Case Report

# **Stanford Type A Acute Aortic Dissection** with Intimal Intussusception

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In case of complete circumferential dissection of the ascending aorta, the dissected flap has the potential to fold backwards, causing several complications. We report two cases of Stanford type A acute aortic dissection (AAD) whose intimal flaps intussuscepted into the left ventricular outflow tract.

Case 1: A 41-year-old man with AAD in whom transthoracic echocardiography (TTE) showed the dissected flap as folded back into the left ventricular outflow tract, causing severe aortic regurgitation (AR) with rapidly progressing acute pulmonary edema. Despite performing salvage surgery, the patient could not be rescued.

Case 2: An 81-year-old man with annuloaortic ectasia developed Stanford type A AAD. TTE showed an extremely mobile intimal flap intussuscepting into the left ventricular outflow tract. However, AR was not severe as it was prevented by the flap itself. The patient was rescued by performance of the modified Bentall procedure.

**Keywords:** thoracic aorta, aortic dissection, aortic regurgitation

#### Introduction

Complete circumferential dissection of the ascending aorta is a rare event. Rosenzweig $^{1)}$  reported that it occurred in 2% of ascending aortic dissections. We report two cases of complete circumferential dissection due to Stanford type A acute aortic dissection (AAD).

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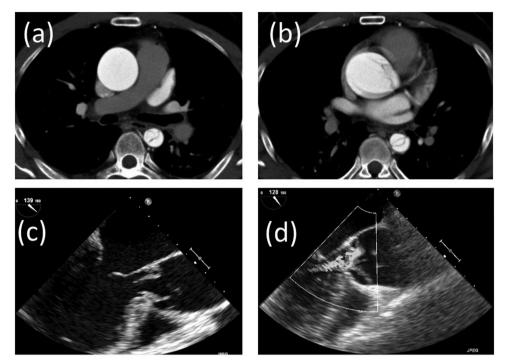
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## **Case Report**

Case 1: A 41-year-old man was taken to another hospital by ambulance with sudden onset vomiting, abdominal pain, and syncope. He regained consciousness at the hospital. Enhanced computed tomography (eCT) showed dissection of the thoracic aorta and pulmonary congestion. Since he was diagnosed with Stanford type A AAD, he was referred to our hospital for immediate surgical treatment. Physical examination revealed a blood pressure of 94/36 mmHg and heart rate of 74 beats/min. Electrocardiography (ECG) showed sinus rhythm and ST segment slightly depression in leads V3-6. eCT seemed to indicate absence of the dissected flap in the ascending aorta although it was present in the aortic root (Figs. 1a and 1b). This imaging finding is called "missing flap." Transthoracic echocardiography (TTE) showed intussusception of the ascending aortic intimal flap into the aortic valve during diastole, along with severe aortic regurgitation (AR). The patient was hypoxic upon arrival, with rapid deterioration in his condition. Since he was coughing up pink frothy sputum and was hypoxic,

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**Fig. 1** Imaging evaluation in case 1. (a) and (b) show eCT images, which seemed to indicate the absence of dissection in the ascending aorta (a). However, the dissected intimal flap was visible at the aortic root (b), the so-called "missing flap." (c) and (d) show TEE images. The dissected intimal flap was intussuscepted into the left ventricular outflow tract (c). Since the intimal flap prevented closure of the aortic valve in diastole, it resulted in severe AR (d). eCT: enhanced computed tomography; AR: aortic regurgitation; TEE: transesophageal echocardiography

emergency intubation was performed and he was transported to the operation room. However, the hypoxia persisted and he developed cardiac arrest. By establishing cardiopulmonary bypass (CPB) as soon as possible, spontaneous circulation was restored during continuing cardiac massage and surgical repair was performed. Transesophageal echocardiography (TEE) showed severe AR due to intussusception of the ascending aortic intimal flap (Figs. 1c and 1d). Under moderate hypothermia and circulatory arrest, the ascending aorta was incised and the entire circumference of the ascending aorta was dissected near the sinotubular junction. The aortic valve was left intact. Although ascending aortic replacement was performed, the patient could not be weaned off CPB because of acute pulmonary edema and myocardial damage due to acute AR. The patient returned to the intensive care unit (ICU) with percutaneous cardiopulmonary support, where he died. The pathology of the dissected aortic wall suggested cystic medial necrosis.

Case 2: An 81-year old man was transported to the emergency room with dizziness and reeling after playing park golf. TTE demonstrated a mobile flap in the ascending aorta. Since the eCT revealed Stanford type A AAD,

the patient was transferred to our hospital for surgical treatment. Physical examination revealed a blood pressure of 80/40 mmHg and heart rate of 50 beats/min. ECG showed sinus rhythm with no significant ST-T segment shift. eCT showed (Figs. 2a and 2b) a "missing flap" in the ascending aorta and dilatation of the aortic root (diameter: 64 mm). Intraoperative TEE during emergency surgery showed an extremely mobile dissected flap which moved in and out of the aortic valve in synchronicity with the cardiac cycle although it did not result in severe AR (Figs. 2c and 2d). Intraoperative findings indicated that the intima of the ascending aorta was completely torn off proximal to the innominate artery. The intimal flap was intussuscepted at the aortic valve. Hence, the modified Bentall procedure was performed. He was transferred to another hospital for rehabilitation on the postoperative day 38. The pathology of the dissected aortic wall did not appear to be cystic medial necrosis.

### **Discussion**

Complete circumferential dissection with Stanford type A AAD, which is rare, can reportedly cause several

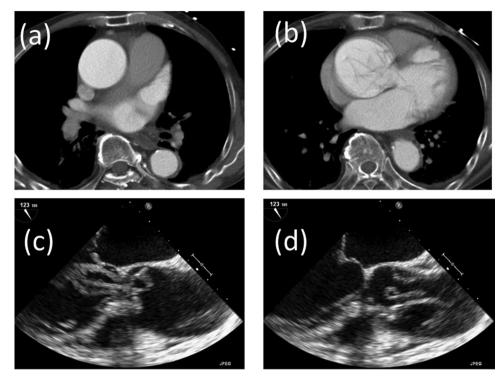


Fig. 2 Imaging evaluation in case 2. (a) and (b) show eCT images. Dissection was not seen in the ascending aorta (a) ("missing flap") although an intimal flap was apparent in the aortic root (b). The aortic root was dilated. (c) and (d) show transesophageal echocardiography images. The dissected intimal flap exhibited dynamic mobility. It was intussuscepted into the left ventricular outflow tract during diastole (c) and returned to the ascending aorta during systole (d). eCT: enhanced computed tomography

complications due to intussusception of the mobile dissected flap.<sup>2)</sup> If the flap folds over distally, it causes syncope;3) if folded over proximally, it can cause occlusion of the coronary artery<sup>4)</sup> or AR.<sup>1,5,6)</sup> In our case 1, the flap was folded over into the left ventricular outflow tract in diastole, preventing closure of the aortic valve, resulting in severe AR and rapid development of acute pulmonary edema. Although salvage surgical repair was performed, the patient could not be weaned off CPB and died soon thereafter. In our case 2, the dissected flap was extensive and extremely mobile, and intussuscepted into the left ventricular outflow tract during diastole. Although the flap interfered with closure of the aortic valve, the excess tissue of the flap itself reduced the regurgitant volume through the aortic valve. The difference between case 1 and case 2 was the position at which the dissected flap was disconnected. In case 1, the circumferential dissected flap was disconnected just above the sinotubular junction. The short-dissected flap was therefore folded back into the left ventricular outflow tract, preventing the aortic valve from closing. In case 2, the dissected flap was disconnected near the proximal portion of the innominate artery. Almost all the ascending aorta was circumferentially dissected, and a large amount of the flap tissue was intussuscepted into the left ventricular outflow tract during diastole. Therefore, we consider that the difference between the AR grade of case 1 and that of case 2 caused the difference in the position of flap disconnection. As a result, although intussusception of the dissected flap occurred in both cases, the degrees of intussusception were different, resulting in different symptoms and, ultimately, different pathophysiological outcomes.

As stated above, AAD with a mobile flap is associated with a high probability of causing severe complications. Hence, it requires urgent diagnosis and treatment.<sup>5)</sup> Usually, Stanford type A AAD cases show a false lumen and dissected flap at the ascending aorta on eCT. However, in cases with circumferential dissection, the eCT image indicates disappearance of the flap because the flap is folded over proximally or distally, with only the adventitia remaining in the dissected part of the aorta. This imaging finding which is called "missing flap" sometimes leads to misdiagnosis.<sup>3,7,8)</sup> The phenomenon of the "missing flap" has been reported in detail by Sanders

et al.<sup>9)</sup> In our patients, although eCT images showed seemed to indicate absence of a flap (missing flap) in the ascending aorta, the dissected flap was visible in the aortic arch and TTE showed the dissected flap in the aortic root. Hence, we could easily diagnose Stanford type A AAD. TTE can easily demonstrate the mobile dissected flap in the ascending aorta. Hence, screening TTE is very important for the diagnosis of AAD.

### Conclusion

Stanford type A AAD with complete circumferential dissection is a serious disease since it can cause severe complications secondary to intimal intussusception. Screening TTE is useful for the diagnosis of a mobile flap in the ascending aorta.

### **Disclosure Statement**

None of the authors has any conflicts of interest to declare.

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