

Case Report

Endovascular Embolization of a Ruptured Distal Lenticulostriate Artery Aneurysm in Patients with Moyamoya Disease

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A ruptured distal lenticulostriate artery (LSA) aneurysm is detected occasionally in moyamoya disease (MMD) patients presented with intracerebral hemorrhage. If the aneurysm is detected in hemorrhage site on angiographic evaluation, its obliteration could be considered, because it rebleeds frequently, and is associated with poorer outcome and mortality in MMD related hemorrhage. In this case report, the authors present two MMD cases with ruptured distal LSA aneurysm treated by endovascular embolization.

Key Words : Moyamoya disease · Cerebral hemorrhage · Therapeutic embolization.

INTRODUCTION

A ruptured aneurysm on collateral vessels is detected occasionally in patients with moyamoya disease (MMD) who present with intracerebral hemorrhage. This aneurysm is usually located in the anterior or posterior choroidal artery, but some can be detected in the distal lenticulostriate artery (LSA). In MMD related hemorrhage, rebleeding is known to be more frequent than spontaneous one⁶, and associated with poorer outcome and mortality^{7,9}. Therefore, obliteration of this aneurysm which is detected in the center of hematoma could be considered.

Herein, we describe technique and follow-up results of two MMD cases with ruptured distal LSA aneurysms treated by embolization using n-butyl-cyanoacrylate (NBCA) for prevention of its rebleeding.

CASE REPORT

Case 1

A 53-year-old woman presented with sudden stuporous mentality. Brain CT scan showed a hemorrhage in the left basal ganglia, which was extended into the lateral ventricle. She was diagnosed with MMD 2 years ago on MRI performed for evaluation of chronic headache. Based on her past history and CT findings,

diagnostic angiography was performed to detect any bleeders, and revealed a ruptured aneurysm in the left medial LSA, supplying the frontal cortex via medullary vessels (Fig. 1A). The aneurysm was located in a branch of this LSA, which was another medullary vessel connected with the insular branch of the M2 segment (Fig. 1B).

Because the patient was uncooperative, the procedure was performed under general anesthesia. Echelon-14 microcatheter (EV3, Irvine, CA, USA) was positioned directly at the target branch with the aneurysm using Synchro-14 wire (Stryker, Fremont, CA, USA) in order to save medullary vessels from this LSA. Selective angiogram showed that delayed filling defect in the M2 branch, which was connected with the LSA. This finding confirmed another collaterals to this lesion (Fig. 1B). The target branch was perfectly wedged by the microcatheter. To prevent a retrograde filling of the aneurysm from the M2 branch, the target branch of the LSA was embolized with a 33% NBCA mixture with lipiodol up to the point connecting with the insular branch of the M2 (Fig. 1C). Postembolization angiogram showed no aneurysmal filling. Patent distal vessels of the LSA and M2 branches were also confirmed (Fig. 1D).

Follow-up CT scan showed the aneurysm and target branch filled with NBCA (Fig. 1E). No infarction in the adjacent structure and cortex area was detected on diffusion weighted imaging

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(DWI). The patient fully recovered and no rebleeding occurred for 1 year after embolization. Aneurysm recurrence was not detected on 1-year follow-up angiography.

Case 2

A 44-year-old woman presented with stuporous mental status. Brain CT scan revealed intraventricular hemorrhage extended from a focal hemorrhage in the left caudate nucleus. The patient had a history of intracerebral hemorrhage in the right temporal lobe 8 years ago and had been diagnosed with MMD. Diagnostic angiography showed a ruptured aneurysm on medullary vessel from the LSA connected with cortical branches supplying frontal area. Additional aneurysmal dilatation was noted in these cortical branches, but its location was not in the hemorrhage site (Fig. 2A).

Under general anesthesia, Echelon-14 microcatheter was navigated into the target LSA using Synchro-14 wire. To avoid a reflux of NBCA into M1 and another vessels, the microcatheter was inserted up to proximal segment of the target LSA over its origin. Selective angiogram showed the aneurysm and cortical branches via medullary vessels (Fig. 2B). These cortical branches

were found to disappear after brief opacification on selective angiogram, suggesting collateral vessels from other LSA. Occlusion of the target LSA with wedging microcatheter into the LSA induce no perfusion defect and branch loss on repeated angiograms. A 25% NBCA mixture with lipiodol was slowly injected via the microcatheter up to the aneurysm (Fig. 2C). Postembolization angiogram revealed complete obliteration of the aneurysm and patent cortical branches (Fig. 2D). No retrograde filling of the aneurysm was noted on venous phase of angiogram.

CT scan taken 3 days after embolization showed the aneurysm and LSA filled with NBCA (Fig. 2E). No infarction was detected on DWI. The patient also fully recovered without neurologic deficit. One-year follow-up angiography did not reveal the aneurysmal recurrence. No rebleeding developed during 2 years after embolization.

DISCUSSION

In MMD, LSAs serve as important transmedullary collateral vessels supplying cortex areas as well as original vascular territories, instead occluded major arteries. Fragile nature of these

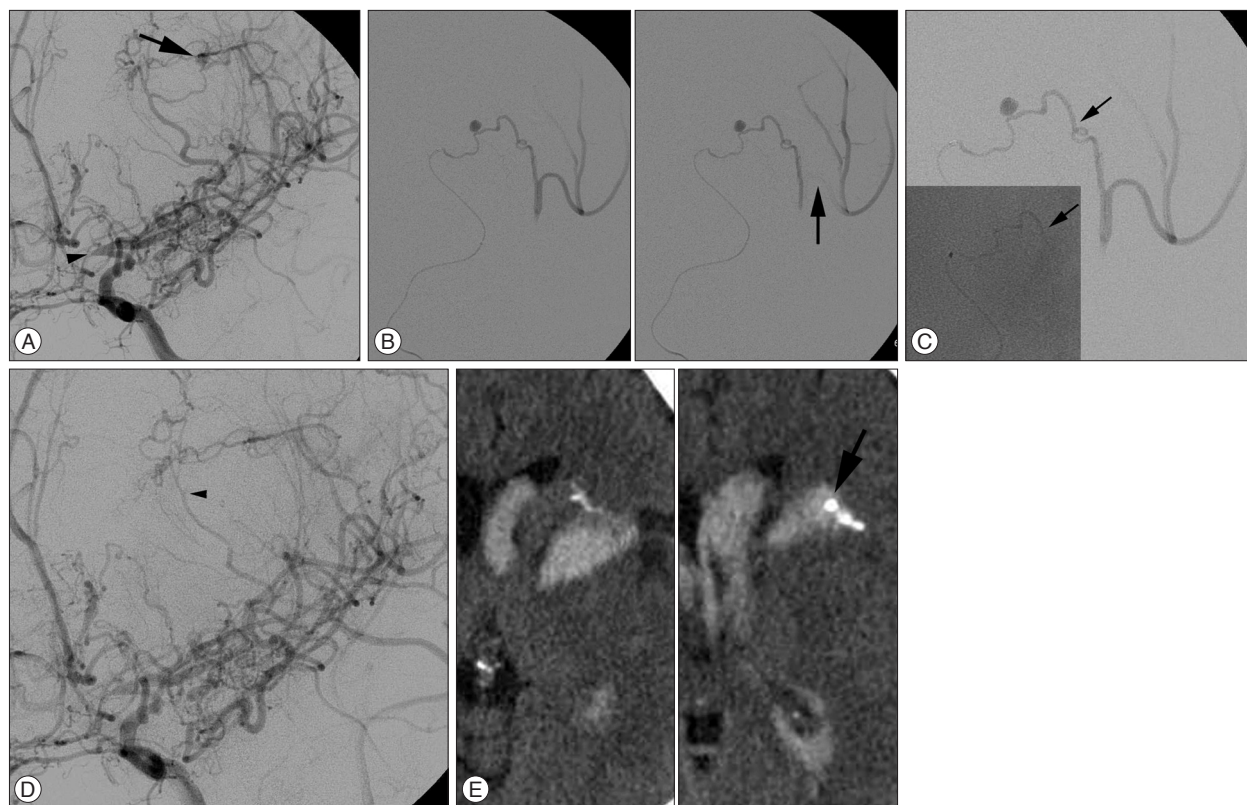


Fig. 1. A : A ruptured aneurysm (arrow) is located on a branch of the lenticulostriate artery (LSA), which connects this LSA with the insular branch of the M2 segments of left middle cerebral artery (MCA). The LSA is originated from the A1 segment of the anterior cerebral artery (arrowhead), which indicates that the LSA is a medial one. Distal medullary vessels from this LSA are also noted. B : The target branch with the ruptured aneurysm is selected directly with a microcatheter to save the distal medullary vessels. Selective angiogram shows the target branch and the MCA branches connected with it. These MCA branches disappear on selective angiogram after brief contrast opacification (arrow), suggesting other collateral flows. C : N-butyl-cyanoacrylate (NBCA) is injected into the target branch, not over the M2 segment. Arrows indicate the intended target point and the final glue endpoint. D : Postembolization angiogram shows patent flows of the medial LSA, its distal medullary vessels, and the MCA branches. The target branch with the aneurysm is no more seen except its stump (arrowhead). E : The target branch and aneurysm (arrow) filled with NBCA are observed on follow-up CT scan.

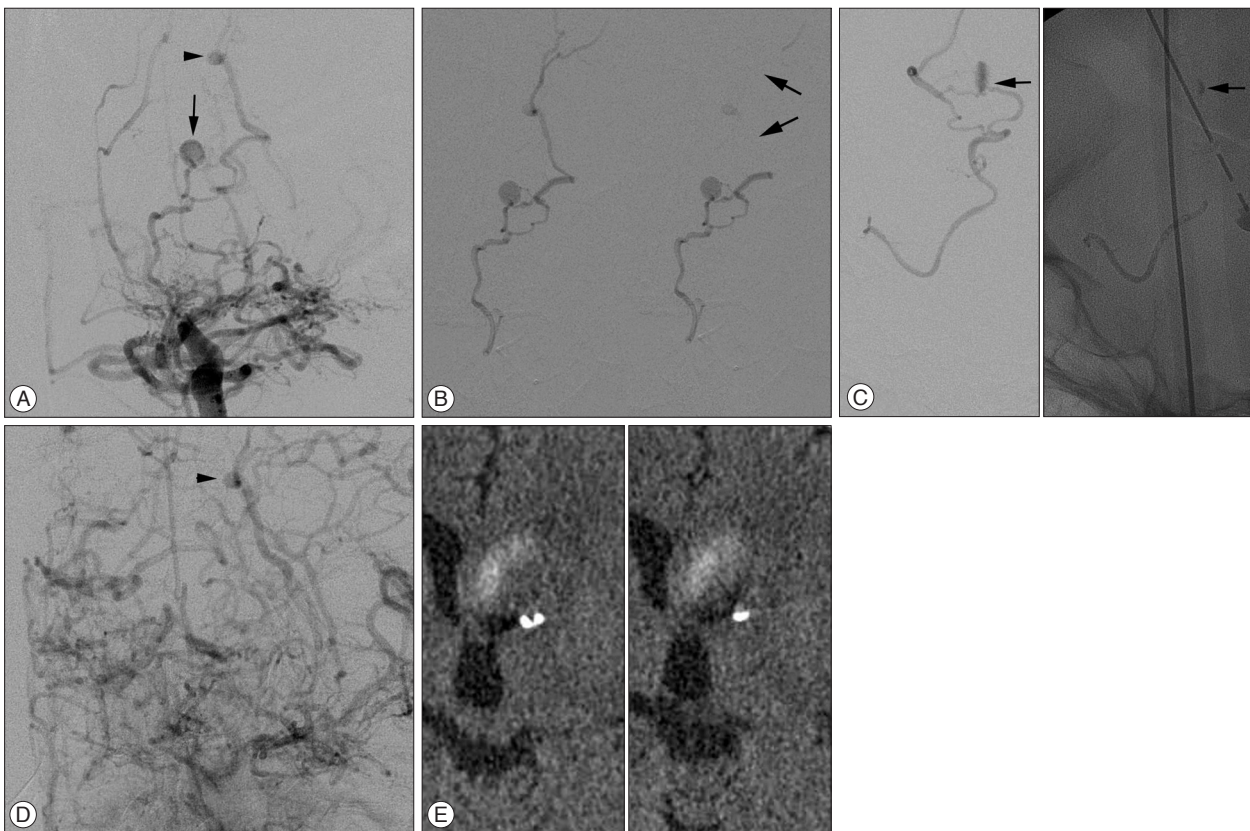


Fig. 2. A : A ruptured aneurysm (arrow) is noted in the distal lenticuloostriate artery (LSA). Another aneurysmal dilatation (arrowhead) is also observed on a cortical vessel via distal medullary vessel from this LSA. B : Selective angiogram shows the aneurysm and the cortical branches. Contrast in the cortical branches washes out after brief opacification, suggesting that the LSA with the aneurysm is wedged with a microcatheter and its distal branches are supplied by blood flow via another collaterals (arrows). C : Treatment working view. N-butyl-cyanoacrylate (NBCA) is injected into the LSA up to the aneurysm (arrows). D : Postembolization angiogram shows complete obliteration of the aneurysm and patent flow of the distal cortical branches with the aneurysmal dilatation (arrowhead). E : NBCA filling in the LSA and aneurysm is detected on follow-up CT scan.

collateral vessels and hemodynamic stress have been considered as a cause of hemorrhage in MMD. Although aneurysmal lesion on collateral vessels is not frequently encountered in MMD patients with hemorrhage, if it is detected on angiography and well correlated with bleeding site, endovascular treatment for aneurysmal lesion could be considered as an option for prevention of rebleeding^{3,4}.

In our experience of ruptured aneurysms in choroidal arteries in MMD patients, these arteries are usually straight, because of their subependymal location covered by ependymal layer along with ventricular wall⁴. Therefore, microcatheter can be easily advanced near the aneurysm. However, because LSAs have originally tortuous course and relatively small diameter, an access to the distal LSA aneurysm with microcatheter can be limited. In this situation, occlusion of long segment of the LSA can make large infarction beginning from its proximal territory and additional infarctions in cortex area supplied by its distal medullary vessels². The easy way to avoid infarction and consequent aggravation of neurological status is the selective embolization for the target branch with the aneurysm limited within hemorrhage site and saving other collateral vessel. Although, we were able to perform the selective embolization in Case 1, it is not always possible.

When embolization of long segment of the LSA is inevitable, pre-embolization provoking test such as amobarbital test can be used³. However, in uncooperative MMD patients with hemorrhage, because this type of test was unavailable, we used careful inspection of repeated angiogram findings after wedging microcatheter into the target LSA as a simulation test, in Case 2. Although some collateral flows were confirmed, fine and complex collateral networks all of which could not be traced were found to make reverse flows, leading to no perfusion defect and branch loss on the repeated angiograms. Other reports described no detailed angiographic information on collateral flows, but reported no infarction and its related sequelae, as our cases^{1,5}. Consequently, these cases suggest that collateral networks which develop extensively in MMD would be critical determinant for successful outcomes and a background for endovascular embolization of ruptured distal LSA aneurysm in MMD patients.

Although these collateral networks prevent infarction, they could make a retrograde filling into the aneurysm and even rebleeding, unless obliteration of the aneurysm itself is achieved. However, the LSA was usually wedged without antergrade flow after placing the microcatheter in our practice. This permitted fine control of NBCA without its reflux. Therefore, after adequate

injection volume, pressure, and speed were confirmed by several test injection with contrast, we were able to penetrate NBCA up to the aneurysm or target point, avoiding occlusion of collateral vessels beyond the target. Compared with Onyx (EV3, Irvine, CA, USA), NBCA has a tendency to go straight to a target. In our cases, therefore, because embolic material had to be injected at a point far from the target lesion, we selected NBCA as adequate embolic material.

Although more long-term follow-up is necessary, aneurysm recurrence and rebleeding were not detected in our and other reports^{3,5,8}. This may indicate that endovascular embolization is effective for prevention of rebleeding from the distal LSA aneurysm in MMD, if successful. However, because of limited cases, more investigation is necessary for proving the benefit of endovascular embolization.

CONCLUSION

A ruptured distal LSA aneurysm is not frequent in patients with MMD who present hemorrhage. However, if it is detected on angiographic evaluation and can be accessed, endovascular embolization could be considered for prevention of rebleeding. Special consideration of collateral network is mandatory.

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