AN ORANGE-SHAPED AORTIC ROOT ANEURYSM IN AORTITIS SYNDROME WITH SEVERE AORTIC REGURGITATION

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A 57-year-old man, who had undergone aorto-coronary bypass surgery 4 years before when the shape of the ascending aorta had been normal, had a unique orange-shaped aortic root aneurysm associated with severe aortic regurgitation and congestive heart failure. Replacement of the aneurysm and the aortic valve was successfully carried out, and histopathological examination revealed that the aneurysm was caused by aortitis syndrome.

(Jpn Circ J 1992; 56: 687–689)

There are wide variations in the clinical features of aortitis syndrome according to the vessels involved.

We report on a patient with aortitis syndrome who manifested a unique orange-shaped aneurysm localized in the aortic root. This lesion had developed within 4 years, and was associated with severe aortic regurgitation and congestive heart failure.

CASE REPORT

A 57-year-old man was admitted to our hospital because of dyspnea.

Four years earlier, he had undergone aorta-coronary saphenous vein bypass graft surgery because of stenosis at the bifurcation of the left anterior descending and circumflex arteries. He was then well until one and half years before entry when a diastolic blowing heart murmur appeared for the first time. He experienced dyspnea on effort one year before admission, followed by nocturnal orthopnea and abdominal fullness. He was referred to our hospital in April, 1989.

On examination, arterial pulsations in the extremities were all easily palpable. Blood pressures measured in the right and left arms were 136/56 mmHg, 134/68 mmHg respectively. Cardiac examination showed third heart sound and grade 3 diastolic blowing murmur on left sternal border. The liver was palpable 2 cm below the right costal margin.

Laboratory data included a moderately enhanced erythrocyte sedimentation rate (48 mm/1 h) and C-reactive protein was positive (1.3 mg/dl). Serological tests for syphilis and autoimmune antibodies of various types were negative. PPD response was positive. A chest X-ray showed cardiomegaly, pulmonary congestion and right pleural effusion. The ECG was compatible with left ventricular hypertrophy. Echocardiographic examination revealed a markedly enlarged aortic root and dilated left ventricle. A Doppler color echocardiography disclosed severe aortic regurgitation.

In contrast to the normal configuration of the ascending aorta 4 years before, an

Key words:
Aortitis syndrome
Aortic root aneurysm
Aortic regurgitation

(Received July 29, 1991; accepted November 27, 1991)
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Fig. 1. In June, 1985, just after the aortocoronary bypass surgery, the ascending aorta was normal in shape (left). In April, 1989, an orange-shaped aneurysm of aortic root appeared and caused severe aortic regurgitation (right).

Fig. 2. Histological section of the aortic wall of the aneurysm showed infiltration of lymphocytes and plasma cells in the media (upper half) and thickened wall of vasa vasorum and diffuse fibrosis in the adventitia (lower half). (hematoxylin and eosin, ×4)

Aortogram showed an aortic root aneurysm of orange-like appearance involving the proximal end of the aorta and sinuses of Valsalva (55 mm in diameter) and severe aortic regurgitation (Fig. 1). Pressure data included the left ventricular end-diastolic pressure of 41 mmHg and the pulmonary arterial pressure elevated to 81/32 (mean 51) mmHg. Additional aortograms showed no abnormalities in other parts of the aorta and its main branches. A $^{99m}$Tc perfusion lung scan was normal.

An operation was performed with a pre-operative diagnosis of aortic root aneurysm of unknown cause with severe aortic regurgitation and congestive heart failure. An aneurysm, the wall of which was thickened and suggestive of inflammatory change, was localized in the aortic root including the lower end of the ascending aorta but not involving the proximal anastomosis site of the bypass graft. This aneurysmal portion and the aortic valve were successfully replaced using a modification of Bentall's method.

Microscopic examination of the aortic wall of the aneurysm showed that there was diffuse infiltration of lymphocytes and plasma cells in the media, wall thickening of the vasa vasorum and marked fibrosis in the adventitia although the intima was normal (Fig. 2). These findings were consistent with a diagnosis of the diffuse-proliferative type of aortitis syndrome! The aortic valve showed mucoid degeneration without in-
flamatory change.

The postoperative course was uneventful and the patient was discharged one month later.

DISCUSSION

Several pathological conditions including Marfan syndrome, hypertension, syphilis, infectious endocarditis and Beget disease or even the normal aging process may bring about aneurysms of the ascending aorta. In annuloaortic ectasia seen in patients with typical or forme fruste of Marfan syndrome, degenerative change usually involves sinuses of Valsalva and the whole ascending aorta, which causes western pear-like appearance.

On the other hand, the aortic root aneurysm seen in our patient was restricted to the lower end of the ascending aorta and three sinuses of Valsalva, and its wall was thickened. So the morphology of the aneurysm was obviously different from that seen in Marfan's syndrome. Furthermore, our patient was not old, had no hypertension, was serologically negative for syphilis, had no signs of Beget disease, and had no history of infectious endocarditis.

Aortitis syndrome is an inflammatory disease of unknown etiology. The lesion is not only obstructive but also ectatic, and often involves the aorta, its main branches and pulmonary arteries. We diagnosed our patient as having aortitis syndrome from the typical histopathological findings of the aortic wall.

Ectatic lesions of the ascending aorta in aortitis syndrome are not infrequent and can be seen in approximately 30% of all cases in some reports and its shape is, in general, a diffuse rod-like one with inflammatory thickening of the whole ascending aorta. Such an orange-shaped aortic aneurysm localized only in the aortic root as was seen in our patient is quite unique, and has been rarely reported. Nakano et al reported a case of aneurysm of the left aortic sinus caused by aortitis syndrome.

The time course for the formation of lesions in aortitis syndrome has not yet been fully clarified. Saito et al reported that dilatation of the ascending aorta and aortic arch becomes common in long-standing (more than 5 years after onset) cases. In our case, no abnormality had been found on the ascending aorta or sinuses of Valsalva at the time of aorto-coronary bypass surgery 4 years before. So it is clear that our patient's aneurysm developed within 4 years. Furthermore, it took at most 2 and half years for the aneurysm to be large enough to cause aortic regurgitation from the time when the diastolic blowing murmur appeared.

It may be arguable whether this patient should be treated with corticosteroid. We have not given him such an agent, and the erythrocyte sedimentation rate was 80 mm/h one year after discharge although there are no symptoms and signs typical for aortitis syndrome.

A coronary stenosis in aortitis syndrome is usually situated at the ostium, and is caused either by direct involvement of inflammation or by secondary intimal thickening due to the lesion on ascending aorta. So, although the erythrocyte sedimentation rate was also elevated (40 mm/h) at the bypass graft surgery 4 years before, we assume that the etiology of coronary artery stenosis of this patient might be an arteriosclerotic one rather than aortitis syndrome itself.

REFERENCES


Japanese Circulation Journal Vol.56, July 1992