IDIOPATHIC CYSTIC MEDIONECROSIS AND ANEURYSMS OF THE ASCENDING AORTA

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SUMMARY

Two patients with ascending aortic aneurysms due to cystic medical necrosis are described. One of them was phenotypically normal while the other had features of Marfan's syndrome. Both were disabled by dyspnoea and angina which required corrective surgery.

INTRODUCTION

Cystic medionecrosis involving the aorta frequently manifests itself in a dramatic manner as a dissecting aneurysm. [1,2] It may occur in pregnancy, [3] Marfan's syndrome, [2,4,] and coarctation of the aorta.

A common manifestation of this condition is aneurysmal dilatation of the ascending aorta. [5,6,7] This is a disease of relatively young men and, if left untreated leads to severe aortic regurgitation, heart failure and death. Early diagnosis is important because, unlike rheumatic aortic regurgitation, progression of disease is rapid and early death is frequent.

We report on two patients with aneurysms of the ascending aorta due to idiopathic cystic medionecrosis. Both patients were young men who presented with rapidly progressive symptoms of heart failure.

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CASE I

A 31 year old Chinese male was initially seen at the University Hospital in 1970 and was then asymptomatic. Rheumatic fever and family history of heart disease were denied. Clinical examination revealed a normal looking adult male with no features of Marfan's syndrome. His pulse was 80/min, blood pressure 120/70 mmHg. Examination of the heart revealed mild cardiomegaly and a soft early diastolic murmur of aortic regurgitation. An electrocardiogram showed left ventricular hypertrophy and chest radiographs showed a large heart and moderate dilatation of the ascending aorta. A diagnosis of aortic incompetence secondary to a bicuspid aortic valve was made. He remained in good health until 1975 when he began to experience frequent attacks of retrosternal pain radiating up the neck on exertion or after heavy meals. Occasionally, the chest pain occurred at rest. He also noted that his exercise tolerance gradually worsened. He was seen at the hospital a year later. On examination his peripheral pulses were frankly collapsing in nature; blood pressure 160/65 mmHg. There was marked left ventricular dilatation clinically. Gross aortic regurgitation associated with an Austin-Flint murmur was evident.Electrocardiography revealed left ventricular hypertrophy with strain. Chest radiography revealed cardiomegaly and marked dilatation of the ascending aorta (Fig. 1) with filling- in of the superior retrosternal free space by the ascending aorta. Serological tests for syphilis were negative. Echocardiography revealed a dilated left ventricle, poorly-moving interventricular

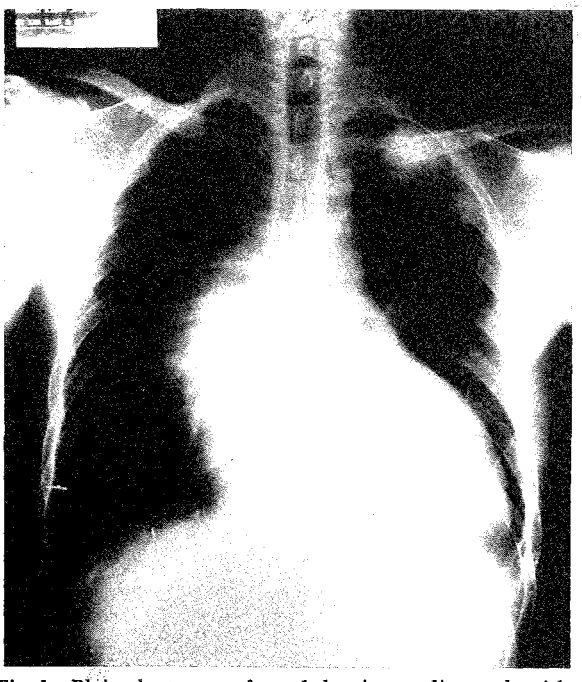

Fig. 1 Plain chest x-ray of case 1 showing cardiomegaly with marked dilatation of the ascending aorta.

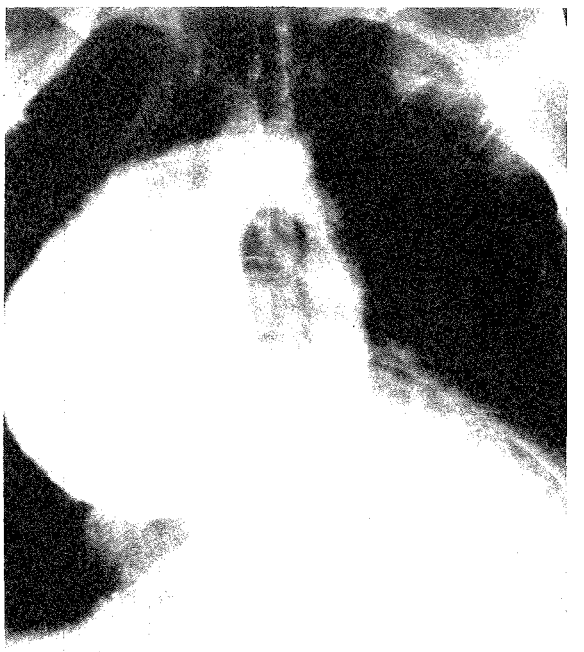

Fig. 2 Ascending aortogram of case 1 showing an aneurysm of the ascending aorta. There is severe reflux of contrast into the left ventricle.

septum and normal mitral valve motion.

At aortography, gross dilatation of ascending aorta, involving the aortic root and the arch just proximal to the origin of the innominate artery was confirmed (Fig. 2). The aortic wall was smooth and no aortic dissection was noted. Ascending aortic cineangiogram revealed gross aortic regurgitation, and normal-sized aortic arch distal to the aneurysm.

The patient's cardiac failure did not respond satisfactorily to digoxin and diuretics but his chest pain was controlled with isosorbide dinitrate. He underwent surgery in December 1976. At operation, the ascending aorta was aneurysmally dilated. A Starr-Edwards aortic valve prothesis with dacron support of the ascending aorta was done. Unfortunately the patient was unable to come off cardio-pulmonary by-pass. Histopathological examination of the resected aortic tissue confirmed the presence of idiopathic cystic medionecrosis.

CASE 2

A 27 year old male meter reader was in good health until 1975 when he began to notice rapidly worsening dyspnoea on exertion. About 18 months later he began to have severe retrosternal chest pain precipitated by moderate exertion, emotional stress and heavy meals. There was no history of previous rheumatic fever and no family history of heart disease. On direct questioning he recalled hypermobility and hyperflexibility of joints ever since childhood. No visual difficulties were ever noted. Examination of his eyes revealed no lenticular subluxation. His upper limbs exhibited arachnodactyly with marked hyperextensibility of the metacarpophalangeal and interphalangeal joints. The knees demonstrated bilateral genu valgii and hyperextensibility. His peripheral pulses were collapsing in nature; blood pressure was 155/40 mmHg. Gross cardiomegaly with severe aortic regurgitation was noted on examination of his heart. Electrocardiography revealed first degree heart block and left ventricular hypertrophy. Chest radiographs revealed a widened aortic root and severe cardiomegaly (Fig. 3). Serologic test for syphilis was negative. Echocardiography showed a dilated left ventricle and decreased ejection fraction. Fluttering of the anterior mitral valve leaflet in diastole was noted. The aortic root was markedly dilated, measuring 6.5 cm in diameter.

Cardiac catheterization revealed increased left atrial mean and left ventricular end-diastolic

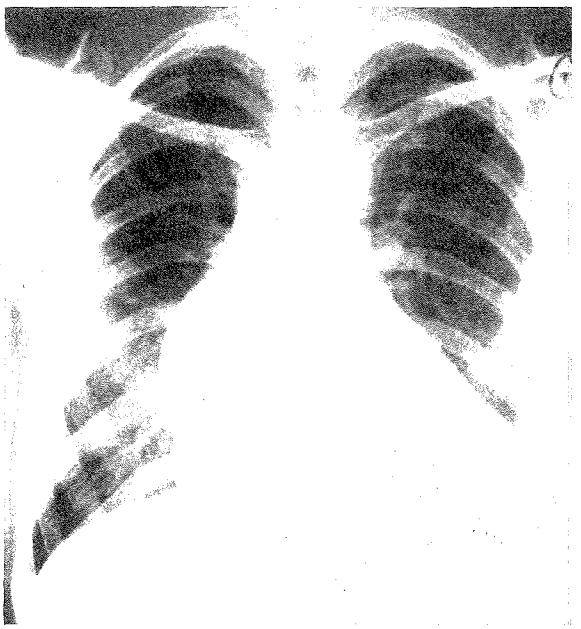

Fig. 3 Plain chest x-ray of case 2 showing gross cardiomegaly with marked dilatation of the ascending aorta.

pressures and a decreased left ventricular ejection fraction of 56%. Left ventricular cineangiocardiography showed no mitral reflux. Aortography demonstrated a large saccular aneurysm of the ascending aorta and gross aortic regurgitation (Fig. 4 and 5). The patient responded well to anti-heart failure therapy and his chest pains were controlled with isosorbide dinitrate and glyceryl trinitrate. In July 1978 he underwent successful aneurysmectomy and aortic valve with ascending aortic replacement (stented aortic homograft). At operation, the pericardial space was free. Marked left ventricular dilatation was noted. A fusiform (maximum diameter 10 cm) ascending aortic aneurysm affecting the aortic valve ring and all three sinuses of valsalva was found. The aortic valve was tricuspid and aortic regurgitation was secondary to dilatation. Histopathological examination of the resected portion of aorta revealed cystic medionecrosis. Following his operation, the patient made remarkable symptomatic improvement and was able to return to normal work. A year post-operatively he was still asymptomatic but mild aortic regurgitation was detected. He began to have deterioration of cardiac function the following year and progressively worsened despite therapy for heart failure. He died from acute pulmonary oedema in late May 1982, almost four years after

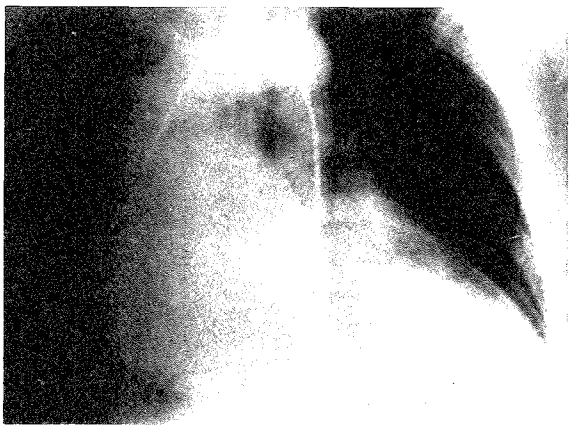

Fig. 4 Ascending aortogram (antero-posterior view) showing the large fusiform aneurysm of the ascending aorta with aortic regurgitation.

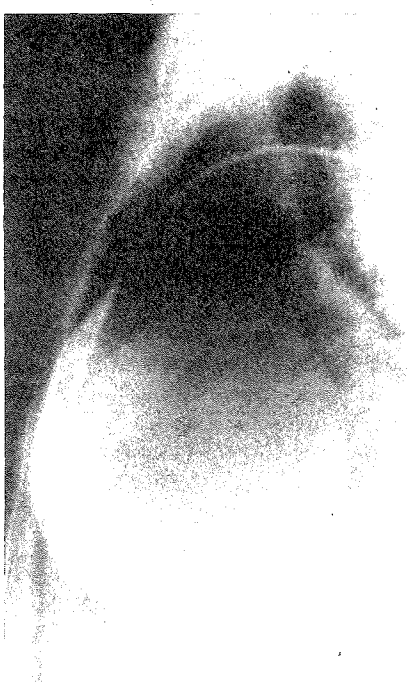

Fig. 5 Ascending aortogram (lateral view) showing the large fusiform aneurysm of the ascending aorta with aortic regurgitation.

aortic valve replacement.

DISCUSSION

Aneurysms of the ascending aorta have been known as "aneurysms of symptoms," [8] the outstanding symptoms being pain, dyspnoea and

cough. The average duration of illness in patients with aortic aneurysms has been reported to vary from 6 months [8] to over 2 years. [9] Death is usually due to rupture, pressure on adjacent vital structures or heart failure.

In both cases, idiopathic cystic medionecrosis of the aorta was the underlying process responsible for the aneurysmal dilatation of the ascending aorta. Aortic regurgitation was secondary to dilatation of the aortic ring. Both patients presented with symptoms of left ventricular failure and ischaemic cardiac pain secondary to the severe aortic regurgitation. This must be differentiated from the deep, diffuse aching sensation which many patients with large aortic aneurysms frequently complain of. The latter type of pain may be due to pressure on surrounding structures, including erosion of the sternum, thoracic vertebrae or ribs. One of our patients had a phenotype compatible with Marfan's syndrome. Whether idiopathic cystic medionecrosis is aetiologically related to Marfan's syndrome is still not clear. Layman and Yang Wang [4] noted that none of their 10 patients had a phenotype compatible with Marfan's syndrome, and yet four of eight cases studied had abnormally high urinary hydroxyproline excretion. Typical cystic medionecrosis was proven histopathologically in seven of 10 patients.

Idiopathic cystic medionecrosis is a frequent underlying cause of aneurysmal dilatation of the aorta. The disease occurs most commonly in young males and manifests as left heart failure with a rapidly deteriorating course. In view of this, early detection and surgical repair of aneurysm and replacement of the aortic valve is necessary.

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