Aortic Dissection Associated with Aortitis Syndrome

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SUMMARY

A 50-year-old woman with aortitis syndrome complicated by aortic dissection, is reported. The dissection was observed at the level of the descending thoracic aorta by aortography and at the intimal side of the dilated aorta on CT. An aneurysm of the right subclavian artery and a diffusely thick wall of the abdominal aorta were also observed. This case suggests that the uneven wall of the aorta in aortitis syndrome might be dissected at the intimal side by dilatation.

Key Words:
Takayasu's arteritis Dissecting aneurysm Separation of intima

AORTITIS syndrome (Takayasu's arteritis) is a chronic non-specific arteritis of the aorta and main branches. Aortic dissection is a rare complication because of fibrous and granulomatous lesions of the aortic wall, although some cases have been reported. The process of aortic dissection, however, remains unknown. We report the case of a patient with aortic dissection associated with aortitis syndrome showing an unusual intimal separation related to the dissection.

CASE REPORT

A 50-year-old Japanese woman had suffered from fatigue and anemia since the age of 20. She had experienced severe back pain 5 years prior to admission. She had had lymphadenitis in her neck and arterial disease was suspected 1 month before. She was admitted to our hospital on March 25, 1989 for further examination. At admission she complained of fatigue but was healthy. Blood pressure was 130/70 mmHg and did not differ among the extremities. Vascular bruits were heard over the second right rib and
epigastrium. There were no other abnormal physical findings.

On examination, red blood cell count was $387 \times 10^4/\mu l$, hemoglobin 8.9 g/dl, and hematocrit 29.4%. C-reactive protein was negative and erythrocyte sedimentation rate was 26 mm at 1 hour. Urinalysis, blood chemistry, STS and immunoglobulins were within the normal range. Chest x ray showed diffuse and high-grade calcification of the aorta and a small abnormal shadow at the right second rib. Pulmonary perfusion scintigraphy did not show any lesion of the pulmonary artery.

Aortography (Fig. 1) showed a right subclavian artery aneurysm (fusiform $34 \times 16$ mm) and aortic dissection of the descending thoracic aorta that opacified in the late phase. The abdominal aorta proximal to the renal artery showed an irregular wall and was stenosed in a short segment, but the distal aorta was normal. The aortic lesion continued from the thoracic to the abdominal aorta. On computed tomogram (Fig. 2), the descending thoracic aorta dilated at the 6th thoracic vertebra, and then had a false lumen at the 7th to 10th vertebrae (Fig. 3). The true lumen was opacified by contrast material, some of which flowed into the false lumen (Fig. 2-D), which was made of the dilated aortic wall and a thin intimal side wall separated by the tear. The wall of the proximal abdominal aorta (Fig. 2-E) was

![Fig. 1. A right subclavian artery aneurysm, and aortic dissection with dilatation and a partial defect of the descending thoracic aorta in the early phase of the aortogram.](image)
Fig. 2. Dilatation and dissection of the descending thoracic aorta on computed tomogram. A: normal size transverse aorta (right) at the 5th thoracic vertebra (left, white). B: dilated aorta with thick and uneven wall at the 6th. C: dissected aorta at the 7th. A part of the thick wall was torn, and the thin intimal side wall (arrows) separation is continuous with the tear, resulting in a false lumen (upper lumen). D: dissected aorta at the 8th. The true lumen (bottom) stained by contrast material. A part of the material flowed into the false lumen (top). E: thick and uneven abdominal aorta at the 11th.
also abnormally thick but the distal aorta had a normal wall.

**DISCUSSION**

The etiology of aortitis syndrome remains unknown. Therefore it is diagnosed by clinical findings as follow: 1) multiple lesions of aorta, its main branches and pulmonary artery, 2) unknown etiology of the arteritis, 3) inflammatory signs.

Our patient showed a diffuse aortic lesion and a subclavian artery aneurysm of unknown etiology. There were no signs of active inflammation but they seemed to have disappeared over the long time course of her disease from 20 years of age.

Aortic dissection associated with aortitis syndrome had previously been reported in 13 patients, 12 of whom were Japanese. It occurred at various stages in the course of the disease, but was located in the descending thoracic and upper abdominal aorta in 12 of the 13.1)-12) Three patients were studied by autopsy, and showed inflammatory changes in the dissected wall.1)-3) One patient was examined by computed tomography, but the detail of the dissected wall was not described.4) One patient was observed at operation for coarctation of the aorta and the false lumen was seen to be divided by the intimal flap.5) Eight other patients6)-12) were observed only by aortography, and details of the dissected wall were not described.
Our patient had dissection of a short segment of the descending thoracic aorta that was similar to previously reported cases. Her dissection occurred in the thickened and calcified aorta, and consisted of an unusual separation of the intimal wall continuous with the torn portion in the dilated aortic lumen. The aortic architecture of aortitis syndrome takes the form of thick intima, fibrous and adhesive adventitia, and thin atrophied media. Therefore the intima could be separated from the media.

These observations suggest, as a process of dissection in aortitis syndrome, that uneven aortic wall might tear at the dilation of the aorta, and the intima might separate continuously with the tear as a result of the inflow of blood, and a false lumen might be formed inside the dilated lumen.

References