SYPHILITIC CORONARY OSTIAL OCCLUSION*

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Syphilitic ostial occlusion or stenosis is a rare disease. Scharfman *et al.*,[1] in 1950, reviewed several large necropsy series and analysed 935 cases of syphilitic aortitis in 33,176 necropsies. The coronary ostia were involved in 24. Heggtveit,[2] more recently, reviewed 100 cases of syphilitic aortitis in a necropsy series of over 13,000 patients from Brooklyn, New York. Coronary ostial stenosis occurred in 26, i.e. in more than a quarter of the patients. The right ostium was involved in 5, the left in 6 and both in 15 cases. The involvement was marked in 15 of these cases, and syphilitic coronary arteritis was present in only 1 case. Cardiac infarction was present in 24 patients. In 17, coronary atherosclerosis was responsible and in 3 both syphilis and atherosclerosis were implicated. Syphilitic ostial occlusion was the main or sole factor in 4 of these. Sudden death due to ostial stenosis occurred in 5. Heggtveit rightly drew the conclusion that cases of syphilitic coronary ostial stenosis are particularly prone to be regarded simply as examples of coronary atherosclerosis.

With the development of cardiac bypass, surgical cure is now possible. Successful treatment by coronary ostial endarterectomy has previously been reported by French workers[3,4] in 5 patients and by ourselves[5] in 2 patients. Connolly *et al.*[6] published a report on a patient in whom ostial occlusion was thought to be due to atheroma, diaphragmatic infarction developing after the procedure.

It is important to think of syphilis as a cause of coronary disease. There may be no clinical clues and the ECG is unhelpful. Of a series of over 150 patients with syphilitic heart disease seen by one of us (V.S.) in the Cardiac Clinic, Groote Schuur Hospital, no more than 5 have been clearly recognized as suffering from coronary ostial occlusion. The diagnosis can now be readily made by coronary arteriography. This procedure was of particular value in a patient with atypical chest pain and progressive heart failure in whom surgery was curative.

CASE REPORT

C.F., a 54-year-old Coloured male, was first seen in the Cardiac Clinic at the age of 48 complaining of left-sided chest pain, burning in nature and first noticed while lifting a heavy weight at work. The pain was not generally related to effort. Clinical examination was negative, the blood pressure was 150/90 mm.Hg and the X-ray was normal. Repeat ECGs showed transient flattening of the T waves in V1 and V6. An unequivocally positive effort test was obtained (Fig. 1). A diagnosis of atypical chest pain with latent coronary artery disease was made.

The patient was lost to follow-up for 5 years, and was then referred back after mass radiography had shown cardiomegaly. He then stated that since his first examination the left-sided chest pain had continued, lasted about ½-hour at a time, could be provoked by effort and was relieved by rest and TNT. This symptom had remained stationary, but for several years he had become increasingly dyspnoeic on effort so that he had been forced to alter his occupation. For 1 month before admission, he had characteristic attacks of paroxysmal cardiac dyspnoea associated with slight left chest pain. This was followed by swelling of the feet. There had been no prolonged attacks to suggest cardiac infarction. On examination he had definite cardiomegaly, with a loud apical triple rhythm.

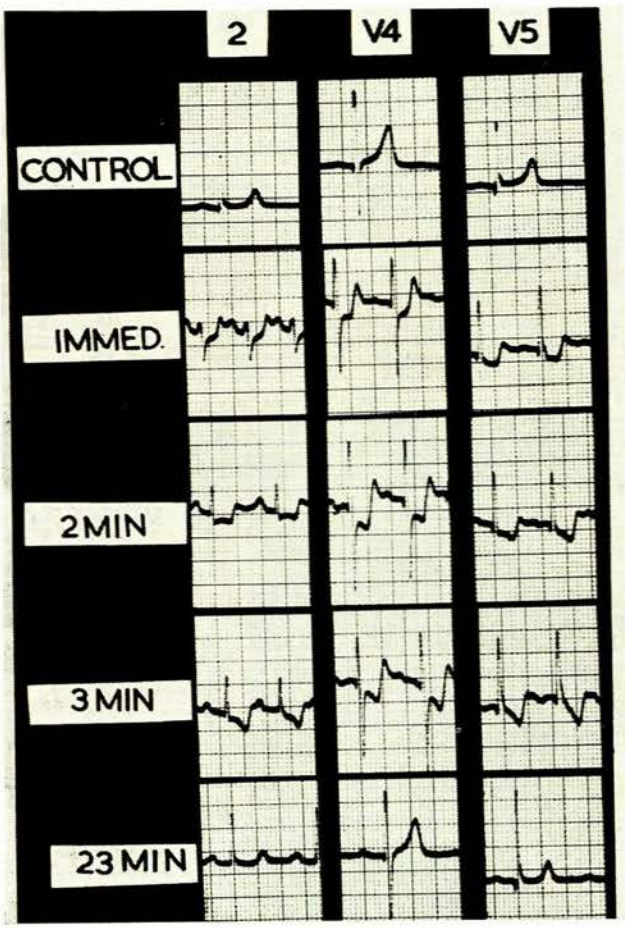

Fig. 1. The resting ECG is normal. Immediately after effort pathological left axis deviation develops, which soon returns to normal. Unequivocal ST segment depression due to cardiac ischaemia develops.

The X-ray confirmed the cardiomegaly, and showed well-marked pulmonary congestion. The ECG had altered, showing pathological left axis deviation, with a widened QRS and abnormal T waves suggesting anterolateral parietal block (Fig. 2).

He was admitted for treatment and study. After digitalization and diuretics, there was a good response. A soft, intermittent, early diastolic murmur became audible in the fourth left space. The serology was positive and the sedimentation rate was 8 mm. per hour. A diagnosis of luetic coronary ostial disease with cardiac ischaemia and slight aortic incompetence was made.

Cardiac catheterization confirmed the presence of mild aortic incompetence with a raised end-diastolic left ventricular pressure—145/5-25. The right coronary artery filled well, but there was narrowing of the lumen at or close to the ostium.

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The left coronary appeared diminutive and filled late and retrogradely from the posterior-intraventricular branch of the right coronary artery (Fig. 3). Ostial occlusion of the main left coronary artery was demonstrated and dilatation of the ascending aorta was present.

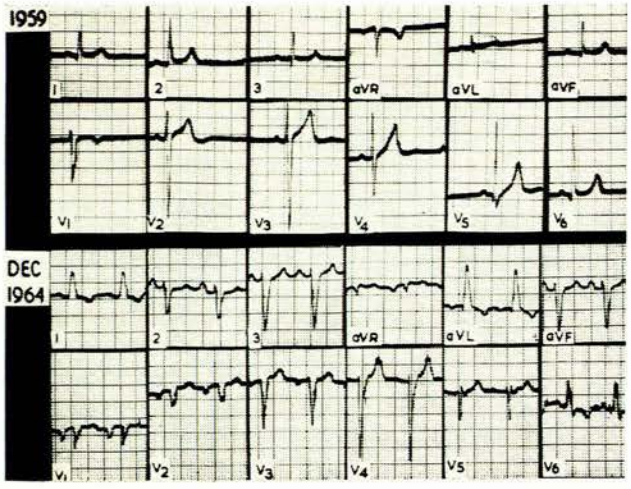

Fig. 2. The two ECGs done at an interval of 5 years are contrasted. The initial tracing is within normal limits; the later tracing shows pathological left axis deviation with a widened QRS and abnormal T waves suggesting anterolateral parietal block.

A week later, surgery was performed with cardiac bypass. Dilatation with several saccular aneurysms of the aorta was found, the aorta being adherent to the surrounding structures. Severe intimal changes with calcification were present. The left coronary ostium was completely occluded and the right ostium partly occluded by syphilitic endarteritis. Minimal aortic incompetence was present. The saccular aneurysms were resected. Great difficulty was encountered in finding the orifice of the left coronary artery. After extensive endarterectomy this area was located. The small orifice was completely cleaned up with good back-flow from the right coronary artery. This artery was not endarterectomized. The postoperative course was complicated by severe mental depression which gradually settled.

Within 2 months of surgery, the patient's condition improved strikingly, all chest pain disappeared and he was free of dyspnoea. The early diastolic murmur was barely audible and the gallop rhythm had disappeared. There was a clear reduction in heart size (Fig. 4). No cardiac therapy was required.

Reinvestigation 3 months after surgery showed patent coronary arteries. Both vessels could now be selectively catheterized. The ostium of the left coronary was half the size of the right, but filled normally (Fig. 5). Collateral filling of the left coronary was no longer present. An aortic angiogram (Fig. 6) showed both coronary arteries filling from the aorta and trivial aortic incompetence was present. The left ventricular pressure curve, however, had not yet returned to normal (LVP 160/15 - 27) and the electrocardiogram was unchanged.

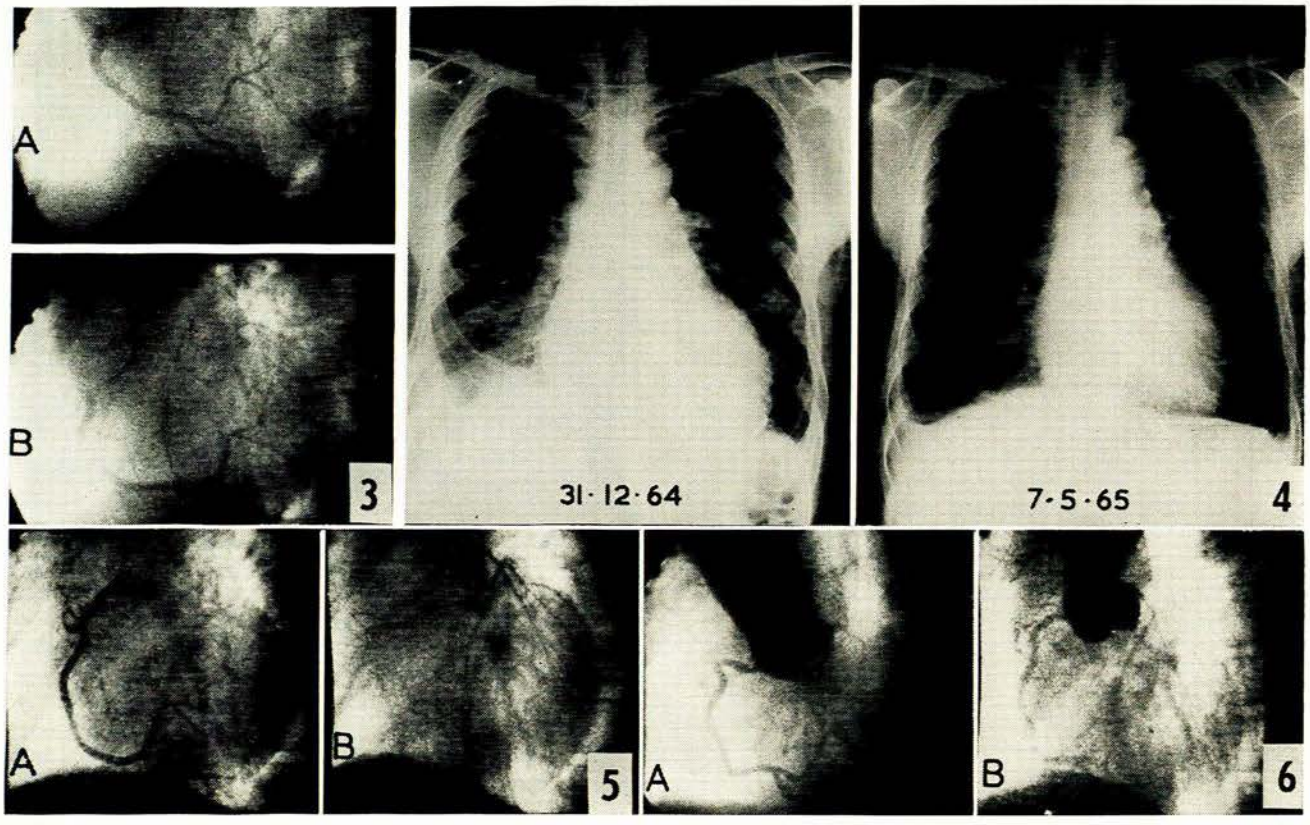

Fig. 3. Selective coronary angiography shows a dominant right coronary artery which is markedly enlarged. The left coronary is diminutive and fills late and retrogradely from the posterior intraventricular branch of the right coronary.
Fig. 4. The marked reduction in heart size and disappearance of pulmonary venous congestion is shown within the short space of 3 months after surgery.
Fig. 5. Selective coronary angiography postoperatively now shows patency of both coronary ostia. Although the left coronary artery is small, it fills directly from the aorta and the retrograde anastomotic channels have disappeared. Note the reduction in the size of the left ventricle.
Fig. 6. An aortic angiogram before and after surgery is contrasted. Before operation only the right coronary artery fills and is enormously dilated. After operation both coronary vessels fill directly from the aorta. Trivial aortic incompetence is present.

DISCUSSION

Cardiovascular syphilis has become a rarity in most countries, since the introduction of penicillin treatment has all but eliminated the tertiary manifestations of the disease. In South Africa, however, syphilis still remains an important cause of heart disease. Thus it accounts for 0·6% of all cases of cardiovascular disease at Groote Schuur Hospital.[7] In non-Whites 1 - 2% of heart disease is due to syphilis—10 times higher than in Whites. Moreover, recent reports in the literature suggest that syphilis is again on the increase.[8]

Syphilis should always be considered when angina pectoris is encountered in the younger age-group, especially if other evidence of aortitis is present. Disproportionate angina in the presence of mild aortic incompetence is strongly suggestive of this condition. Positive serological tests are supportive evidence, but may be an incidental finding in a patient with coronary atherosclerosis.

The only certain way of establishing the diagnosis is by coronary angiography; the obstruction is localized to the coronary ostium, the rest of the coronary vascular tree being normal. The appearance is quite unlike coronary atherosclerosis where the obstructions are generally multiple and situated distal to the ostia.

A full course of penicillin may sometimes be of value but has been very disappointing in our experience. Surgery is required to relieve the narrowed coronary ostia. It was of considerable benefit to our patient, even though the main manifestations were those of progressive heart failure due to myocardial disease. Presumably restoration of coronary blood flow relieved myocardial ischaemia and the heart failure was reversible.

SUMMARY

A case of atypical left chest pain, probably anginal in nature, with progressive heart failure culminating in acute paroxysmal nocturnal dyspnoea and severe disability, has been described. Auscultation initially revealed a gallop rhythm only, but later with control of the heart failure a soft early diastolic murmur became audible. Syphilitic coronary ostial disease was confirmed by coronary arteriography, normality restored by surgery, and a striking benefit resulted for the patient.

Syphilitic ostial disease must always be considered if angina pectoris occurs in a young man; if aortic incompetence is present; if the ascending aorta is dilated or calcified; or if disproportionate angina pectoris is present in the presence of aortic incompetence.

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REFERENCES

1. Scharfman, W. B., Wallach, J. B. and August, A. (1950): Amer. Heart J., **40**, 603.
2. Heggtveit, H. A. (1964): Circulation, **29**, 346.
3. Blondeau, P. and Dubost, C. L. (1962): Ann. Chir. thorac. cardiov., **1**, 802.
4. Michau, P., Froment, R., Pont, M., Saubier, E. and Aimard, G. (1963): Lyon chir., **59**, 575.
5. Beck, W., Barnard, C. N. and Schrire, V. (1966): Brit. Heart J., **27**, 911.
6. Connolly, J. E., Eldridge, F. L., Calvin, J. W. and Stemmer, E. A. (1964): New Engl. J. Med., **271**, 213.
7. Schrire, V. (1964): S. Afr. Med. J., **38**, 598.
8. US Department of Health, Education and Welfare (1962): *VD Fact Sheet*. Publication no. 341. Washington, DC: Dept. of Health, Education and Welfare, Public Health Service.