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【 CASE REPORT 】

Successful Surgical and Endovascular Multidisciplinary Therapy for Mid-aortic Syndrome with Complicated Atherosclerotic Comorbidities in an Older Patient

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Abstract:
Mid-aortic syndrome (MAS) is a rare vascular disorder that causes refractory hypertension. A 76-year-old woman was hospitalized for acute heart failure (HF) with drug-resistant hypertension; other comorbidities included epigastric artery rupture, old myocardial infarction, an intraventricular thrombus, and a cerebral artery aneurysm. Angiography revealed severe narrowing of the descending aorta, which led to the diagnosis of MAS. Although intensive medical treatment improved her HF, optimal blood pressure (BP) could not be achieved. Percutaneous coronary intervention and surgical bypass for diseased aorta was then performed in two stages, resulting in the achievement of optimal BP and alleviation of HF.

Key words: blood pressure, heart failure, mid-aortic syndrome, revascularization

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Introduction

Coarctation of the thoracic aorta at the level of the ligamentum arteriosum and occasionally in the aortic arch is well recognized (1). However, obstruction of the distal thoracic aorta, abdominal aorta, or both, collectively called mid-aortic syndrome (MAS), is much less common, accounting for only 0.5% to 2.0% of aortic coarctation cases. Cases of MAS are often found in childhood renovascular hypertension (2). Furthermore, MAS also induces various organ disorders due to severe and uncontrollable hypertension, including intermittent claudication, congestive heart failure, and renal insufficiency in childhood (3). MAS that develops in childhood is rarely diagnosed in the elderly, and limited cases have been reported previously.

We herein report an elderly case of MAS, who was successfully treated with surgical and endovascular multidisciplinary therapy.

Case Report

A 76-year-old woman with drug-resistant hypertension prescribed 3 kinds of antihypertensives by her primary care physician was admitted to our hospital for acute decompensated heart failure. Chest X-ray showed pulmonary congestion and cardiac enlargement. Echocardiography revealed left ventricular hypertrophy, akinesis of antero-septal wall and apical aneurysmal change with mural thrombus. She was treated with non-invasive positive-pressure ventilation, vasodilator and anticoagulation of vitamin K antagonist. Although her heart failure was improved, at hospital day 23, she suddenly complained of abdominal pain and entered a state of shock. Contrast-enhanced computed tomography (CT) showed a dilated and ruptured left epigastric artery (Fig. 1A, C), and narrowing of the thoracic descending aorta (Fig. 1B). Transcatheter arterial embolization of the left superior and inferior epigastric artery was performed, and she

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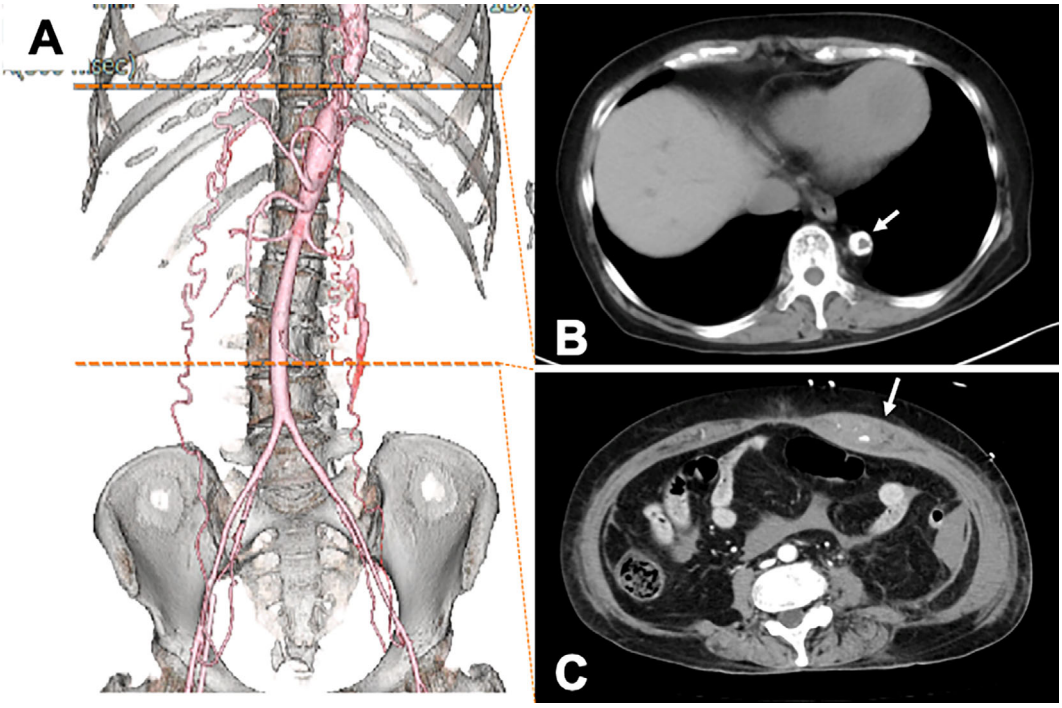

Figure 1. **(A) An overall view of 3D contrast-enhanced computed tomography. (B) Aorta at the level of stenosis with a luminal diameter of 6 mm and an external diameter of 15 mm (arrow). (C) Aorta distal of the dilated and ruptured left epigastric artery (arrow).**

recovered from the hemorrhagic shock. At hospital day 51, she was discharged after rehabilitation.

After hospital discharge, her blood pressure (BP) remained high despite the use of a calcium-channel blocker, beta blocker (BB), diuretics and angiotensin II receptor blocker (ARB). She complained of claudication and visited our hospital again. The bilateral ankle-brachial index (ABI) values were diminished to 0.51 on the right and 0.53 on the left. She was re-hospitalized for a further investigation.

On a physical examination, a BP of 168/62 mmHg and heart rate of 50/min were noted. A grade 3/6 ejection systolic murmur at the right sternal border and abdominal bruit were audible. Pulsation of the femoro-popliteal arteries was weakened bilaterally. An electrocardiogram showed left ventricular hypertrophy with an inverted T wave in leads V2 through V6. Chest X-ray revealed cardiomegaly with a cardiothoracic ratio of 60% and no pulmonary congestion. The laboratory data revealed a poor kidney function (serum creatinine of 1.31 mg/dL) and elevated n-terminal pro-brain natriuretic peptide (NT-pro BNP) level of 5,470.0 pg/mL. Other inflammatory, immunological and hormonal values, including serum renin, aldosterone and catecholamine, were within normal limits.

Cardiac magnetic resonance imaging (MRI) revealed transmural late gadolinium enhancement in the apex of antero-septal wall and thrombus in the left ventricular apex (Fig. 2A). Brain MRI revealed a 3-mm cerebral aneurysm. No inflammatory signs in the aorta were evident on positron emission tomography (PET)-CT (Fig. 2B). A coronary angiogram showed diffuse 90% narrowing of the left anterior descending (LAD) artery (Fig. 3A) and 75% narrowing of the left main (LM) trunk to the left circumflex (LCx) artery (Fig. 3B). An aortogram demonstrated 90% stenosis of the distal part of the thoracic descending aorta, and a waveform analysis using a pressure wire showed a pressure gradient of 79 mmHg (Fig. 3C) across the lesion. In addition, the minimal lumen diameter was 6 mm, and the outer diameter was 15 mm (Fig. 1B) on enhanced CT. This finding of significant stenosis of the lower thoracic aorta with no significant stenosis below the iliac artery suggested a diagnosis of MAS.

Based on the above findings, we determined that the hypertension induced by MAS was the primary cause of heart failure, old myocardial infarction with severe coronary artery disease, left ventricular thrombus, the rupture of epigastric artery and cerebral artery aneurysm. We therefore decided to perform revascularization of the coronary artery and aorta as treatment, with percutaneous coronary intervention (PCI) and surgical repair for the diseased aorta performed in two stages. Co-Cr everolimus-eluting stents were first percutaneously implanted in the left main bifurcation using culotte stenting technique (Fig. 3D, E), and one month after PCI, descending aorta-abdominal aorta bypass surgery was performed (Fig. 3F) with aspirin interrupted one week before and oral anticoagulant (OAC) stopped on the day before the surgery. The right lateral decubitus position was selected, and we reached the abdominal aorta via the peritoneal route after a paramedian incision and the descending aorta via fifth intercostal thoracotomy. The bypass was performed by using a 16-mm strait graft (J-graft Japan Lifeline, Tokyo, Ja-

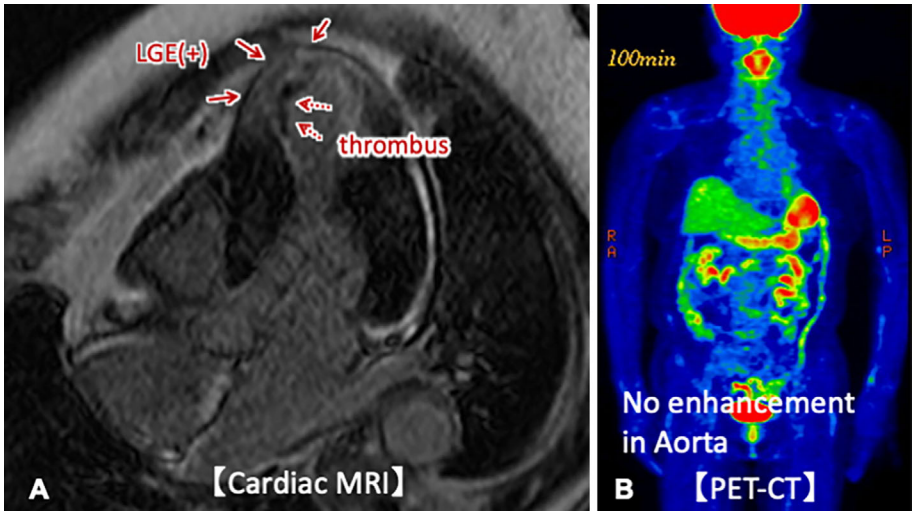

Figure 2. **(A) Cardiac magnetic resonance imaging (MRI) revealed transmural late gadolinium enhancement at the apex of the antero-septal wall and thrombus in the left ventricular apex. (B) There was no enhancement in the aorta on PET-CT. MRI: magnetic resonance imaging, LGE: late gadolinium enhancement, PET-CT: positron emission tomography computed tomography**

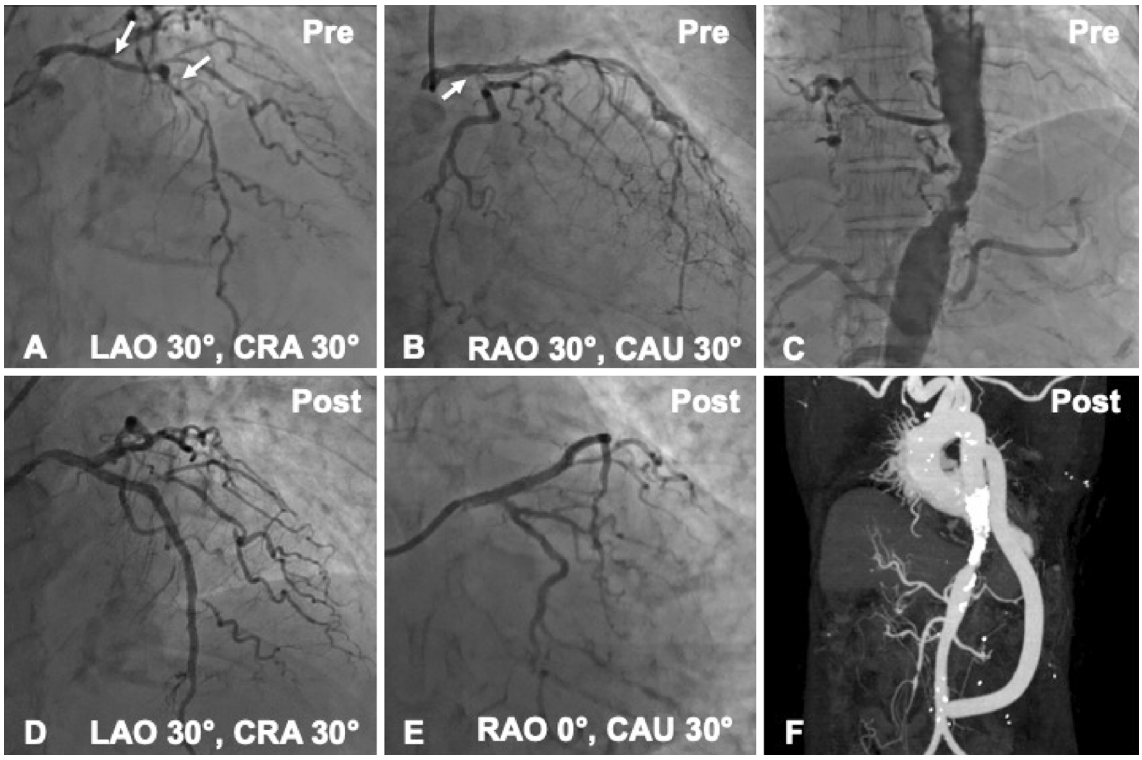

Figure 3. **(A) A preoperative coronary angiogram (CAG) of the LAO 30° and CRA 30° view showing diffuse 90% narrowing of the LAD (arrow). (B) A preoperative CAG of the RAO 30° and CAU 30° view showing 90% narrowing of just proximal of the LCx (arrow). (C) A preoperative aortagram showing marked stenosis in the descending aorta. (D, E) A post-culotte stenting technique CAG revealing good dilation of both the LAD and LCx. (F) Postoperative CT of descending aorta-abdominal aorta bypass surgery. CAU: caudal, CRA: cranial, CT: computed tomography, LAD: left ascending descending artery, LAO: left anterior oblique, LCx: left circumflex artery, RAO: right anterior oblique**

pan) with an F-F bypass for extracorporeal circulation.

The surgery was completed without any complications, and she was discharged on the 13th post-surgical day. Immediately after this surgery, her BP was stabilized from 150 to 120 mmHg, and her antihypertensive drug prescriptions were reduced from the maximum dose of 5 agents to the

standard dose of 2. Serum creatinine and NT-pro BNP levels were improved to 1.00 mg/dL and 300 pg/mL, respectively. In addition, the ABI also was improved to 1.19 on the right and 1.05 on the left after surgery.

Discussion

MAS is a rare vascular disorder consisting of narrowing or stenosis of the distal thoracic or abdominal aorta, accounting for 0.5-2% of all cases of aortic narrowing (4). The mean age of a diagnosis with MAS is reportedly 14.3 years old (range 19 days to 49 years old) (4), and a previous review suggested that 55% of untreated patients died at a mean age of 34 years old (5). Although MAS is an important cause of renovascular hypertension in children and adolescents, elderly cases have been infrequently reported.

The majority of cases of MAS are idiopathic (61%), with the pathogenesis of the disease unknown. Aortitis (26%), atherosclerosis (5%), neurocutaneous syndrome (5%), Williams syndrome (2%) and others (1%) are classified as secondary causes of MAS (4). Older cases of MAS may be mainly caused by aortitis or atherosclerosis, whereas idiopathic MAS is usually detected during childhood. Given the small outer diameter and lack of inflammatory signs of a diseased aorta, despite the patient's advanced age, the present case was considered idiopathic MAS. It was thought that MAS was originally present in this patient, but the progress of arteriosclerosis due to aging reflected the current condition.

MAS is often incidentally found in drug-resistant hypertension, and other presenting features include symptoms of the cardiovascular (heart failure, palpitations, heart murmur) and central nervous (headache, hypertensive encephalopathy and cerebrovascular accident) systems (6). However, to our knowledge, no MAS cases presenting various complications as those seen in our case have been reported.

Regarding the treatment of MAS, surgical operation or endovascular treatment is indicated in cases that are drug treatment-resistant. There have been changes in the operative strategy for MAS over the decades (7). Many repair techniques have been utilized (8), and surgical therapy has been associated with a low risk of recurrence of coarctation (9-11). Although anatomical repair, such as end-to-end bypass or patch-angioplasty, is the most common procedure for managing coarctation of the aorta (1), aortic extra-anatomical bypass is also a common procedure in MAS (7, 9). However, significant developments in endovascular therapy have been achieved, and there are fewer complications in the acute phase than with surgical repair (12). Nevertheless, the re-intervention rate remains high, and the long-term outcomes have yet to be elucidated (12, 13). It thus remains unclear which procedure is the better option for revascularization in MAS.

In the present case, some intercostal arteries arose from the stenotic segment of the aorta and would require reconstruction should we select anatomical bypass, and reattachment seemed difficult because of the vessel calcification. Furthermore, anatomical bypass via the thoracoabdominal aorta approach requiring a large dissection area and a long surgical time would be highly invasive compared to extra-anatomical bypass surgery, which only requires dissection of the anastomotic areas. Regarding endovascular stent grafting, based on the CT findings of heavy calcification and a small external diameter of the aorta, in the present case, endovascular therapy might not have obtained sufficient luminal enlargement, and serious concerns remained regarding spinal ischemia as well. Thus, extra-anatomical bypass surgery was selected for MAS in our patient.

Regarding the coronary revascularization, we considered performing coronary artery bypass and ascending aorta-abdominal bypass surgery simultaneously. However, neither of the internal thoracic arteries of this patient were suitable for CABG, as they were important collateral sources of the lower extremities, and the left internal thoracic artery had been embolized for occlusion of the epigastric artery beforehand. We therefore chose PCI for coronary revascularization. Our initial plan with PCI was to implant a stent only in the mid-segment of the LAD, as the major purpose of PCI was to ensure that subsequent aortic surgery was safe. However, given the huge plaque burden in the LM bifurcation on intravascular ultrasound (IVUS), we decided to also treat the LM bifurcation lesion. The access site for mechanical support in case of an emergency was a matter of concern, so we requested a cardiac surgeon be present as on-site back up before starting this procedure.

In conclusion, we encountered a rare case of MAS with severe complications that was successfully treated with surgical and endovascular combination therapy, achieving optimal BP and improvement of the renal function and heart failure. Our case suggests that resistant hypertensive patients presenting with several hypertensive organ damage might have MAS and require not only antihypertensives but also multidisciplinary invasive treatment.

Author's disclosure of potential Conflicts of Interest (COI).

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