A CASE OF SEVERE ANNULOAORTIC ECTASIA DETECTED COINCIDENTALLY FROM AN ANNUAL MEDICAL CHECK

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Abstract: This report describes a case of severe annuloarotic ectasia (AAE) who presented without any symptoms. A 33-year-old man was noted to have a cardiac murmur at an annual medical checkup. Evaluation of the cardiac murmur revealed severe AAE with moderate aortic regurgitation (AR). He underwent immediate surgical treatment successfully and remains in good clinical condition. Therefore, even in asymptomatic patients, the possibility of severe AAE should not be overlooked in patients with a murmur. Further, echocardiographic evaluation should be performed in such patients.
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Key words : annuloaortic ectasia, aortic regurgitaion, echocardiography

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The term annuloaortic ectasia (AAE) was first used by Ellis et al. in 1961 to describe a clinicopathologic condition seen in a subset of patients with thoracic aortic aneurysms in whom idiopathic dilation of the proximal aorta and the aortic annulus causes aortic regurgitation (AR)[1]. Insufficient coaptation of the aortic valve due to aortic root dilation may produce the AR. In patients with AAE, aortic valve replacement is performed not so rarely for the treatment of AR. In fact, patients with AAE make up about 5 to 10 percent of the patients undergoing surgical therapy for AR. The sudden onset of symptoms of severe AAE, followed by rapid hemodynamic compromise, is occasionally seen. Therefore, to improve the prognosis of patients with severe AAE, it is important to detect AAE in the early or asymptomatic stage. In this report, we describe an asymptomatic patient with severe AAE which was detected because of the presence of a cardiac murmur.

CASE REPORT

A 33-year-old man had been without complaint, until he was noted to have a cardiac murmur at the time of a routine annual medical checkup in July 1997. He was admitted to our hospital for evaluation of the cardiac murmur. There was no significant past medical history or family history. On physical examination, the patient was 174 cm tall and weighed 62 kg. The extremities appeared normal in length and no skeletal deformities were evident. Ophthalmoscopic examination revealed no evidence of lens displacement. The blood pressure was 118/64 mmHg and the pulse was regular at 70 beats per minute. He had a grade II/IV systolic ejection murmur and a grade III/VI diastolic blowing murmur, both of which were loudest in the third

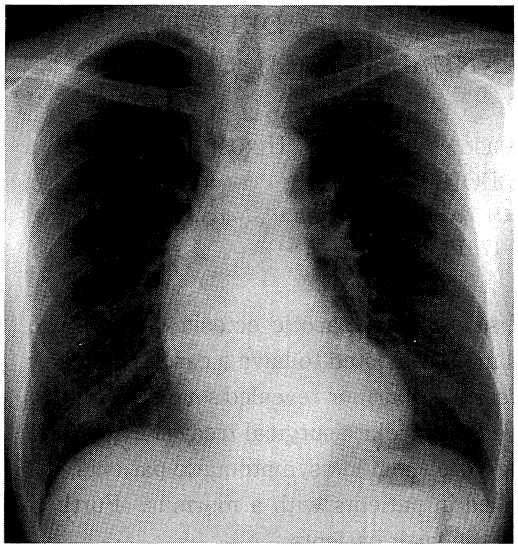

Fig. 1. Chest radiograph.
Chest radiograph showed a large bulge in the region of the ascending aorta. These is no evidence of cardiomegaly or pulmonary congestion.

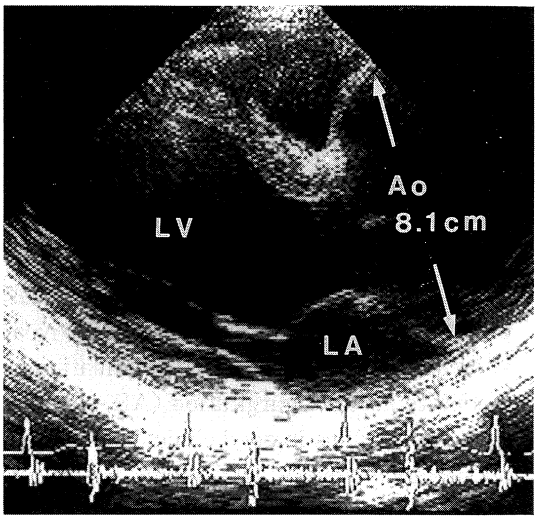

Fig. 2. Two-dimensional echocardiograph.
Two-dimensional echocardiograph showed a grossly dilated aortic root and asecending aorta associated with left ventricular enlargement.

intercostal space along the left sternal border. A chest radiograph showed a large bulge in the region of the ascending aorta, but there was no evidence of cardiomegaly or pulmonary congestion (Fig. 1). Electrocardiography revealed high voltage on the left lateral chest leads. Two-dimensional echocardiography revealed massive dilation of the ascending aorta. The diameter of the aorta was 8.1 cm at the level of the ascending aorta maximally, and 3.4 cm at the level of the aortic annulus. Left ventricular enlargement was also noted with a left ventricular end-diastolic dimension (LVDd) of 6.5 cm, a left ventricular end-systolic dimension (LVDs) of 4.8 cm, and a fractional shortening (FS) of 25 % (Fig. 2). Doppler flow studies revealed moderate aortic regurgitation, but no evidence of mitral regurgitation. Retrograde aortography confirmed the above findings. On January 9, 1998, the patient underwent Yacoub's type cusp sparing aortic remodeling, and the left and right coronary arteries were reimplanted using Carrel patch and Hemashield graft. Histologic examination of the ascending aorta demonstrated severe cystic medial necrosis. Based on these findings, the diagnosis of idiopathic cystic medial necrosis was made. The patient's postoperative recovery was uneventful. Postoperative retrograde aortography demonstrated no residual enlargement of the ascending aorta or AR. The patient was discharged one month after operation, and remained in good clinical condition 3 months after operation.

DISCUSSION

The common pathologic feature shared by patients with AAE is cystic medial degeneration of the affected aortic wall, causing progressive dilation[2]. AAE may or may not be accompanied by the stigmata of Marfan's syndrome. In the present case, the typical skeletal and

ophthalmic abnormalities of the disease were not observed. The AAE lesion poses a double threat to the patient's life. First, the thin-walled aneurysm may rupture, resulting in fatal hemorrhage. Second, the presence of severe aortic insufficiency may cause intractable congestive heart failure. It has been reported that in patients with AAE, the incidence of AR is significantly greater if the aortic root is >5 cm in diameter. Further, the incidence of aortic dissection increases if the aortic root is >6 cm in diameter[3]. Lemon and White found that two features, the acute or subacute development of symptoms and the presence of associated chest pain, were more common in patients with AAE than in patients with primary AR[4]. As in primary AR, the presence of congestive heart failure or chest pain indicates poor prognosis in patients with AAE[5]. Patients with AAE can die within several years from the onset of the above symptoms[6]. Therefore, in order to improve survival, it is important to detect AAE in the early or asymptomatic stage and to follow carefully. Moreover, it is essential that surgical treatment be performed before symptoms of congestive heart failure appear or the ascending aorta increases markedly in size. However, the lesion is difficult to detect in the early stage, particularly in patients without symptoms. In our patient, the cardiac murmur noted at a routine annual medical checkup led to the detection of severe, but asymptomatic, AAE. Severe AAE can exist without symptoms, because the associated AR may be relatively mild. The discrepancy between the degree of dilation of the ascending aorta and the severity of AR may be due to a rather small diameter of the aortic annulus. Therefore, asymptomatic AR in the setting of severe AAE might not be so rare. In conclusion, even for asymptomatic cases presenting with isolated cardiac murmurs, an evaluation using echocardiography may be necessary.

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