Coronary Artery Ectasia with Annuloaortic Ectasia

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SUMMARY
A 50-year-old Japanese woman with annuloaortic ectasia was found to have total coronary artery ectasia without evident atherosclerosis. The coronary ectasia may have been secondary to or of similar etiology to the annuloaortic ectasia. There was neither stigmata of Marfan’s syndrome nor any sign of dissection of the ascending aorta or coronary arteries. Furthermore, the patient was not elderly, and had no hyperlipidemia, diabetes mellitus, or history of smoking. There was a marked blood pressure difference between the arms, and linear calcification was present in the aortic wall. A stenotic lesion was present in the right mid-subclavian artery. Although it is impossible to rule out atherosclerosis as the etiology of these findings, the possibility that they may be a manifestation of Takayasu’s arteritis is discussed. (Jpn Heart J 35: 389–394, 1994)

Key words: Coronary artery ectasia Annuloaortic ectasia Takayasu’s arteritis

The prevalence of coronary artery ectasia is approximately 1–2%, and its pathogenesis is most commonly atherosclerosis.1,2) However, coronary ectasia in association with other diseases has also been reported. We present a case of diffuse coronary ectasia with annuloaortic ectasia.

CASE REPORT

A 50-year-old Japanese woman, in whom cardiomegaly had been identified at the age of 40, was admitted for further examination. She had a history of palpitations and dyspnea after pregnancy, but on admission had no symptoms and no history of syphilis or myocardial infarction. Physical examination revealed

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blood pressure of 168/90 mmHg in the right arm and 142/96 mmHg in the left arm. An aortic regurgitation murmur was audible. There were atherosclerotic changes in both eyes, but no stigmata of Marfan’s syndrome. Laboratory examination revealed mild liver dysfunction, and plasma renin activity and aldosterone level were elevated to 5.1 ng/ml/hr and 120 pg/ml, respectively. Erythrocyte sedimentation rate was 12 mm/hr, C-reactive protein was negative, and gamma globulin was 1.9 g/dl. Serological test for syphilis and tests for rheumatoid factor and antinuclear factor were all negative. Other laboratory examination results were normal, including total cholesterol, triglyceride and fasting blood sugar. A chest X-ray revealed moderate cardiomegaly and diffuse calcification along the thoracic aorta. The electrocardiogram revealed mild left ventricular enlargement. Echocardiography and computed tomography revealed massive dilation of the ascending aorta, which was 84 mm in maximal diameter. The aortic annulus diameter was 38 mm. Mitral valve prolapse was not detected. Although the left ventricle was also enlarged, it appeared diminutive compared to the aorta.
Aortography revealed a large fusiform aneurysm of the ascending aorta (Figure 1) and grade I aortic valve regurgitation. Moreover, a stenotic lesion was observed in the right mid-subclavian artery (Figure 2). Selective coronary angiography demonstrated coronary ectasia of all three coronary arteries without atherosclerotic stenotic lesions. The diameters of the proximal right coronary artery, left main trunk, proximal left anterior descending artery, and proximal left circumflex artery were 6.6 mm, 13.6 mm, 9.6 mm, and 6.6 mm, respectively (Figure 3).

**DISCUSSION**

In this patient, the coronary arteries were extremely dilated without any irregular lesion suggesting an atherosclerotic etiology. There is a possibility that the dilation of the coronary arteries may have been secondary to or of similar etiology to the annuloaortic ectasia. Doud et al. reported that coronary ectasia is associated with abdominal aortic aneurysms, and Guy et al. suggested that it
may be pathogenetically linked to systemic aneurysmal vascular disease.

Annuloaortic ectasia results from cystic medionecrosis of the aorta, and is the most common cardiovascular manifestation of Marfan’s syndrome. Hartnell et al\(^2\) reported that of 4993 consecutive patients who underwent coronary angiography, 70 had coronary ectasia, one of whom had Marfan’s syndrome with a thoracic aortic aneurysm. McKeown\(^3\) reported a case of arachnodactyly complicated by medionecrosis of the right coronary artery, leading to a dissecting aneurysm without a dissecting or fusiform aneurysm of the aorta. Becker et al\(^6\) microscopically studied the coronary arteries of patients with Marfan’s syndrome, and found that the coronary artery changes closely resembled those frequently described as cystic medionecrosis for the aorta.
Dissecting aneurysms of the coronary arteries are most often secondary to dissecting aneurysm of the ascending aorta.\(^7\) According to Brody et al,\(^8\) primary dissecting aneurysms of coronary arteries may result either from cystic medionecrosis or from primary hemorrhage into the media, and pregnancy may play a role in their pathogenesis. Our patient had neither stigmata of Marfan’s syndrome nor signs of dissecting aneurysm of the aorta or coronary arteries. The relation between pregnancy and coronary ectasia is uncertain.

Cystic medionecrosis is seen in Marfan’s syndrome, in aortic dissection in pregnancy and in idiopathic aortic dissection.\(^9\) However, in addition to its relationship to advancing age, cystic medionecrosis was consistently more common in hypertensive subjects than in normotensive subjects.\(^10\) Because our patient had hypertension and atherosclerotic changes in both eye grounds, the finding may be relevant to the etiology of the condition. But our patient was not elderly, and did not have hyperlipidemia, diabetes mellitus, or a history of smoking.

We speculate that coronary ectasia may be caused by Takayasu’s arteritis. Takayasu’s arteritis is a chronic inflammatory arteriopathy of unknown etiology, which commonly affects the aorta, its main branch, and the pulmonary artery.\(^11,12\) Several investigators reported that dilative and aneurysmal changes of the ascending aorta are not infrequently found in Takayasu’s arteritis.\(^13-15\) Saito et al,\(^16\) in an evaluation of 16 autopsy cases of the disease, found that the ascending aorta and aortic arch were without exception dilated in those with long-standing disease. Annuloaortic ectasia may result from both the inflammatory lesion and the hemodynamic condition of the aorta which are found in Takayasu’s arteritis. Ishikawa\(^17\) reported that five of 96 patients with Takayasu’s arteritis showed annuloaortic ectasia, and it is one of the 12 proposed criteria for the clinical diagnosis of Takayasu’s arteritis. Coronary artery involvement was found in three of 20 patients\(^15\) and ten of 107 patients\(^12\) with Takayasu’s arteritis; it consisted mostly of stenotic lesions, especially coronary ostial stenosis, which is due to extension of the process of proliferation of the intima and contraction of the fibrotic media and adventitia in the aorta.\(^18,19\) Rose et al\(^20\) found coronary artery aneurysms in two of the 16 autopsy cases, but reported that it was apparently exceptional in Takayasu’s arteritis. There are thus few case reports of coronary aneurysm\(^21\) and ectasia\(^22\) in Takayasu’s arteritis. Unfortunately, in our 50-year-old patient, who, as indicated by the normal erythrocyte sedimentation rate, was not in the active inflammatory stage, angiography of the left subclavian artery and abdominal aorta was not performed. However, we noted a marked blood pressure difference between the arms, linear calcification of the aortic wall, and a stenotic lesion in the right mid-subclavian artery (Figure 2). It may be difficult to determine whether the coronary ectasia and annuloaortic ectasia are due to atherosclerosis or to Takayasu’s arteritis,\(^17\) but we suggest this is a rare
case of Takayasu’s arteritis with annuloaortic ectasia and coronary ectasia.

REFERENCES