Syphilitic ostial occlusion

S. HOLT

From Liverpool Regional Cardiac Centre, Liverpool

A patient presenting the picture of congestive cardiomyopathy was found to have syphilitic obliteration of the left coronary artery. Aortic regurgitation, angina, and myocardial infarction were notably absent.

Syphilitic stenosis or obliteration of the coronary ostia is a rare disease (Schröter *et al.*, 1966). Heggteit (1964) reviewed the major clinical and pathological features of 100 cases of syphilitic aortitis derived from a necropsy series of 13 082 patients and found syphilitic coronary ostial stenosis or occlusion to be present in 26 cases. Bruenn (1934) analysed the clinicopathological findings in 118 cases of cardiovascular syphilis to define the role of syphilitic disease of the coronary arteries in relation to the general picture of cardiovascular syphilis. Of 118 cases described, 39 (33%) had coronary ostial stenosis. Of these 39 cases, 34 (87%) had aortic regurgitation; the right coronary ostium was found to be stenosed eight times more commonly than the left.

A patient with syphilitic obliteration of the left coronary ostium presented with clinical features suggesting a diagnosis of congestive cardiomyopathy.

Case report

A 49-year-old car mechanic complained of progressive exertional breathlessness for 6 weeks. He claimed previous good health and in particular denied any history of chest pain or excessive alcohol consumption. He showed oedema, raised venous pressure, sinus rhythm, and a blood pressure of 120/80 mmHg. The apical impulse was sustained, forceful, and displaced to the left, but there was no parasternal lift. There was a summation gallop but no sign of aortic or mitral valve disease. The jugular venous pressure was raised to 6 cm above the clavicle but showed a normal wave form. The electrocardiogram showed left ventricular hypertrophy with the pattern of left anterior hemiblock. The radiological cardiothoracic ratio was 65 per cent. The ascending aorta was prominent and no valvular or aortic calcification was visible. A diag-

nosis of myocardial disease of uncertain cause was made. The heart failure completely responded to treatment in hospital within two weeks.

Three months later, despite maintenance treatment, the fluid retention recurred, with symptoms of acute left ventricular failure but again without chest pain. Clinical examination at this stage indicated severe congestive cardiac failure with mild icterus. There was pronounced jugular venous engorgement with dominant V waves in the jugular venous pulse. A summation gallop rhythm was present and a midsystolic murmur was audible at the right sternal edge, which increased in intensity on deep inspiration, suggesting tricuspid valvular regurgitation. The electrocardiogram and chest film were unchanged. Persistent slight rise in serum aminotransferase enzymes was considered to reflect hepatic congestion. A pyruvate tolerance test gave a normal result. Blood cultures were repeatedly negative. Antinuclear factor was not detected and lupus erythematosus cells could not be seen. Serological tests for syphilis were found to be strongly positive, shortly before the patient's sudden death from asystole.

Necropsy showed syphilitic aortitis of the ascending aorta but a normal aortic valve. The left coronary ostium was totally obliterated by fibrous tissue and the left main coronary arterial lumen was narrow but patent 5 mm from the ostium (Fig.). The left coronary arterial branches showed no sign of atheroma or other occlusive disease. The right coronary ostium and artery were large and healthy. There was no histological evidence of syphilitic periarteritis or endarteritis of the coronary arteries.

The heart was enlarged, weighed 620 g, and was hypertrophied with dilatation of the ventricle. There was moderate but widespread subendocardial fibrosis. Mural thrombi and amyloid material were absent. There was no evidence of syphilis outside the cardiovascular system.

Fig. The ostium of the coronary artery has been occluded by inflammatory tissue.

Discussion

Syphilitic cardiovascular disease occurs more frequently than is recognised clinically. An accurate clinical diagnosis was established in only 17 per cent of a large group of cases of syphilitic aortitis reviewed in a clinicopathological necropsy study (Heggveit, 1964). Beck *et al.* (1965) emphasised the importance of suspecting syphilitic ostial occlusion when angina is disproportionate in the presence of aortic regurgitation, but in this patient there was no history of angina pectoris and no clinical or necropsy evidence of aortic valve involvement. In Bruenn's series (1934) 62.5 per cent of cases with stenosed coronary ostia experienced cardiac pain, the inference being that the absence of angina pectoris does not rule out the presence of such stenosis.

Virtually all patients with luetic aortitis will have hearts which display some degree of interstitial fibrosis of the myocardium (Heggveit, 1964). It is uncertain whether myocardial fibrosis in these cases results from ostial stenosis, or whether it is due to syphilitic myocarditis or coexistent coronary atherosclerosis. It seems likely that the subendocardial

fibrosis seen in this patient was a significant factor causing progressive cardiac failure. The relation of ostial stenosis to so-called syphilitic myocardial disease is unclear (Warthin, 1922; Saphir and Scott, 1930; Storey, 1958). Since a large proportion of previously described cases was complicated by aortic regurgitation and coronary atheroma, it is difficult to assess the contribution of ostial stenosis to left ventricular hypertrophy and myocardial fibrosis. Congestive cardiac failure was found in 8 of 15 cases of syphilitic ostial stenosis (Love and Warner, 1934), and was even the presenting feature in 2, but it was not specified whether these patients had aortic regurgitation or coronary disease in addition. Heggveit (1964) reports congestive heart failure in 7 of 11 patients with ostial stenosis without aortic regurgitation but only 3 of 11 had clinical cardiac enlargement and 2 had hypertension.

If coronary ostial stenosis or occlusion can give rise to myocardial failure, in the absence of aortic regurgitation, angina, and myocardial infarction, then this entity will need to be included in the differential diagnosis of congestive cardiomyopathy (Oakley, 1974), particularly in the younger adult.

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Requests for reprints to: Dr. Stephen Holt, University Department Therapeutics, The Royal Infirmary, Edinburgh EH3 9YW.