

**Case
Report**

Extremely Localized Aortic Dissection and Intussusception of the Intimal Flap into the Left Ventricle

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Stanford type A aortic dissection frequently deforms the aortic root and causes aortic regurgitation (AR). On the rare occasion, massive AR can occur due to circumferential intimal disruption and prolapse of the cylinder-shaped intimal flap into the left ventricle. Because of the critical, general, and hemodynamic state of such patients, surgery for this condition carries a high risk. A 62-year-old woman suffered acute chest pain and fell into cardiogenic shock. Computed tomography and transthoracic echocardiography failed to identify the etiology of this rapid hemodynamic collapse. Transesophageal echocardiography (TEE) demonstrated circumferential intimal disruption, 3 centimeters above the aortic valve annulus; a very localized aortic dissection in the proximal ascending aorta; and a to-and-fro motion of cylinder-shaped intima causing severe AR. The dissection did not affect the aorta beyond the intimal tear, and TEE was the only useful modality for the diagnosis. Emergency replacement of the ascending aorta and resuspension of the aortic valve was successfully performed. Residual AR was absent, and the postoperative course was uneventful.

Key words: localized acute aortic dissection, aortic regurgitation, intussusception of the intimal flap

Introduction

Stanford type A acute aortic dissection is frequently associated with aortic regurgitation (AR). In the vast majority, deformity of the aortic root is the cause of AR. We encountered a rare case that circumferential intimal disruption and intussusception of the cylinder-shaped intima into the left ventricular cavity caused massive AR and cardiogenic shock.¹⁻⁴⁾ Diagnosis was very difficult in this patient because dissection was localized just around the aortic root and did not affect the aorta beyond the

circumferential intimal tear. Transesophageal echocardiography (TEE) was the only modality that was useful to determine the therapeutic strategy for this critically ill patient.

Case

A 62-year-old woman, who had been treated for hypertension, suffered acute chest pain and dyspnea. Blood pressure was 66/20 mmHg, heart rate was 128/min, and the patient was transferred to our hospital. Shock state, orthopnea, and anuria persisted despite strenuous inotropic support. Moist rale at bilateral lung field and Levine III/VI to-and-fro murmur along the left sternal border was noted. Critical hypoxia (arterial oxygen tension of 48mmHg under maximal oxygen inhalation) prompted endotracheal intubation. Results of routine check-up were unremarkable except severe pulmonary congestion on chest X-ray. Computed tomography (CT) of the chest was also unremarkable except multiple linear

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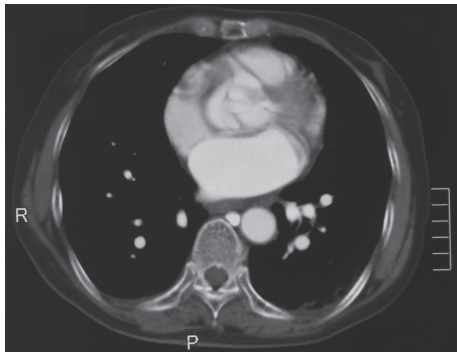


Fig. 1 Multiple linear structures were revealed in only two images around the aortic valve level, which were not distinguishable from the artifact.

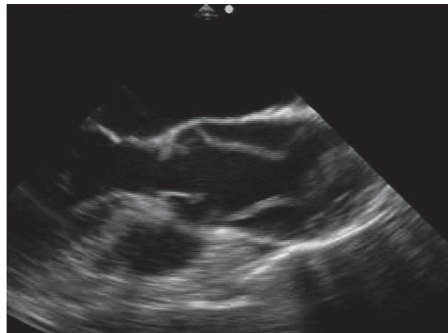


Fig. 2 (a) Very short and cylinder-shaped intimal flap fell into the left ventricle during the diastolic phase, causing severe AR. (b) The intimal flap ejected back into the ascending aorta during the systolic phase. AR, aortic regurgitation

a | b

structures, which were not distinguishable from artifact, in only two images around the aortic valve level (**Fig. 1**). Transthoracic echocardiography showed severe AR with normal size and function of the left ventricle (end-diastolic diameter of 42 mm, end-systolic diameter of 25 mm, ejection fraction of 69%). Acute AR was suggested, but its etiology could not be identified.

Medical therapy obviously failed to manage cardiogenic shock due to AR. Emergency operation appeared to be mandatory, although surgical procedure was undetermined. At operative theater, TEE demonstrated circumferential intimal disruption 3 centimeter above the aortic valve annulus and localized aortic dissection proximal to the intimal tear. Very short and cylinder-shaped intimal flap fell into the left ventricle during diastolic phase, causing severe AR, and was ejected back into the ascending aorta during systolic phase (**Fig. 2**).

Through a midline sternotomy, mildly hypothermic (28°C) cardiopulmonary bypass was instituted between both vena cava and right femoral artery. As dissection affected only around the aortic root (**Fig. 3**), the unaffected ascending aorta was clamped. There was a cir-

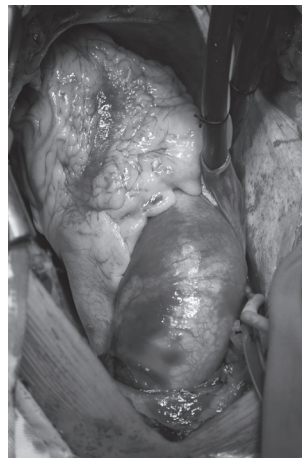


Fig. 3 Aortic dissection was localized at only around the aortic root.

cumferential intimal tear 15mm above the right coronary ostium and 20mm above the left coronary ostium. The aorta proximal to this tear was dissected circumferentially and was inverted to the left ventricular outflow tract. Dissection did not affect the aorta distal to the circumferential intimal tear at all. Resuspension of the aortic valve by sewing all commissures in place and

replacement of the ascending aorta was performed. Post-operative echocardiography showed no residual AR, and clinical course was uneventful. The patient discharged home on foot 14 days later.

Discussion

Dissection limited in the ascending aorta alone (DeBakey type II) is the least common mode of aortic dissection with frequency of about 20%.⁵⁾ In the present case, dissection affected just around the aortic root, 3cm in length. Such an extremely localized dissection is rare, and it is unknown why the aorta beyond the intimal tear was completely free from dissection. A very large intimal tear might play some role.

More than mild degree AR is observed in 60%–70% of patients with Stanford type A acute aortic dissection, and deformity of the aortic root, namely, annulus or commissure, is the most common cause.⁴⁾ On the other hand, circumferential intimal disruption is rare but hazardous. Intussusception of the cylinder-shaped intima into the left ventricle causes massive AR. When intimal flap is swung proximally, coronary artery flow may be compromised.^{6–8)} When it is swung distally, aortic arch vessels may be obstructed, and cerebral malperfusion may ensue.⁹⁾ In general, patients with this type of tear are critically ill and require an emergency surgery, which carries a very high operative mortality of 40%.¹⁾ Some patients including the present one may undergo surgery without a definitive diagnosis.¹⁰⁾

Infective endocarditis, aortic dissection, avulsion of the aortic valve commissure, rupture of fibrous strands and so forth should be considered as a cause of acute AR.^{11, 12)} The main problem of this moribund patient was an unclear etiology, although the indication and maneuver of surgery are highly dependent on the diagnosis. Judging from negative laboratory tests and the afebrile state, infective endocarditis was unlikely. However, area of dissection was too limited to be diagnosed definitively with CT. The aorta was completely normal except in two images around the aortic valve annulus, and sanguinous pericardial effusion or hematoma was absent. In addition, there was no sign of cardiac ischemia, which is a well known complication of aortic dissection. The only useful diagnostic modality in this patient was TEE that was performed in the operating theater. However, TEE is not always suitable for patients in a serious condition. There-

fore, physicians should be aware of this rare entity, and that such an extremely localized dissection can cause hemodynamic collapse and necessitate emergency surgery.

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