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【 CASE REPORT 】

Difficulty Diagnosing Retrograde Type A Aortic Dissection with Intramural Hematoma and Risk of Re-dissection and Rupture: A Report of Two Cases

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Abstract:
Acute type A aortic dissection is a potentially fatal disease, and emergency surgery should be considered when it is diagnosed. We herein report two cases of retrograde type A aortic dissection with intramural hematoma, followed by re-dissection, rupture, and cardiac tamponade. The diagnoses in these cases had to be made carefully, as the false lumen of the ascending aorta was sometimes unclear on contrast-enhanced computed tomography.

Key words: retrograde type A aortic dissection, ulcer-like projection, cardiac tamponade, contrast-enhanced CT

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Introduction

Acute type A aortic dissection is a potentially fatal disease, and it is very important to decide whether to perform emergency surgery or follow-up with conservative treatment. When deciding on a treatment policy, the results of contrast-enhanced computed tomography (CT) are considered important in daily clinical practice.

We encountered two cases of retrograde type A aortic dissection (retrograde TAAD) with intramural hematoma (IMH). These cases were difficult to diagnose because the false lumen was sometimes unclear in the ascending aorta on contrast-enhanced CT.

Case Reports

Case 1

A 63-year-old woman with no remarkable medical history was transferred to our hospital for persistent chest pain. A physical examination revealed a heart rate of 92 bpm and blood pressure of 91/64 mmHg. A 12-lead electrocardiogram showed no ST-T changes. Echocardiography revealed mild pericardial effusion. The laboratory data showed an elevated white blood cell count (9630/μL), but the D-dimer level was within the normal range (0.6 μg/mL). Emergent contrast-enhanced CT showed pericardial effusion but no apparent dissection in the aorta. The diameter of the ascending aorta was 41×41 mm (Fig. 1A-C). The patient was hospitalized and placed under observation. Her blood pressure was maintained below 120 mmHg. CT performed on the second day of hospitalization showed no change in the findings of the ascending aorta (Fig. 1D). On the third day of admission, the patient suddenly went into cardiopulmonary arrest and died.

A pathological autopsy showed the entry of dissection at the descending aorta of the arch, dissection of the ascending/descending aorta, and perforation of the pericardial cavity from behind the ascending aorta (Fig. 2). The pericardial cavity was perforated from the dorsal side of the ascending aorta and further into the left thoracic cavity from the left

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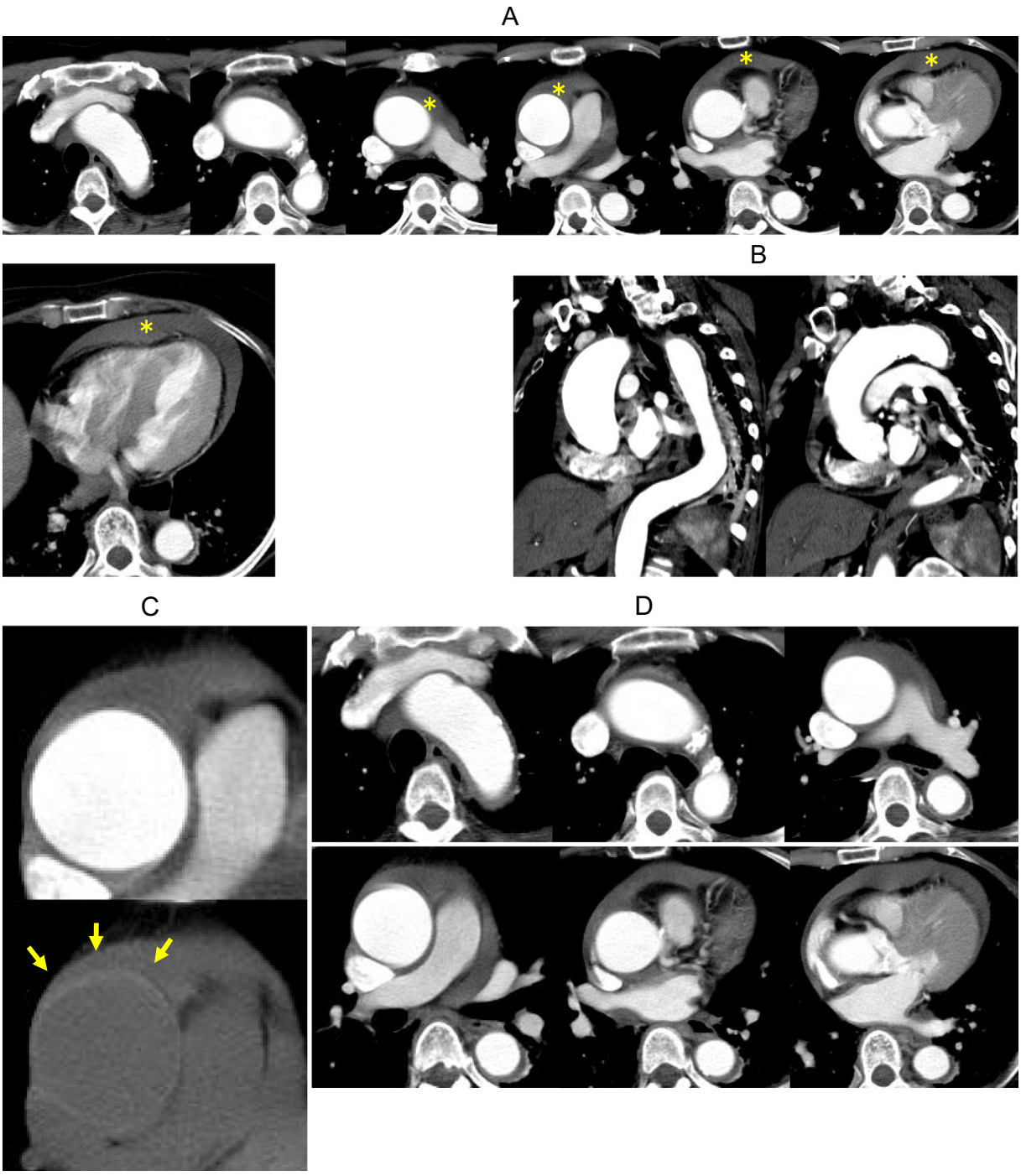

Figure 1. **Contrast-enhanced CT in case 1 (A: horizontal, B: sagittal). Pericardial effusion (*) was found, but no apparent dissection or ULP in the aorta was observed. A thin dense layer of tissue was observed completely encircling the ascending aorta (A, yellow arrows) in plane CT (C). Such findings were not observed on contrast-enhanced CT. CT performed on the second day of hospitalization showed no change in the findings of the ascending aorta (D).**

side of the pericardium.

Case 2

A 65-year-old man with essential hypertension was transferred to our hospital for chest and back pain. A physical examination revealed a blood pressure of 210/100 mmHg. A 12-lead electrocardiogram showed no ST-T changes. Echocardiography revealed mild pericardial effusion. Emergent contrast-enhanced CT showed pericardial effusion and IMH with Ulcer like projection (ULP) in the descending aorta of the arch but no apparent dissection in the ascending aorta (Fig. 3). The diameter of the ascending aorta was 37×41 mm. Echocardiography 30 minutes after CT in the emergency outpatient department showed an increase in pericardial effusion, and rupture of the TAAD was suspected. He was transferred to another hospital for cardiovascular sur-

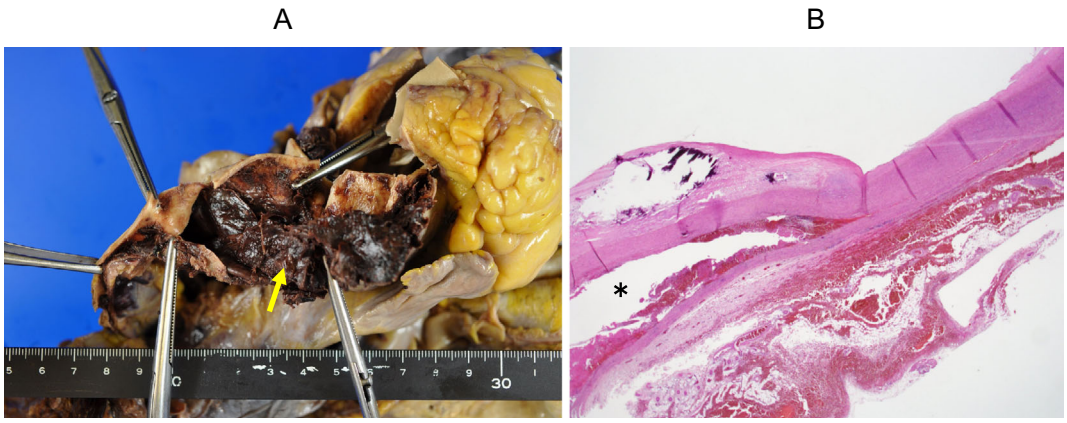

Figure 2. **Findings of the pathological autopsy in case 1. A: Resected specimen of the ascending aorta. The pseudolumen and IMH were confirmed (arrow). B: Microscopic findings of the ascending aorta. Dissociation in the medium was confirmed (*).**

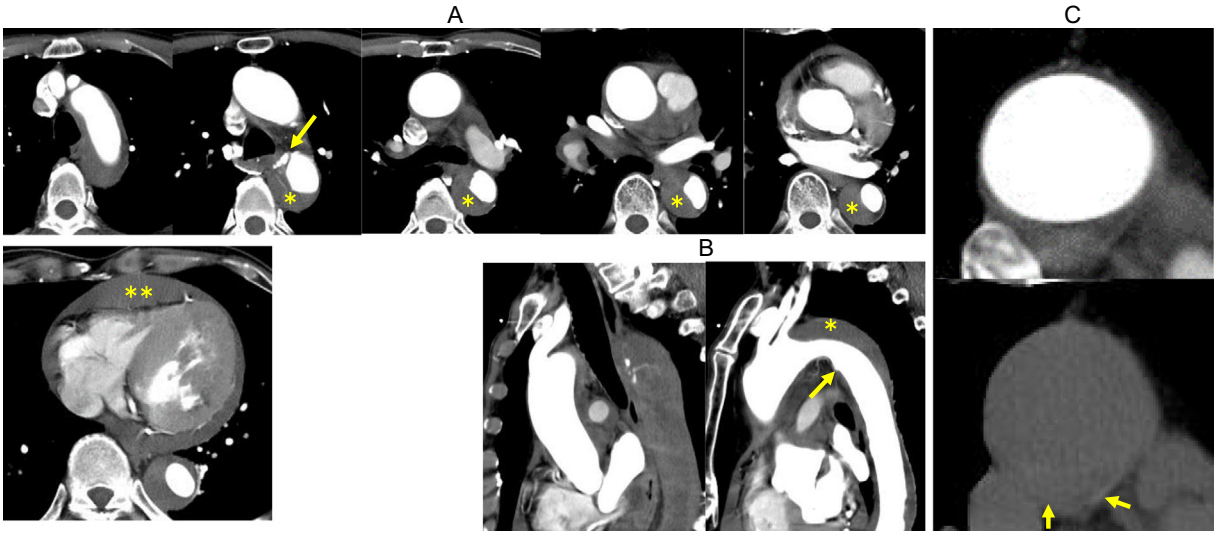

Figure 3. **Contrast-enhanced CT in case 2 (A: horizontal, B: sagittal). Pericardial effusion (**) and thrombosed false lumen in the descending aorta of the arch (*) with ULP (arrow) were found. Dissociation was observed up to the left subclavian artery branch (*) but not in the ascending aorta. A dense layer of tissue on the crescent moon confined to the posterior side of the ascending aorta (yellow arrows) was observed by plane CT. Such findings were not observed on contrast-enhanced CT.**

gery. He was administered with electric shocks in an ambulance during transport and suffered cardiac arrest after arriving at the transfer destination. He underwent emergent pericardial drainage and surgery and survived. Intraoperative findings showed dissection of the ascending aorta and rupture of the adventitia near the aortic root.

In Case 1, a thin dense layer of tissue was observed that completely encircled the ascending aorta, and in Case 2, a dense layer of tissue crescent moon and confined to the posterior side of the ascending aorta was observed on non-contrast CT (Fig. 1C, Fig. 3C).

Discussion

In the two cases presented here, the false lumen and entry of dissection were unclear in the ascending aorta on contrast-enhanced CT.

In Case 1, pericardial effusion was observed, but the entry and false lumen of the dissection were unclear. Contrast-enhanced CT was repeatedly performed after the patient's admission to hospital, and aortic findings did not change. Histopathological findings showed the entry of dissection at the descending aorta of the arch, dissection of the ascending/descending aorta, and perforation of the pericardial cavity from behind the ascending aorta. Based on the patient's clinical course, the dissection that occurred in the descending aorta of the arch extended to the ascending aorta retrogradely and was closed with a thrombus. Re-dissociation then occurred on the third day, ruptured into the pericardial cavity, and led to cardiac tamponade.

In Case 2, IMH with ULP was found in the descending thoracic aorta. Initially, we presumed the patient's diagnosis

to be Stanford type B aortic dissection. However, because repeated echocardiography confirmed a rapid increase in pericardial effusion, we suspected dissection of the ascending aorta and rupture of the pericardial cavity. An emergency operation was performed, which saved the patient's life. Considering the clinical progression, images, and surgical discoveries, the dissection that developed in the descending aorta of the arch expanded retrogradely to the ascending aorta was temporarily closed with a thrombus and then redissociated.

It is difficult to diagnose TAAD when dissection of the ascending aorta is unclear on contrast-enhanced CT, as in the cases presented here. There have been some reports of aortic dissection in which the false lumen is not imaged by contrast CT (1, 2). The pathological condition is considered to involve closure of the false lumen with thrombosis early after the onset. These cases are referred to as IMH and reportedly account for 10-30% of aortic dissections (3-5). It was also reported that the hyperdense crescent sign, in which the occluded false lumen is highly absorbed by plain CT, aids in making a diagnosis (6), and the same findings were confirmed in the present two cases (Fig. 1C, 3C).

Regarding the treatment policy for patients with Stanford TAAD and IMH, some reports state that a good prognosis can be expected with medical treatment (7-10), but others recommend emergency surgery (3, 5, 11); thus, there is no clear guideline at present. In particular, there have been very few reports on treatment policies for retrograde TAAD.

Kaji et al. reported a good long-term prognosis with a 5-year survival rate of 93% after medical treatment of 14 cases of retrograde TAAD, in which the false lumen of the ascending aorta was completely thrombosed; however, in the discussion, they considered the following clinical conditions as indications for emergency surgery:

1) enlargement of the ascending aorta and arch,

2) fatal complications including aortic regurgitation, and

3) rupture of the affected aorta (12).

Cardiac tamponade has been reported to be associated with 18.7% of Stanford type A acute aortic dissections and is considered to carry a major postoperative mortality risk (13). Kim et al. recommended urgent surgical intervention for aorta-related complications with signs of malperfusion or significant pericardial effusion (14). Ascending aorta replacement is performed in patients with ordinary TAAD in which the tear is in the ascending aorta, whereas retrograde TAAD is often performed when the tear is present in the distal arch and arch aortic replacement is required. There is a greater difficulty of surgery in cases of retrograde TAAD than in cases of ordinary TAAD.

Based on the clinical course of the two cases presented here, the following points were presumed to be important in retrograde TAAD diagnosis and treatment:

1) When diagnosing patients with chest pain who have no ST changes and pericardial fluid retention, it is important to suspect acute TAAD and to perform CT to examine the ascending aorta.

2) When contrast-enhanced CT does not reveal a clear dissection in the ascending aorta, the presence or absence of HCS on plane CT should be fully evaluated. Transesophageal echocardiography may be useful for confirming dissection of the ascending aorta.

3) If conservative treatment is selected because the false lumen of the ascending aorta is thrombotic and hemodynamics are stable, a CT reexamination on the day after admission is recommended.

4) When dissection with ULP in the thoracic descending aorta or pericardial effusion is observed, emergency surgery should always be considered.

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

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