

Aortic Arch Aneurysm of Takayasu Arteritis Associated with Entero-Behçet Disease

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We report a case of a ruptured aortic arch aneurysm due to Takayasu arteritis concomitant with entero-Behçet disease. A 53-year-old woman with total left lung atelectasis underwent emergency total aortic arch replacement with a modified Bentall operation and elephant trunk procedure. The postoperative course was highly eventful. A pseudoaneurysm of the left coronary button occurred with mediastinitis due to fistula of the left bronchus into the remnant of the aneurysmal wall. The left main trunk was reconstructed with a saphenous vein graft. The left bronchial fistula into the esophagus was exposed and an esophageal stent was placed. Finally, the saphenous vein graft ruptured and the patient expired. The autopsy diagnosis was Takayasu arteritis. This is the first reported case of concomitant Takayasu arteritis and entero-Behçet disease. (Ann Thorac Cardiovasc Surg 2007; 13: 216–219)

Key words: aortic arch aneurysm, aortitis, Behçet disease, Takayasu arteritis

Introduction

We report an unusual case of a patient who suffered from a huge ruptured aortic arch aneurysm. There was a history of entero-Behçet disease. The surgery performed was an aortic root reconstruction and total arch replacement concomitant with an elephant trunk procedure. Various complications developed. The patient died of a coronary event 4.5 months after the first operation. Ultimately, Takayasu arteritis was diagnosed at autopsy. Coexistence of Takayasu disease and entero-Behçet disease has not been reported in the literature to date.

Case Report

A 53-year-old woman, who had been diagnosed with entero-Behçet disease and medicated with Tharazopilin

for 10 years, was referred to our department for an emergency operation due to a ruptured aortic arch aneurysm. Her entero-Behçet disease was stable in the pre-operative state, and the C-reacting protein (CRP) value was around 3 to 5 mg/dL without steroid therapy. Computed tomography (CT) revealed a true aneurysm was over 8.0 cm in diameter, compressing the trachea and left main bronchus (Fig. 1A). She suffered from complete atelectasis of the left lung and was intubated due to respiratory failure (Fig. 1B). Moderate aortic valve regurgitation was revealed in the echocardiography. We performed an emergency operation for a ruptured aneurysm.

A median sternotomy was made. The aneurysmal wall was white, very thick and solid, thus confirming inflammatory change due to aortitis. Cardiopulmonary bypass (CPB) was instituted with right atrial venous drainage and central perfusion via the aneurysm. The aneurysm was opened under hypothermic circulatory arrest. The distal end of the aneurysm was at the level of the descending aorta. The left carotid artery and left subclavian artery were occluded. The aortic valve was floppy and incompetent. Initially, an arch reconstruction was performed by grafting only the right brachiocephalic artery using a composite graft with 4 branches (Hemashield Gold®). Under antegrade brain perfusion, a distal anastomosis to

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the descending aorta was performed by the elephant trunk method over a 10 cm length, and systemic perfusion was started thereafter. On the proximal side, we performed a modified Bentall procedure. The aortic valve annulus was completely separated from the Valsalva sinus because the wall of the Valsalva sinus is considered part of the aortic wall. A composite graft, consisting of a 26 mm Hemashield Gold® graft and a 25 mm CarboMedics mechanical valve, was sutured to the annulus by including a deep stitch into the ventricular wall from outside the annulus with interrupted, pledget-supported, 2-0 polyester mattress sutures, reinforced by a Teflon felt strip placed surrounding the outside of the annulus. Two short pieces of graft (8 mm) were placed for the right and left coronary arteries because the coronary buttons were not mobilized. The time of total circulatory arrest was 19 min, cardiac arrest 184 min and CPB 264 min. Left total lung atelectasis improved and she was extubated the next day.

Steroid therapy was started on the second postoperative day on the basis of a clinical diagnosis of vascular Behçet disease. Her postoperative course was highly eventful. At one month after operation, a fistula between the left main bronchus and aortic aneurysmal wall was found (Fig. 2A), and we performed an omentopexy into the fistula concomitant with wrapping the graft.

At 3 months, a pseudoaneurysm formed at the left coronary button. An emergency coronary bypass on the left main trunk was performed by switching the Pielar's interposed graft to the saphenous vein graft (SVG) (Fig. 2B). Four months after the initial operation, another fistula developed between the left main bronchus and the esophagus. Emergency treatment was performed using an esophageal stent (Ultraflex® 10 cm). After 4.5 months, she suffered hemorrhagic shock due to a perforation of the replaced SVG, and could not be resuscitated despite an emergency operation.

The pathological findings in the aorta revealed fibrous thickening of the adventitia and spotty destruction of the media, which confirmed the scar stage of Takayasu arteritis (Fig. 3A). The brachiocephalic artery, the only patent branch of the aortic arch, definitely proved the Takayasu arteritis histopathologically. The outer side of media was destroyed in a spotty fashion (Fig. 3B). No other vessels involved active inflammation, thus supporting the differential diagnosis of vasculo-Behçet disease.

Discussion

We encountered a rare case of a huge aneurysm in a pa-

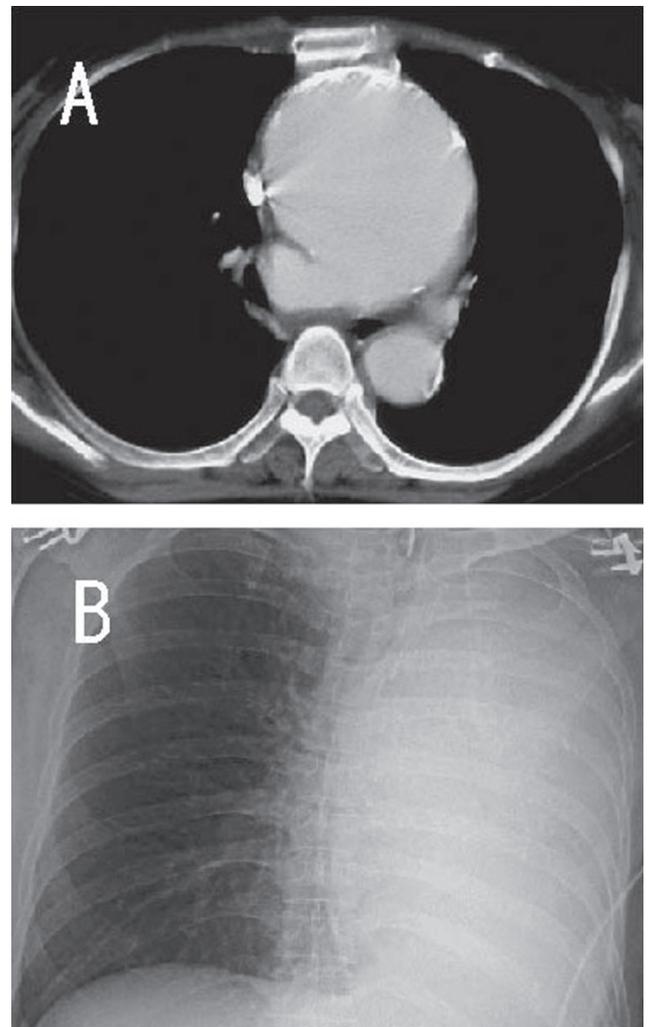


Fig. 1.

A: Computed tomography shows that the left main bronchus was compressed by a huge aneurysm.

B: Chest radiograph reveals complete atelectasis in left lung.

tient with Takayasu arteritis with entero-Behçet disease. The entero-Behçet disease in this patient had been confirmed 10 years previously. She had a history of genital ulceration and aphthous stomatitis. It was natural that this aneurysm could have been caused by vasculo-Behçet disease; however, it was finally diagnosed as Takayasu arteritis. There are differences between vasculo-Behçet disease and Takayasu arteritis in the pathology of the large arteries. A typical aortic change in vasculo-Behçet disease has been thought to be a nonspecific mesoaortitis with severe destruction in all layers of the media and secondary fibrous thickening of the intima and adventitia in the aorta.¹⁾ In Takayasu arteritis, the slow spotty loss of elastic fibers occurs in the adventitia and the outer me-

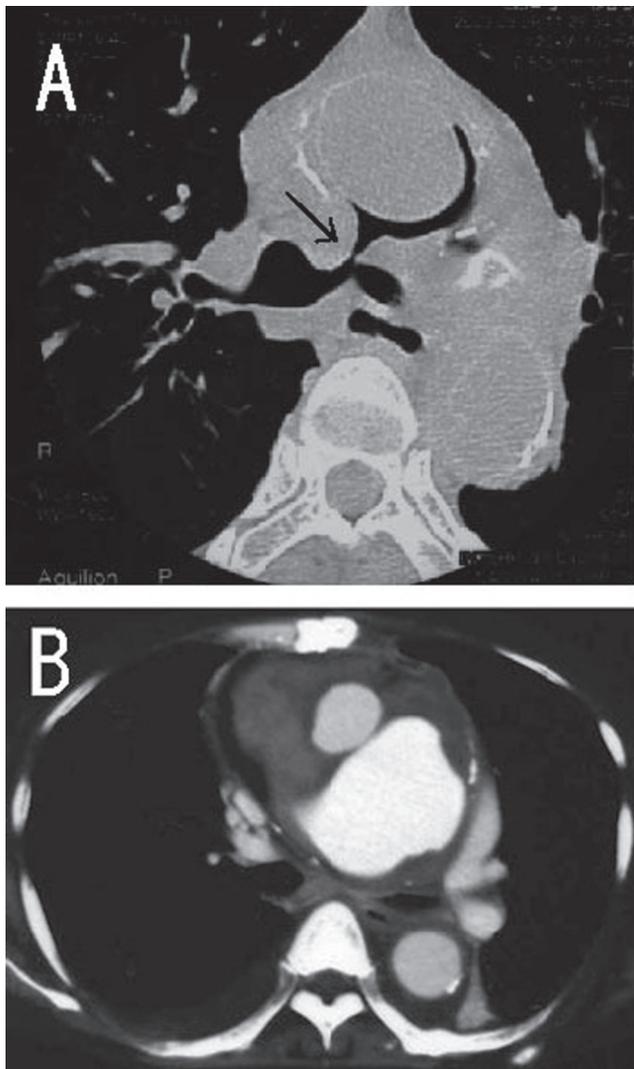
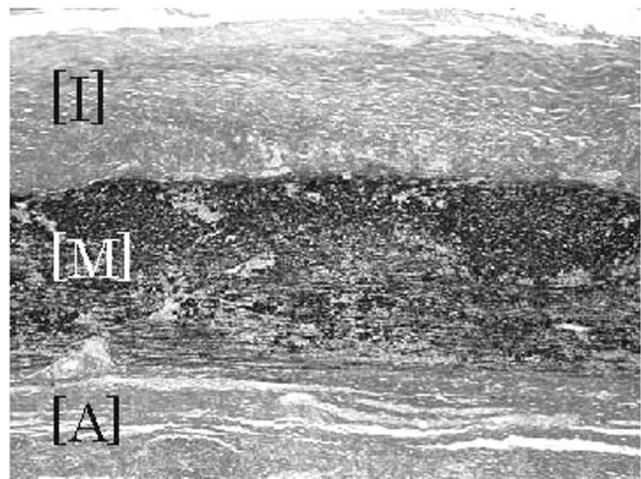
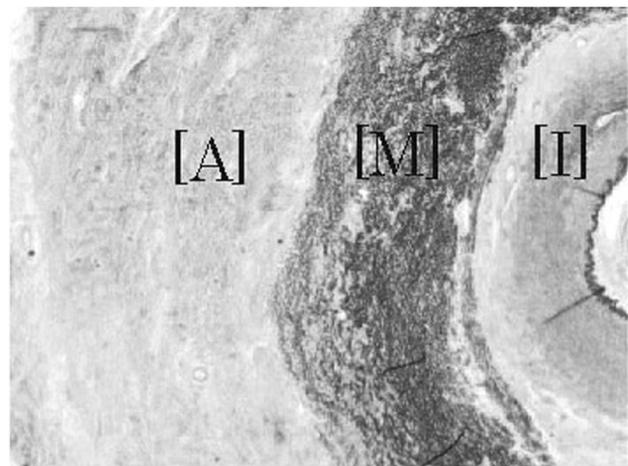


Fig. 2.
A: Computed tomography showing the fistula between the left main bronchus and the aneurysmal wall (arrow).
B: Computed tomography showing pseudoaneurysm in the mediastinal space.

dia. The pathology in this patient was diagnosed as Takayasu arteritis from the findings of elastic fiber spotty loss on the outer side of the media in the brachiocephalic artery and descending artery. Female sex and occlusive changes of the left carotid and left subclavian artery are characteristic of Takayasu arteritis, but we could not diagnose this etiology clinically because of the history of entero-Beçet disease. Thus, both Takayasu arteritis and entero-Beçet disease coexisted in this patient. A 1985 review indicated that 3 of 57 autopsies of vasculo-Beçet disease had findings that suggested Takayasu arteritis.²⁾ However, the coexistence of both diseases had not yet



A



B

Fig. 3. Pathological finding of aortitis (Elastic van Giessen stain).
A: Descending aorta. Note spotty destruction of loss of elastic fibers in outer side of media [M] and fibrous thickening of adventitia [A]. [I], intima.
B: Brachiocephalic artery. Note destruction on outer side of media [M] and thickening of adventitia [A]. [I], intima.

been reported in the literature. This report may be the first concerning a patient suffering from both Takayasu arteritis and entero-Beçet disease.

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