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Case Report

Transbrachial Mechanical Thrombectomy for Acute Ischemic Stroke in Marfan's Syndrome: A Case Report

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Objective: Marfan's syndrome (MFS) is a systemic connective tissue disorder with autosomal dominant inheritance. Cardiovascular complications of MFS such as aortic root or valve disease and aortic aneurysm or dissection are potential cause of access route problems of mechanical thrombectomy (MT) for acute ischemic stroke (AIS). Here, we report a case of a patient with MFS who underwent MT for AIS.

Case Presentation: A 58-year-old woman with MFS presented with a sudden onset of consciousness disturbance and right hemiparesis, and was referred to our hospital. After the infusion of tissue plasminogen activator, CTA showed a type III arch in the aortic arch and severe tortuosity of the thoracoabdominal aorta; thus, angiography was performed using the transbrachial approach. Left common carotid angiogram showed complete recanalization of the left middle cerebral artery. On the sixth day, the patient presented a sudden consciousness disturbance and left hemiparesis. MRA showed right internal carotid artery occlusion. MT was performed by the transbrachial approach, and complete recanalization was achieved on the first pass.

Conclusion: MT via the transbrachial approach is a treatment option that should be considered, especially in MFS, where the transfemoral approach is difficult due to anatomical problems.

Keywords ▶ Marfan's syndrome, transbrachial approach, mechanical thrombectomy, acute ischemic stroke

Introduction

Marfan's syndrome (MFS) is an autosomal dominant genetic pathology affecting the cardiovascular system, respiratory system, skeletal muscles, bones, and eyes. This syndrome results from a mutation in the gene encoding the fibrillin-1 protein, and its protein abnormalities cause tissue fragility and damage. In patients with MFS, the risk of cerebrovascular diseases, including carotid and vertebral artery dissection; cardiogenic cerebral infarction; and subarachnoid hemorrhage due to a ruptured cerebral aneurysm is high. Acute ischemic stroke (AIS) is rare in MFS, and major guidelines about the safety and efficacy of endovascular treatment are not presented.[1)]

In some studies in the literature, thrombolytic therapy with tissue plasminogen activator (tPA) or mechanical thrombectomy (MT) for AIS associated with MFS has been reported[2–5)]; however, the safety and efficacy have not been well known.

Here, we present a case of a patient with MFS who underwent endovascular MT for cardiogenic cerebral infarction using the transbrachial approach.

Case Presentation

A 58-year-old woman had been diagnosed with MFS at our hospital. The patient has a medical history of MFS, chronic heart failure, atrial fibrillation, aortic artificial blood vessel replacement, mitral valve replacement, artificial blood vessel replacement, and stent-graft insertion for thoracoabdominal aortic aneurysm. The patient was taking warfarin (3 mg) prior to admission.

The patient presented with sudden onset of consciousness disturbance and right hemiparesis, and was transferred to our hospital. On arrival, Glasgow Coma Scale (GCS)

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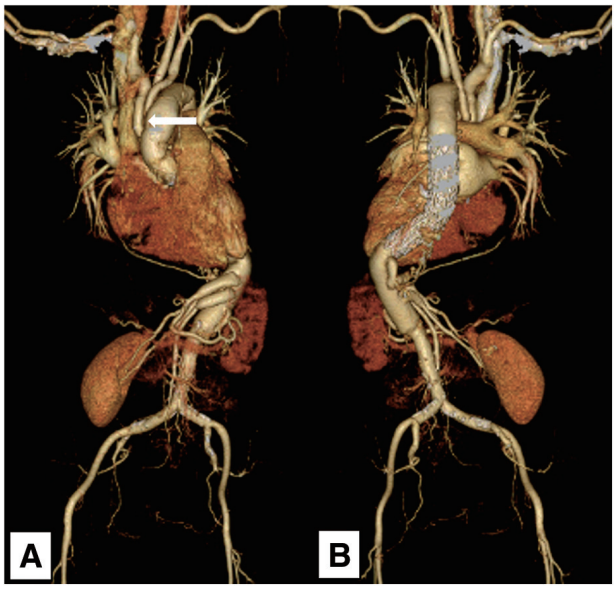

Fig. 1 (**A**) In the anterior view, preoperative 3D CTA showed that flexion of the aorta and the angle between left common carotid artery (white arrow) and the aortic arch were very steep. (**B**) In the posterior view, an artificial vascular stent was placed in the thoracoabdominal aorta and a thoracoabdominal aortic aneurysm was also observed.

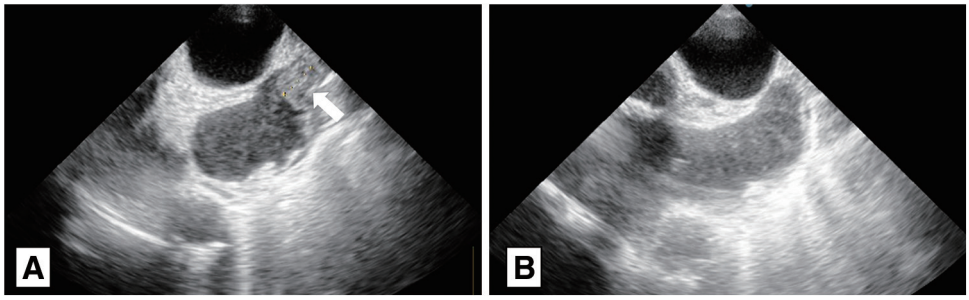

Fig. 2 Postoperative findings in the imaging studies. (**A**) On day 1, echocardiography revealed a 17-mm thrombus in the left atrial appendage (white arrow). (**B**) After MT, the thrombus in the left atrial appendage had disappeared on day 7. MT: mechanical thrombectomy

score of the patient was 10 (E2V4M4), and the National Institutes of Health Stroke Scale (NIHSS) score was 16. The coagulation/fibrinolysis system showed a normal prothrombin time (PT) (17.7 s), PT/international normalized ratio (INR) (1.48 s), and activated PT (31.8 s); however, D-dimer was slightly increased (14.6 μg/mL). Diffusion-weighted imaging (DWI) on MRI revealed a hyperintense area in the left insular cortex and corona radiata. MRA showed obstruction of the M1 segment of the left middle cerebral artery (MCA). After tPA infusion, CTA performed following MRA showed a type III arch in the aortic arch and severe tortuosity of the thoracoabdominal aorta (**Fig. 1A** and **1B**); therefore, angiography was performed using the transbrachial approach. Angiography of the left common carotid artery showed revascularization of the left MCA.

On the first day of admission, echocardiography revealed a 17-mm thrombus in the left atrial appendage (**Fig. 2A**); thus, anticoagulation with warfarin was increased from 3 mg to 3.5 mg based on the INR. The patient's symptoms improved thereafter, and she could independently perform physical activities on the fifth day.

On the sixth day after the initial presentation, the patient presented with a sudden consciousness disturbance (GCS score of 8) and left hemiparesis. DWI-MRI showed a hyperintense area in the right insular cortex, and MRA showed an occlusion of the right internal carotid artery (ICA) (**Fig. 3A**–**3C**). The NIHSS score was 25. Because of the patient's recent stroke, intravenous tPA was contraindicated; therefore, we performed endovascular MT for the ICA occlusion. MT was performed under local anesthesia using dexmedetomidine and pentazocine hydrochloride. Angiogram showed poor visualization of the ICA (**Fig. 3D** and **3E**). Then, a 9-Fr Optimo balloon guide catheter (BGC; Tokai Medical Products, Aichi, Japan) was advanced into the origin of the right ICA along with a 6-Fr JB2 inner catheter

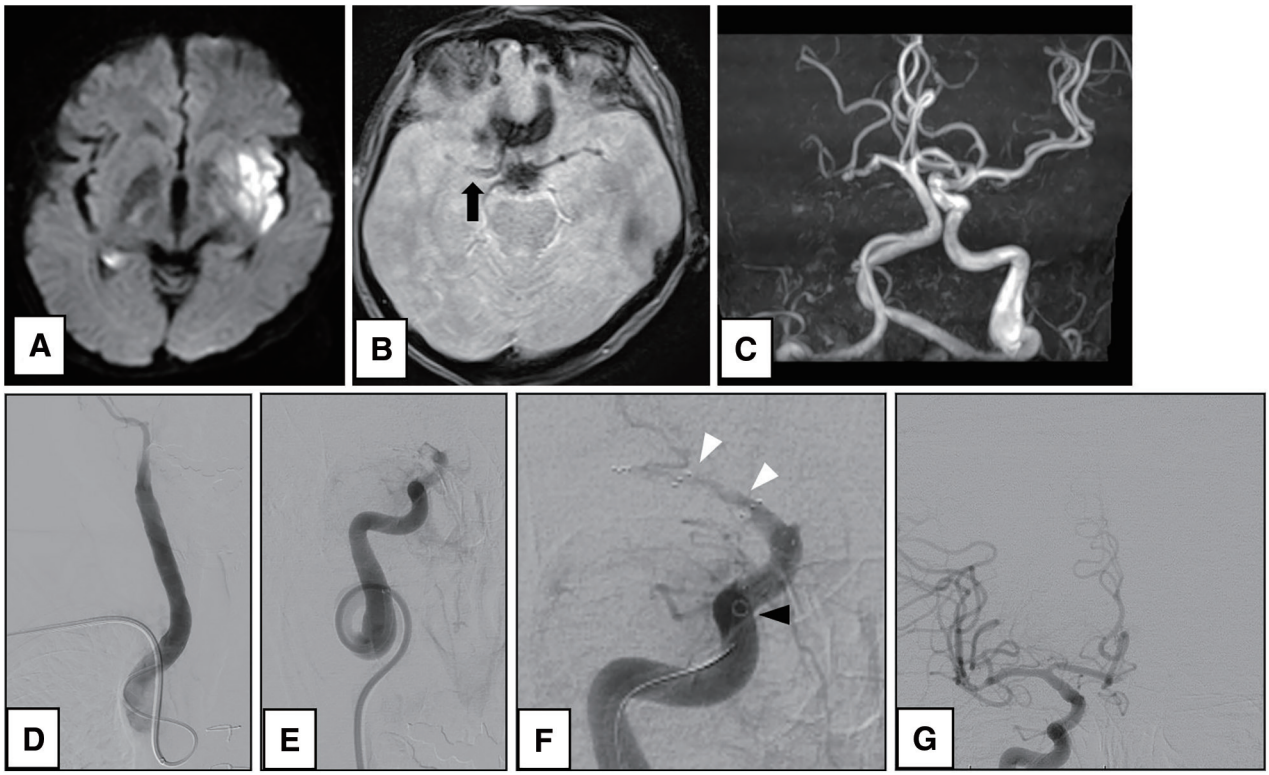

Fig. 3 (**A**–**C**) On day 6, DWI showed a high-intensity area in the right insular cortex and deep white matter; T2-weighted imaging showed a low-intensity area (black arrow) in the right ICA; and MRA showed right ICA occlusion. (**D**) In the anterior view, the transbrachial approach was selected again, and cerebral angiography before MT showed right carotid artery occlusion. (**E** and **F**) In the anterior view, we performed MT using a combined technique with a stent retriever (white arrowheads) and an aspiration catheter (black arrowhead). (**G**) Angiogram after MT showed complete recanalization of the right ICA (TICI 3). DWI: diffusion-weighted imaging; ICA: internal carotid artery; MT: mechanical thrombectomy; TICI: thrombolysis in cerebral infarction

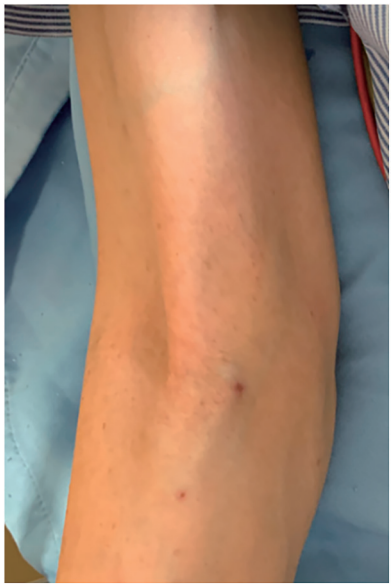

Fig. 4 On 30 days after MT, no local complications were found in the puncture site. MT: mechanical thrombectomy

(Medikit, Tokyo, Japan) and 0.035-inch guidewire (Fukuda Denshi, Tokyo, Japan). First, the React 71 (Medtronic, Irvine, CA, USA) was advanced to the petrous portion of the ICA through a Phenom 27 microcatheter (Medtronic, Minneapolis, MN, USA) and CHIKAI-14 micro guidewire (Asahi Intecc, Aichi, Japan). The Optimo BGC was inflated to arrest blood flow. Then, the Phenom 27 microcatheter was advanced to the M2 segment of the MCA. The Solitaire 6 mm/41 mm (Medtronic) was fully deployed using the push and fluff technique over the thrombus (**Fig. 3F**). The stent retriever and aspiration catheter were retrieved as a unit while suctioning from Optimo BGC using a VackLok syringe. A large amount of thrombus was aspirated from the Optimo BGC. Complete recanalization was achieved on the first pass (**Fig. 3G**), and the procedure was finished after hemostasis of the puncture site using a compression device for 10 hours.

After MT, consciousness disturbance and left paralysis improved promptly, and the thrombus found in the left atrial appendage disappeared due to postoperative anticoagulant therapy on the seventh day (**Fig. 2B**). Postoperative ultrasound echo revealed that the brachial artery diameter was 4.7 mm, and the 9-Fr introducer sheath was fully inserted. No obvious puncture site complications were observed on the 30 days (**Fig. 4**).

Table 1 Summary of case reports with thrombolysis or endovascular MT for MFS patients

Case	Age	Sex	NIHSS	Etiology	Medications	Case reports
1	57	Male	12	Unknown	Thrombolysis	Chembala et al. 2012[2]
2	34	NA	NA	Unknown	Thrombolysis	Gójska-Grymajło et al. 2014[3]
3	NA	NA	25	Cardiogenic embolism	MT	Reznik et al. 2017[5]
4	47	Female	15	Dissection was suspected	Thrombolysis	Lai et al. 2020[4]
5	58	Female	16	Cardiogenic embolism	Thrombolysis + MT	Our case

MFS: Marfan's syndrome; MT: mechanical thrombectomy; NA: not available; NIHSS: National Institutes of Health Stroke Scale

Warfarin was administered for atrial fibrillation, and the patient's modified Rankin Scale score 3 months after surgery was 4.

Discussion

In patients with MFS, the risk of cerebrovascular diseases including carotid and vertebral artery dissection and cardiogenic cerebral infarction is high, that estimated to occur in 10% to 20%[6]; however, only four case reports of patients with MFS with AIS receiving thrombolysis or endovascular thrombectomy have been reported in the literature (**Table 1**). Three of the four cases were described as cerebral infarction due to cardiac embolism,[2,3,5] and only one case suggested MCA dissection.[4] In the literature, AIS in MFS is mainly due to cardiac embolism. Wityk et al.[7] described that the cardiac source of embolism was present in 10 out of 13 AIS patients (77%), including prosthetic cardiac valves in nine patients and atrial fibrillation in one. All ten patients were under treatment with anticoagulants. Of the remaining three patients, two suffered mitral valve prolapse and one had no identifiable source of embolism. Reznik et al.[5] have reported a case of MT for right MCA occlusion in an MFS patient with a history of chronic aortic dissection. MT was performed using a transfemoral approach, which yielded successful recanalization with thrombolysis in cerebral infarction (TICI) 2b flow. The patient had mechanical aortic valve and suggested the cardiogenic embolism.

Patients with MFS are complicated by scoliosis and thoracoabdominal aortic aneurysms, and if there is a history of artificial vessel or valve replacement, it would be better to avoid navigation of the catheter through the artificial vessel and valve. In the present case, the size of the thoracoabdominal aortic aneurysm gradually increased. Severe tortuosity of the thoracoabdominal aorta and type 3 arch caused by scoliosis were observed; thus, the transfemoral approach was considered critical, and the transbrachial approach was adopted.

In this patient, the diameter of the brachial artery was 4.7 mm, which was longer than the outer-diameter of a 9-Fr introducer sheath (outer diameter: 3.6 mm). It is reported that the diameter of the brachial artery is approximately 4.5 mm for men and 3.5 mm for women, and that it expands in association with coronary artery diseases.[8,9] Blood vessel diameter should be checked using ultrasound echo before the operation, and if inserting a 9-Fr introducer sheath is difficult, it should be considered to change to a smaller introducer sheath; however, this may restrict the use of BGC or a large-bore aspiration catheter.

Currently, there are several studies that MT via the transradial approach (TRA) is effective,[10,11] where the transfemoral approach is difficult due to anatomical problems. The advantage of the TRA is that hemostasis is easy; in contrast, the disadvantage is that the sheath size is limited, which restricts the use of the largest aspiration catheters as well as the usage of BGC. Throughout the MT procedure, to prevent distal migration of thrombi, proximal balloon protection using a BGC is commonly used. Beak et al.[12] and Zaidat et al.[13] have reported that the use of BGC was associated with higher recanalization success rates, shorter procedure times, and better clinical outcomes in MT. We adopted the transbrachial approach in MT as the first-line treatment, which allows the insertion of an 9-Fr introducer sheath and the use of BGC.

Conclusion

Endovascular MT and thrombolysis for AIS in MFS are extremely rare, and patients with MFS have often thoracoabdominal aortic aneurysms and multiple cardiovascular diseases; therefore, the choice of access route can be a problem during MT. MT via the transbrachial approach is a treatment option that can use BGC and a large-bore aspiration catheter.

Disclosure Statement

The authors declare that there are no conflicts of interest.

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