

[ CASE REPORT ]

# Atypical Aortic Coarctation in a Patient with an Acute Exacerbation of Multiple Organ Failure: Successful Endovascular Therapy and Spontaneous Retroperitoneal Bleeding

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## Abstract:

We experienced a case of acute multiple organ ischemia and multiple organ failure due to atypical aortic coarctation (AAC). Since the patient's hemodynamics were too unstable to perform surgical revascularization, we performed urgent endovascular therapy (EVT) with a stent. Eventually, the patient achieved remission from multiple organ failure and a satisfactory clinical outcome. We feel that EVT for AAC is a sufficiently effective treatment option if the purpose of EVT is to save a patient's life in the acute phase. In the present case, spontaneous retroperitoneal bleeding (SRB) occurred after EVT of AAC, but this is a rare incident, although noteworthy in the clinical course.

**Key words:** atypical aortic coarctation, atherosclerosis, stent, secondary hypertension, multiple organ ischemia, spontaneous retroperitoneal bleeding

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## Introduction

Atypical aortic coarctation (AAC) is known to be associated with Takayasu arteritis (TA), atherosclerosis, congenital vascular abnormality, and fibromuscular dysplasia. Most cases are caused by TA (1, 2). When AAC develops anywhere other than the ascending aorta, its signs include secondary hypertension of the upper half of the body and hypotension of the organs anatomically lower than the stenotic lesion, with associated consequences (e.g., intestinal ischemia, ischemic nephropathy, intermittent claudication) (3).

Occasionally, acute exacerbation of heart failure due to afterload mismatch occurs, and multiple organ failure is induced if the stenosis of AAC is sufficiently severe. Surgical revascularization is often performed to treat AAC. However, there have been a few reports of endovascular therapy (EVT) with good clinical outcomes obtained (4).

We experienced a case of atherosclerotic AAC in which an elderly woman developed exacerbation of heart failure and multiple organ failure. We successfully achieved bailout

with EVT in the acute phase by implanting a stent in the severe stenotic lesion of AAC.

However, spontaneous retroperitoneal bleeding (SRB) after EVT occurred, hampering the treatment procedure.

EVT for AAC is particularly effective for saving a patient's life in the acute phase.

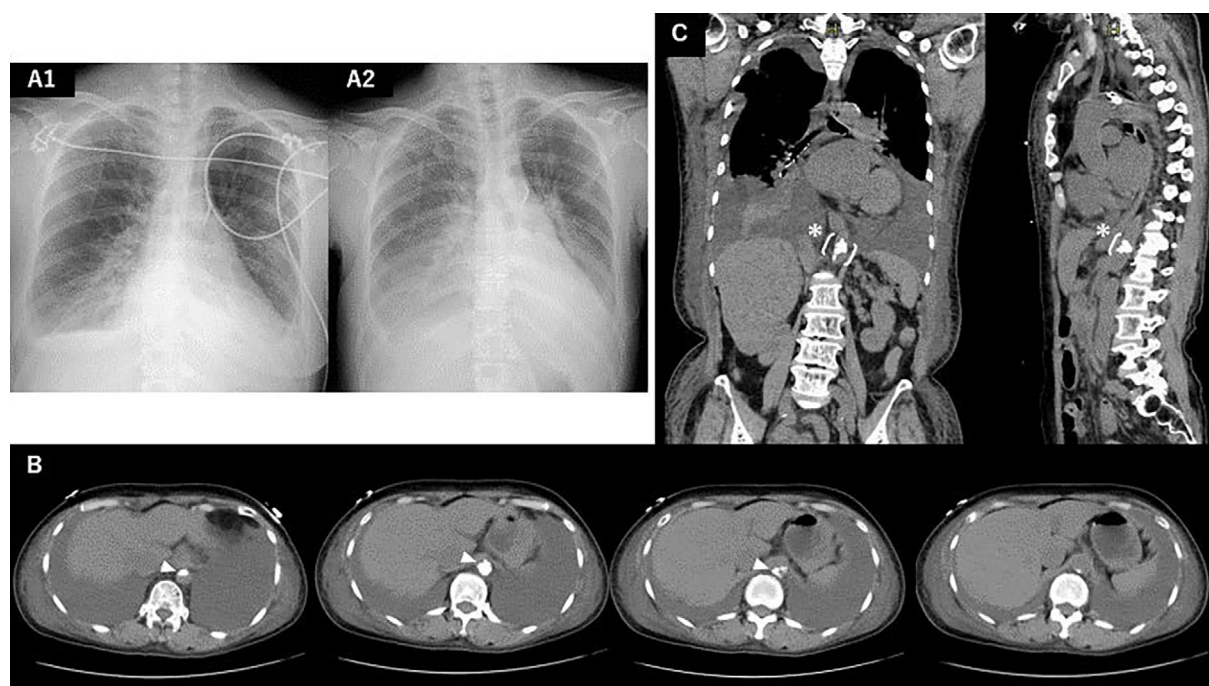
In addition, we feel that retroperitoneal bleeding should be recognized as a potential complication of stent implantation.

We herein report a rare case in which EVT for AAC was quite effective and highlight the possibility of retroperitoneal bleeding occurring after revascularization.

## Case Report

A 74-year-old woman was transported to our hospital by ambulance with presenting complaints of anasarca and dyspnea.

At 63 years old, she started taking oral medication (calcium channel blocker, angiotensin receptor blocker, and diuretic) for hypertension prescribed by a local hospital.



**Figure 1.** A1: Chest X-ray on admission. A2: Chest X-ray on day 8. B, C: Plain CT scan showing the aorta by axial cross-section (B) and coronal cross-section (C). There was severe calcification in the lesion. Part of the nodular calcification protruded into the aortic lumen, and there was calcification along the entire circumference of the lesion (B: white arrowheads). In addition, the lesion was very focal, with a length of about 20 mm (C: \*).

However, despite these treatments, her hypertension was difficult to control, and her systolic blood pressure remained between 160 and 180 mmHg.

She suffered from lower extremity edema two weeks before admission and was additionally prescribed other diuretics by a local doctor, but still there was no improvement. After that, her dyspnea began to manifest and gradually became exacerbated, so she was transferred to our hospital.

On admission, she was conscious, the blood pressure in the upper limbs was 160/52 mmHg, the heart rate was 78/min, and the oxygen saturation as indicated by A pulse oximeter was 91% under nasal oxygen flowing at a rate of 2 L/min.

There was no marked difference in the blood pressure between the left and right upper limbs. Facial edema, jugular dilation, and pitting edema in the lower limbs were observed. We heard coarse crackles in both lower lung fields. We also detected the vascular murmur of the abdominal aorta and could not feel the pulse of either femoral artery. Chest X-ray revealed a dilated heart (Cardio thoracic ratio=61.1%), bilateral pleural effusion, and pulmonary congestion (Fig. 1 -A1). An electrocardiogram showed mild ST depression in V4 to V6.

Hematological findings showed mild anemia (Hb 10.5 g/dL) and elevation of serum brain natriuretic peptide (BNP) (1,313.7 pg/mL), Cr (3.95 mg/dL), liver enzyme [aspartate transaminase (AST) 127 U/L, alanine aminotransferase (ALT) 100 U/L] and CRP (5.89 mg/dL) levels with a reduction in serum Na (122 mEq/L) and Cl (81 mEq/L) levels

and a normal serum K (4.3 mEq/L) level.

Echocardiography revealed good contractility of the left ventricle (ejection fraction 65%) and mild diastolic dysfunction. There was no obvious myocardial hypertrophy of the left ventricle.

The right ventricular systolic pressure was increased (estimated at 50 mmHg). Given these findings, we diagnosed her with acute exacerbation of chronic heart failure and started in-patient medical treatment.

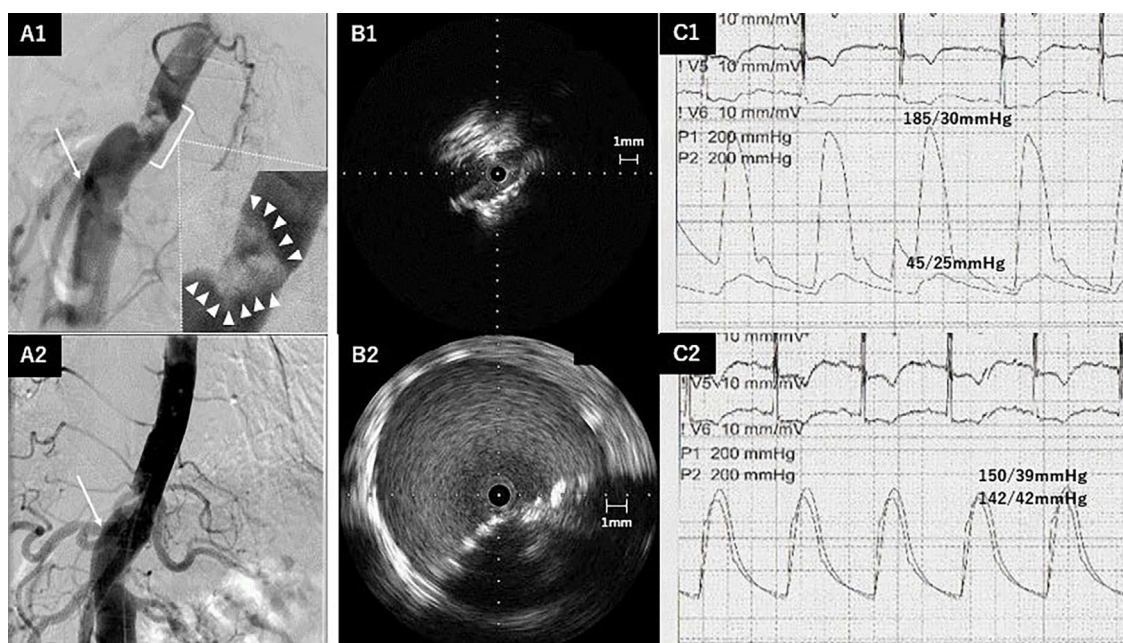
From admission (day 1), we administered carperitide (0.03  $\gamma$ ) and furosemide (20 mg) for diuresis, and her edema and dyspnea showed an improving trend.

However, her systolic blood pressure remained between 180 and 240 mmHg, so we increased the dose of carperitide gradually day by day.

From day 5 of hospitalization, her urine output decreased significantly, and her respiratory status deteriorated.

The findings of pleural effusion and pulmonary edema on chest X-ray were exacerbated, so we added continuous intravenous administration of nitroglycerin to correct afterload mismatch. In addition, we administered noninvasive positive-pressure ventilation therapy at the same time. Her urine output showed no further increase after that, and her systolic blood pressure remained 180 mmHg.

However, this improvement was not satisfactory, so we added continuous intravenous administration of nicardipine hydrochloride. The systolic blood pressure finally improved to 140-160 mmHg with the combination of 3 drugs. We also administered tolvaptan (7.5 mg/day), but she became anuric



**Figure 2.** A1: Angiography findings before endovascular therapy (EVT). There was a large nodular calcification (white arrowheads) protruding into the aortic lumen that made the lumen narrow. The celiac artery branched off from the distal side of the lesion (white arrow). A2: Angiography findings after EVT. Obvious enlargement of the lumen was seen compared to before EVT. B1: Intravenous ultrasound (IVUS) findings before EVT. IVUS showed a severe stenotic lesion with a minimum lumen diameter (MLD) of 1.49 mm and minimum lumen area (MLA) of 4.3 mm<sup>2</sup>. B2: IVUS findings after EVT. The lumen of the lesion was expanded to an MSA of 65.6 mm<sup>2</sup> by implanting the stent. The stent of the lesion had stretched the healthy wall opposite the area of calcification of the target lesion. C1: Pressure pulse wave of the right brachial artery sheath and left femoral artery sheath before EVT. A marked pressure gradient was found, and the left femoral artery pressure was extremely low. C2: Pressure pulse wave of the right brachial artery sheath and left femoral artery sheath after EVT. The pressure gradient had almost disappeared.

with exacerbation of generalized edema and dyspnea.

On day 7, the hematological findings showed that the liver enzyme levels had further increased [AST 481 U/L, ALT 353 U/L, lactate dehydrogenase (LDH) 1,005 U/L], and plasma renin activity was also high (23.2 ng/mL/h).

On the same day, plain CT revealed that the pleural effusion had significantly increased compared to that found upon admission, and there were no morphological abnormalities of the kidney or urinary tract that could cause anuria. However, we did detect on CT a calcified lesion in the part of the aorta at the thoracoabdominal transition, suggesting occlusion or severe stenosis (Fig. 1-B).

The stenotic lesion was just above the celiac artery, and the lesion length was approximately 20 mm (Fig. 1-C), indicating a very focal lesion. There was no stenosis or calcified lesion in any other parts of the aorta or other branched arteries.

Considering our inability to palpate the pulsation of either femoral artery and the above CT findings, we suspected the obstruction of blood flow was due to an AAC-induced organ perfusion disorder in the distal part of the lesion that had impaired the blood flow to multiple sites, including the kidney, liver, and lower limbs.

Since it was difficult to maintain oxygenation, continuous hemodiafiltration (CHDF) was performed for volume reduction on day 7.

Her generalized edema and respiratory status were improved after CHDF was started, but laboratory data showed that the levels of liver enzymes and LDH were further elevated on day 8 (AST 1,892 U/L, ALT 1,371 U/L, LDH 1,724 U/L).

She also complained of abdominal pain that suggested intestinal ischemia.

Dehydration via CHDF improved her respiratory status, but her findings on chest X-ray were not improved (Fig. 1-A2). Furthermore, the decreased intravascular volume due to CHDF clearly exacerbated her multiple organ ischemia, suggesting that saving her life through pharmacological treatment, CHDF, and noninvasive positive-pressure ventilation therapy would be difficult.

Recognizing that her general condition was too poor to tolerate surgical treatment, we performed EVT urgently on day 8 (Fig. 2). We performed EVT via the bidirectional approach using the right brachial artery and left femoral artery. When we inserted the sheath, the right brachial artery pressure was 183/30 mmHg, and the left femoral artery pressure

was 45/25 mmHg. The initial pressure gradients of the right brachial artery and left femoral artery were remarkable (Fig. 2-C1).

It was easy to pass the wire through the lesion, and we performed intravascular ultrasound (IVUS), which revealed an exceedingly focal lesion whose minimum lumen distance (MLD) was 1.49 mm and minimum lumen area (MLA) 4.3 mm<sup>2</sup>, with thick calcified plaque involvement ranging from 180° to 270° (Fig. 2-B1).

We inflated a noncompliant balloon YOROI 6.0/40 mm (Kaneka Medix, Osaka, Japan) with 20 atm at the lesion. The femoral artery pressure increased just after inflation but then immediately dropped, so the improvement was poor.

We then selected a larger balloon (MUSTANG 10.0/40 mm; Boston Scientific, Marlborough, USA) and dilated the lesion again; however, as before, the pressure improvement disappeared immediately after inflation.

IVUS showed that the expandability of the lesion was also insufficient, so we concluded that the inflation of the balloon had expanded only toward the healthy side of the vessel with the lesion and then recoiled immediately.

We therefore decided to implant a stent to restore the vessel lumen, and an EPIC stent 12.0/40 mm (Boston Scientific) was placed at the lesion.

There was some concern that a self-expandable stent might fall off during placement because the aorta has a large diameter and a high flow velocity. We therefore performed rendezvous and externalization with a 300-cm treasure floppy guidewire (Asahi Intecc, Nagoya, Japan) via the left femoral artery to the 6-Fr guiding catheter at the right brachial artery, implanting the EPIC stent through contrast guidance.

After stenting, IVUS showed that the expansion of the stent was insufficient, so we changed the balloon from a MUSTANG 10.0/40 mm to a MUSTANG 12.0/40 mm (Boston Scientific) and performed inflation at 14 atm at in-stent to gain a larger lumen.

Consequently, the right brachial artery pressure was 150/39 mmHg, and the left femoral artery pressure was 142/42 mmHg. The pressure gradient between the right brachial artery and the left femoral artery almost disappeared (Fig. 2-C2).

IVUS revealed that the minimum stent area (MSA) post-EVT had improved to 65.6 mm<sup>2</sup> from 4.3 mm<sup>2</sup> pre-EVT (Fig. 2-B2).

Because the pressure gradient had improved and acceptable expansion of the AAC lesion had been obtained, we terminated the EVT procedure.

Urine output began to be observed after EVT, and on the same day, the total urine volume was 1,150 mL, so we were also able to reduce the dehydration volume of CHDF.

She was able to be withdrawn from noninvasive positive-pressure ventilation therapy on day 10 and from CHDF on day 11, as her urine output had sufficiently increased.

However, she complained of low back pain at dawn on day 12 and went into shock, so we performed emergency

contrast CT, which revealed a huge hematoma at the right retroperitoneal area, opposite from the puncture performed during EVT. In addition, multiple points of active hemorrhaging were found from an unidentifiable small peripheral lumbar artery (Fig. 3A).

After consulting with the radiologist, we concluded that intravascular treatment would be difficult because the bleeding points from a very thin peripheral artery were numerous and extensive, which might necessitate a wide embolization range if endovascular embolization were performed.

Therefore, we selected conservative treatment instead.

We discontinued the heparin-based anticoagulant therapy because she had been successfully withdrawn from CHDF and performed a blood transfusion appropriately.

After several days of transfusions of erythrocytes and fresh-frozen plasma, the hematoma showed no increase on plain CT on day 18.

On day 32, contrast CT was repeated, revealing no bleeding from any lumbar artery, and the hematomas had begun to shrink.

Finally, the renal and liver functions normalized (Fig. 4), and the serum BNP level improved to 72.4 pg/mL on day 39.

The serum renin activity was also normalized, and blood pressure control was good without increasing the dose of antihypertensive drugs (Fig. 5).

Two weeks after her discharge from the hospital, CT findings showed that the retroperitoneal hematoma had been further reduced (Fig. 3B), and the celiac artery was not occluded by the stent at aorta (Fig. 6). In addition, 18-fluorodeoxyglucose-positron emission tomography (<sup>18</sup>F-FDG PET) showed that there was no abnormal accumulation at the stent implantation site, other aorta sites, or their branches (Fig. 3C).

## Discussion

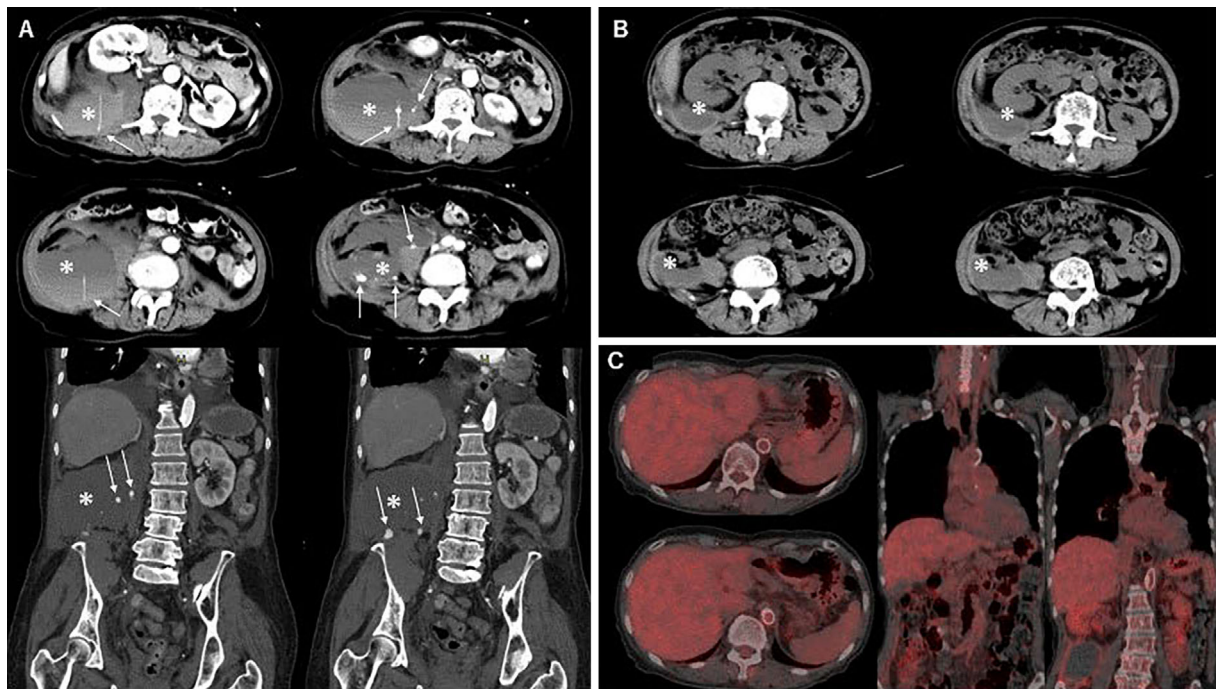
In the present case, we saved the patient's life by EVT for AAC that had caused refractory hypertension, acute heart failure, and multiple organ failure.

Most cases of AAC are known to be associated with TA, but other causes, such as arteriosclerosis, congenital vascular abnormalities, and fibromuscular dysplasia, have also been reported (1, 2).

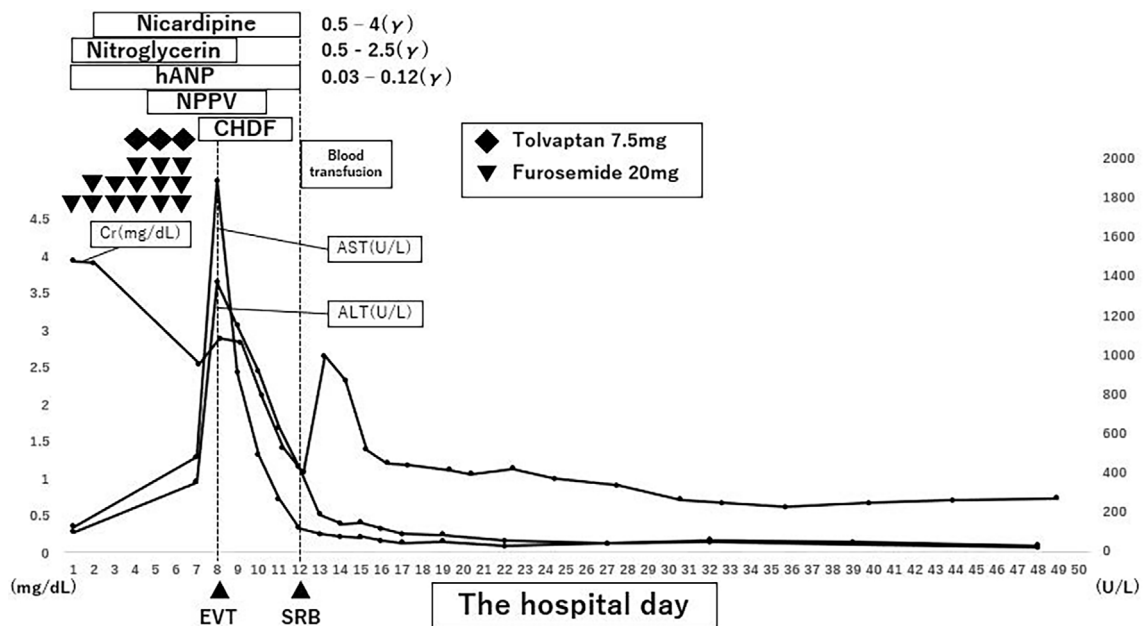
In the present case, AAC was most likely due to atherosclerosis, as the patient had no history of suspected chronic inflammation, and there were few findings suggesting persistent inflammation according to the laboratory data after the improvement of the pathological condition.

Furthermore, the imaging findings showed a focal lesion in the thoracoabdominal aortic transition, and no lesions were found in the major branch arteries, such as the carotid artery, subclavian artery, or renal artery.

<sup>18</sup>F-FDG PET performed after discharge from the hospital showed no abnormal accumulation at the target lesion or other parts of the aorta, including branches.



**Figure 3.** A: Contrast CT revealed retroperitoneal hemorrhaging with multiple bleeding points (white arrows) and a huge hematoma (\*). B: Plain CT performed two weeks after discharge showed a reduction in the retroperitoneal hematoma (\*). C:  $^{18}\text{F}$ -FDG PET showed no abnormal accumulation at the stent implantation site, other aorta sites, or branches.



**Figure 4.** Clinical course. EVT: endovascular therapy, SRB: spontaneous retroperitoneal bleeding

Of note, the present patient was 74 years old, which is markedly older than the typical patient with TA (<40 years old).

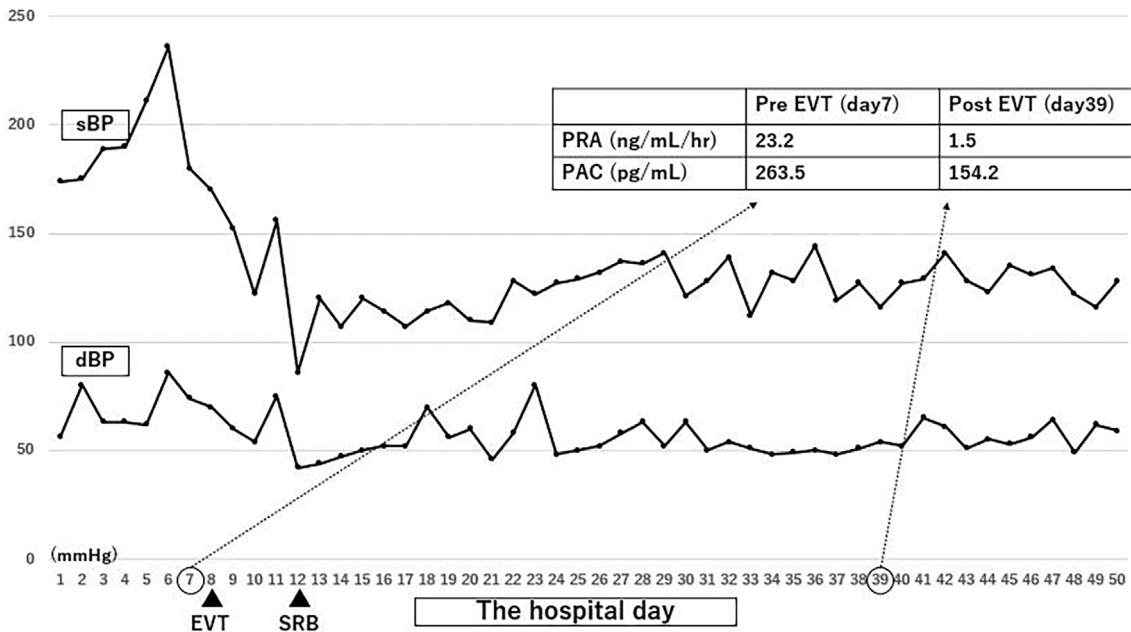
The above findings and clinical course did not suggest TA, so we feel it is highly possible that atherosclerosis caused the AAC in this case.

However, in TA, once the inflammation invades the intima, edematous changes occur, resulting in the infiltration of lipids and blood cells. Eventually, these inflammatory

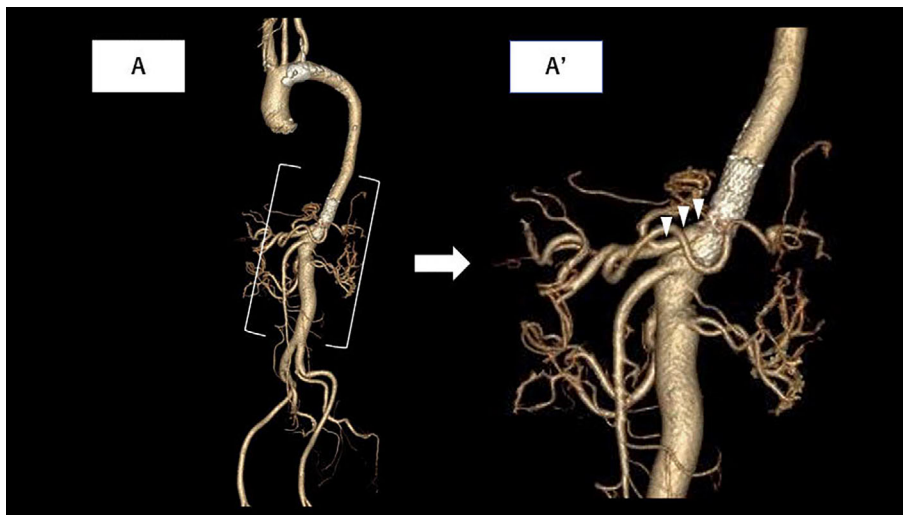
changes cause atherosclerosis. Calcification in particular is found over five years after the onset of the disease (5).

Numano et al. (6) reported difficulty distinguishing the scarring stage of TA from atherosclerosis, and pathologically, rupture, and fibrosis of intimal elastic fibers as well as fibrous thickening of the adventitia and characteristic cellular infiltration are found in TA.

Differentiating TA from atherosclerosis is thus generally recognized as difficult, so we cannot completely deny the



**Figure 5.** Changes in blood pressure, plasma renin activity (PRA), and plasma aldosterone concentration (PAC) during hospitalization.



**Figure 6.** Contrast CT revealed that the celiac artery was jailed by the EPIC stent, but there was no stenosis at the ostium of the celiac artery (white arrowheads).

involvement of TA in this case.

Regarding the treatment of AAC, many reports have described the efficacy of surgical revascularization (1, 3). Even in critical situations, surgical treatment is often chosen.

Major surgical treatments include aortocervical bypass, cervicosubclavian bypass, aortic replacement, aortocoronary bypass, replacement of aortic aneurysm, aorto-aortic bypass, aortorenal bypass, reconstruction of a renal vessel, and nephrotomy (5).

However, even among reports focusing on TA, there have been studies examining the utility of EVT (5, 7, 8).

Regarding the revascularization of TA, previous reports mentioned that, whether by surgical treatment or EVT, the high rate of restenosis is a serious problem, and there are

very few cases of EVT being performed for aorta lesions, as in this case. In addition, there have been few reports of EVT being performed for AAC caused by atherosclerosis.

In the present case, the perfusion pressure of organs distal to the stenosis site of the AAC was reduced due to antihypertensive treatment. Furthermore, the volume reduction due to diuretics and CHDF adversely affected organ perfusion, which exacerbated her condition.

The abnormal elevation of the serum renin activity also suggested a decrease in the renal blood flow.

Regarding treatment methods, the present patient was suffering from exacerbation of heart failure, and liver disorder, renal disorder, and intestinal ischemia were also present; her general condition was thus extremely poor, making it diffi-

cult to perform invasive surgical treatment. We therefore had no choice but to perform EVT which is a low-invasive strategy.

Unlike the above reports, however, it is unlikely that the AAC in this case was caused by TA, as the target lesion was in the aorta rather than the renal artery. Given that the patient's general condition was too poor to perform invasive surgical treatment was difficult, we opted to perform EVT as a more feasible treatment strategy.

However, there have been few reports of EVT for atherosclerotic AAC, so the utility and safety of this approach are poorly understood.

As mentioned above concerning TA, we should consider the possibility that restenosis may occur in the chronic stage.

If restenosis does occur, performing aortic replacement or aorto-aortic bypass may be appropriate provided the general condition is stable enough to support such approaches.

Of note, from the perspective of minimal invasiveness, several reports have described the efficacy of axillofemoral bypass (3).

Four days after EVT, sudden bleeding from multiple points at peripheral points of the lumbar artery occurred, which caused hemorrhagic shock. This was a significant clinical occurrence, and we selected a conservative treatment method to manage the retroperitoneal bleeding. Transcatheter embolization has been suggested to be effective for managing massive retroperitoneal hemorrhaging (9), even in cases with multiple bleeding points, as in the present patient (10).

Fortunately, we were able to save our patient's life without having to deal with abdominal compartment syndrome, which can be caused by massive retroperitoneal hematomas. However, substantial blood transfusion was required, so we should have considered catheter embolization.

The cause of the bleeding from multiple points at the periphery of the lumbar artery in this case is unclear.

The site of retroperitoneal hemorrhaging was opposite the sheath insertion point. Furthermore, it is unlikely that the EVT procedure caused multiple perforations at peripheral portions of the lumbar artery.

Furthermore, there was an interval of four days between the EVT procedure and the state of shock that occurred after, so we do not believe that the retroperitoneal hemorrhaging was a complication of EVT.

We considered several potential causes of retroperitoneal hemorrhaging in our case. For example, the distal part of the AAC was experiencing low-pressure hemodynamics due to stenosis of the aorta. However, treatment by dilating the aorta through EVT suddenly induced high-pressure hemodynamics from the previous low-pressure hemodynamic state. This sudden change in the circulatory system from low pressure to high pressure may have caused a collapse in the peripheral region of the lumbar artery.

Several mechanisms have been proposed as underlying the pathogenesis of SRB.

In previous reports, it was suggested that forceful muscular strain might be one such mechanism, as sites of bleeding were mostly intramuscular, occurring in the posterior region of the iliopsoas or gluteal muscles (10).

In particular, limitations were placed on changing the patient's position, with the patient required to maintain a supine position for a long period during treatment with CHDF; this might have resulted in the compression of the posterior side of the involved muscle as a possible mechanism underlying the blood vessel rupture in SRB (11).

In the present case as well, the patient was forced to remain in the supine position for a long time due to CHDF, and we administered anticoagulants during CHDF. This treatment may have caused the SRB in our patient.

Generally, the prognosis of TA is poor, and a high rate of restenosis is recognized after revascularization by EVT and surgical treatment.

However, the AAC in this case was believed to have been caused by atherosclerosis, and we were able to save the patient's life by performing EVT in the acute phase.

In addition, the patient's hepatic and renal disorders were completely resolved, and the plasma renin activity was also decreased to a normal range, so the patient achieved good blood pressure control.

Our patient thus showed a good outcome after EVT for AAC.

When it is difficult or impossible to perform surgical treatment, as was the case here, EVT to save the patient's life may be an effective alternative method of treating AAC.

However, we should consider the risk of restenosis in the chronic phase and continue monitoring the lesion carefully.

The fact that SRB occurred after EVT is an interesting point in the present clinical course. While we cannot firmly conclude the cause of SRB, we cannot deny the possibility that rapid changes in arterial pressure due to revascularization by EVT had a role in the etiology.

In cases such as the present one, it is necessary to keep in mind the possibility of SRB occurring; as such, we should follow the postoperative course carefully.

### Conclusion

We were able to save the present patient's life by performing emergent EVT for AAC, which seemed to have been caused by atherosclerosis.

The high rate of restenosis when EVT is performed in TA patients is a known issue, and restenosis is a critical problem in the clinical course. However, in cases when atherosclerotic AAC is suspected, EVT may be an effective alternative for revascularization that can save a patient's life.

Furthermore, we failed to clearly determine the etiology of SRB. However, rapid changes in the arterial pressure might trigger SRB.

**The authors state that they have no Conflict of Interest (COI).**

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