformation in a Child : Case Report and Review of the Literature

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Arteriovenous malformations (AVM) are generally considered to be cured following angiographically proven complete resection. However, rare instances of AVM recurrence have been reported in both children and adults with negative findings on postoperative angiography. The authors present the case of a 12-year-old boy with recurrent AVM. The AVM was originally fed by the pericallosal arteries on both sides, and it showed changing patterns of supply at recurrence. The authors concluded that a negative postoperative angiogram is not necessarily indicative of a cure. Repeat angiography and regular follow-up examinations should be performed to exclude the possibility of recurrence, especially in children.

KEY WORDS : Arteriovenous malformations · Cerebral angiography · Recurrence.

INTRODUCTION

Arteriovenous malformation (AVM) is the most frequent cause of hemorrhage in children[9]. Complete excision or obliteration of the lesion usually eliminates the risks of hemorrhage, and a negative cerebral angiogram is generally considered indicative of a cure. Although recurrence of cerebral AVMs is rare, there are several reports of cerebral AVM recurrence in both children[1,2,4,9-11] and adults[3,5,8,16] after complete resection verified by postoperative angiography.

Although the pathogenesis underlying the recurrence of cerebral AVMs after total extirpation is unknown, dysregulation of angiogenesis by vascular endothelial growth factor (VEGF) and the concept of hidden compartments have been suggested[6,14].

We present a case of recurrent AVM in a child whose angiographic findings showed some pathogenetic evidence of AVM recurrence, provide a brief review of the literature regarding this type of recurrent lesion and discuss the possible mechanisms of regrowth.

CASE REPORT

A 12-year-old boy visited our hospital due to the onset of

a headache after falling down. Brain computed tomography showed an incidental vascular lesion in the left frontal lobe and corpus callosal area. Angiography confirmed that the 4×3 cm sized AVM was fed by the pericallosal and callosomarginal arteries on both sides and drained by the superficial cortical and deep veins (Fig. 1). Variceal changes and evidence of an arteriovenous fistula were also noted in the venous phase. The AVM was completely extirpated using an interhemispheric approach. Right pericallosal artery was truncated at the just distal portion of branching callosomarginal artery and left pericallosal and left callosomarginal arteries were cut away together. Angiography performed at the 9th postoperative day showed no residual nidus or early draining vein (Fig. 2). Routine postoperative angiography performed one year later showed a recurrent AVM at the same location as the original lesion (Fig. 3). The recurrent AVM was fed by numerous fine arteries from the right pericallosal artery and left lateral lenticulostriate artery. Draining was only via the deep venous system. His parents did not wish to repeat the surgery, and he was transferred for radiosurgery.

DISCUSSION

Although the actual rate of AVM recurrence is not known due to the lack of routine long-term follow-up and postoperative documentation of complete removal, a surgical series of children with AVM reported that the rates of recurrence ranged from 1.5 to 5.5%[2]. These are all treated with
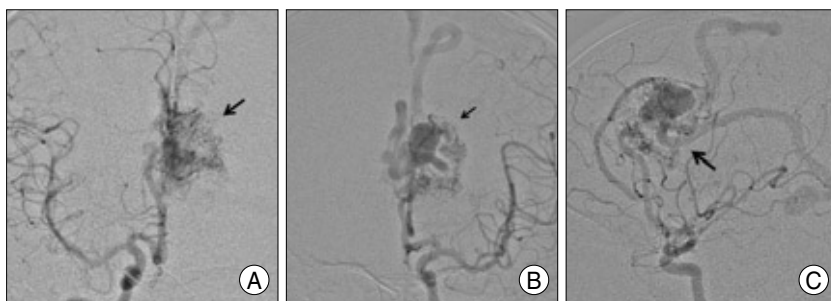
Fig. 1. Angiography of the internal carotid artery. Initial right (A) and left (B) anteroposterior and lateral (C) angiogram reveals an arteriovenous malformations of the pericallosal area mainly fed by pericallosal and callosomarginal artery. Draining veins are the superficial cortical vein and deep vein.
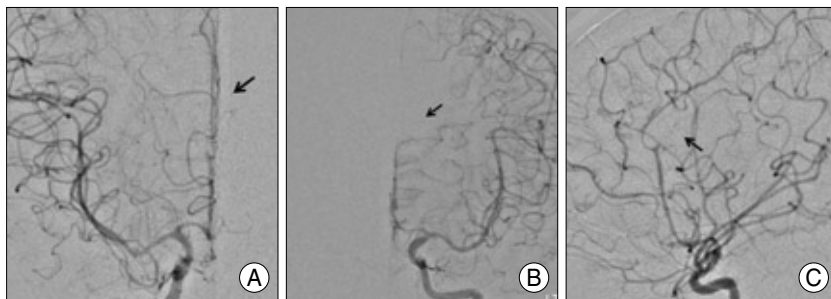

Fig. 2. Postoperative angiogram shows no residual nidus or early draining vein. A : Right. B : Left. C : Lateral.
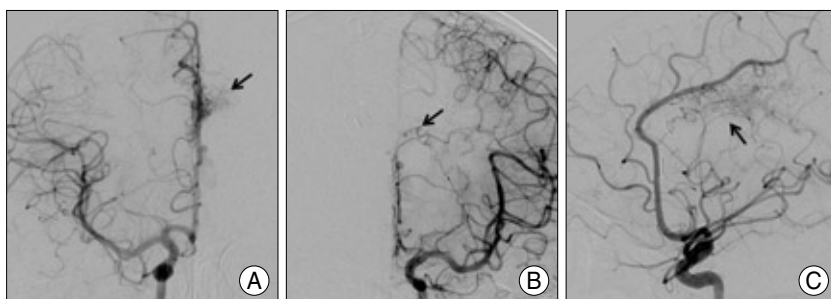

Fig. 3. One-year postoperative angiogram shows a recurrent arteriovenous malformations at the original site and is fed by right pericallosal artery and left lenticulostariate arterial branch. A : Right. B : Left. C : Lateral.

microsurgical resection, and total extirpation was documented with a postoperative angiogram.

There are some concepts about plausible mechanisms of AVM recurrence. Hladky et al.[9] documented two cases of recurrence, one located in the corpus callosum and the other located in the parietal lobe, among the 62 pediatric cases in which AVM surgery had been performed. They suggested that recurrence caused by microshunts might remain invisible on early postoperative angiography. Some authors suggested that regrowth of childhood AVM may be attributable to the inherent nature of the development of an original abnormal vasculature[4]. Recurrence is definitely more common in children than in adults. The early evolution of AVMs is consistent with the diffuse patterns of arteriovenous shunting observed in children as opposed to the more distinct fistulous pattern seen in adults[13]. If more mature

AVMs develop along a continuum as the brain develops, children are more likely to have immature vasculature as part of their AVM. Immature vessels left in the surgical bed may not be visible on angiography, but they may retain the ability to regrow and form a new malformation in the same location[5]. This hypothesis is supported by the findings of Klimo et al.[11], who reported that diffuse lesions were disproportionately represented in cases of failed obliteration, residual lesion, or recurrence. Sano et al.[15] proposed a concept called "reserve nidus" which refers to abnormal vascular groups around the main nidus. They suggested that the "reserve nidus" should be extirpated, or it may become nidus several years later. Although they described the reserve nidus as an illustration, it seemed not to be based on angiographic or surgical evidence rather an inference from recurrence of the AVM near the original site (cingulated and callosal region at first and ventricle at recurrence). A few years later, Pelletieri et al.[14] introduced the concept of "hidden compartments", that is, unfilled compartments that may exist within an AVM but are not seen on angiograms. Within the predetermined boundaries of the malformed vascular tree, these compartments can have separate feeders and drainage. The hidden compartments could be visualized as 'recurrence' after hemodynamic changes such as embolization and serial filling of those compartments could increase the AVM size named 'growth'. However, there are no definite proven mechanisms to explain the recurrence of extirpated AVM.

Dysregulation of angiogenesis by VEGF was also suggested as a plausible mechanism of recurrence[17]. The level of VEGF expression in children with recurrent AVMs was higher than that reported in nonrecurrent groups, suggesting that VEGF may play a role in AVM recurrence. Hashimoto et al.[7] reported that AVM vessels from younger patients tended to have a higher Ki-67 index. These results provided evidence of increased endothelial cell turnover in AVMs, which may be indicative of active vascular remodeling or angiogenesis.

In the present case, a new feeder arose from the lateral lenticulostriate artery, which was not included to the original feeders, and the corpus callosum is not usually supplied by the lenticulostriate arteries. In addition, the new feeder had to span quite a long distance in order to form the new nidus in the original site. The scenario concerning AVM recurrence or regrowth seemed to be inferred from its angiographic course. The scattered anomalous compartment surrounding the main AVM nidus, which had a low flow rate, could not be opacified because of internal steal. After high flow feeders have been eliminated, scattered anomalies recruit the small and slow current vessels. Regardless of the presence of a hidden compartment, unorganized abnormal vasculatures around the nidus core can recruit the new feeders and finally form the reorganized AVM at the original site. In this case, the pericallosal arteries on both sides had been truncated anatomically. The recurrent nidal core had to gather vessels from remote areas.

Table 1. Recurrent arteriovenous malformations reported in the literature

Authors and published year	Age	Sex	Location	Interval	Feeding and draining
Kadar et al.[10] 1996	8	F	Orbitofrontal and sylvian	6 years	1st : branches of ICA, MCA, ACA and superior sagittal sinus drain, 2nd : ethmoidal branches and choroidal branches
	6	F	Sylvian	9 years	1st : branches of MCA, anterior sylvian vein drain, 2nd : lenticulostriate branches and SSS drain
	11	M	Medioparietal	3 years	1st : bilateral ACA, unilateral posteromedial choroidal and splenial branches and superficial drain, 2nd : not performed
	5	F	Sylvian	7 years	1st : branches of MCA and SSS and transverse sinus drain, 2nd : branches of MCA, anterior choroidal artery and superficial drain
	13	F	Parietooccipital	15 months	1st : branches of PCA, galenic system drain, 2nd : branches of PCA, galenic system drain
Klimo et al.[11] 2007	11	M	Parietal	7 months	1st : branches of MCA, ACA, 2nd : branches of MCA, ACA
	12	M	Parietal	6 years	Not mentioned
	7	F	Interhemispheric	7 months	1st : branches of pericallosal artery, 2nd : not mentioned
	1	F	Medial temporal	2 years	1st : branches of PCA, posterior choroidal artery, 2nd : not mentioned
	9	M	Parietal	6 months-8.5 years	Not mentioned
Hladky et al.[9] 1994	NA	NA	Corpus callosum	NA	Not mentioned
	NA	NA	Parietal	NA	Not mentioned
Andaluz et al.[2] 2004	4	M	Temporal	5 years	1st : MCA and PCA branches and Rosenthal draining, 2nd : MCA branches only
	6	F	Corpus callosum	3 years	1st : ACA branches feedings, 2nd : not mentioned
Ali et al.[1] 2003	7	M	Frontal	8 years	1st : not mentioned, 2nd : ACA, ECA and cortical draining
	7	M	Parietal	8 years	1st : not mentioned, 2nd : MCA and choroidal artery and
Freudenstein et al.[4] 2001	17	?	Parietooccipital	5 years	1st : anterior and posterior choroidal arteries, galenic system drain, 2nd : branches of PCA and callosomarginal artery
Sano et al.[15] 1978	14	M	Cingulate and callosal region	9 years	2nd : posterior choroidal artery
	7	M	Frontal	13 years	2nd : branches of ACA
Kondziolka et al.[12] 1992	NA	NA	Temporal	3 years	Not mentioned
	NA	NA	temporal	3 years	Not mentioned
Yasargil[18,19] 1987	17	F	Frontal operculum	7 years	1st : M1, M2, A1, A2 feeding and superficial ascending vein, internal cerebral vein drain, 2nd : not mentioned
Present case	12	M	Corpus callosum	12 months	1st : bilateral pericallosal and callosomarginal arteries and superficial cortical vein and galenic system drain, 2nd : unilateral pericallosal artery and unilateral lenticulostriate arterial branch and galenic system drain

ICA : internal carotid artery, MCA : middle cerebral artery, ACA : anterior cerebral artery, SSS : superior sagittal sinus, PCA : posterior cerebral artery, ECA : external carotid artery, NA : non-available

From the review of literatures, we found 4 cases of recurrent AVMs that recruited the feeders from remote area for forming new nidus; three of four[10] had new deep arterial feeding vessels (choroidal arteries, lenticulostriate arteries) that were not present originally and the one of four[4] had deep arterial feeders (anterior and posterior choroidal arteries) initially and superficial arteries (branches of PCA and callosomarginarl artery) were then involved. Therefore, recurrence or regrowth should be an active process, and VEGF or innate angiogenetic potential may play a role in these processes. There would likely to be some correlation between location and recurrence. The pericallosal area is along the midline and can be supplied bilaterally by the anterior cerebral artery branches. Deep location can make vascular recruitment more available and less difficult. The series of recurred AVMs were summarized in Table 1.

The latency from the initial presentation to recurrence varied from 6 months to 9 years. The second presentation would be more serious. There were two cases in which the lesion recurred twice in the literature,[3,4,11] and one case involved a woman who initially presented at the age of 33 years and experienced the second recurrence at the age of 42[3].

CONCLUSION

AVM recurrence may occur via an active vascular recruitment process. The authors conclude that a negative postoperative angiogram is not necessarily indicative of a cure. Repeat angiography and regular follow-up should be performed to exclude the possibility of recurrence, especially in children.

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