Severe isolated left mainstem coronary artery stenosis

A case report

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Summary

A 33-year-old white man had exertional angina pectoris, followed by angina pectoris at rest, and then episodes of ischaemic acute pulmonary oedema associated with angina pectoris. Selective coronary angiography delineated an isolated long-segment stenosis of the left mainstem coronary artery with no other lesions. We believe that the mainstem obstruction was due to coronary artery fibromuscular hyperplasia, a condition rarely affecting the coronary artery circulation. At operation three coronary artery bypass grafts were inserted, one to the left anterior descending artery and two to the left circumflex coronary artery, with a most successful result. The patient's recurrent acute pulmonary oedema was due to severe myocardial ischaemia; the possibility of superadded coronary vasospasm aggravating the obstruction cannot be entirely discounted.

Case report

Clinical presentation

The patient was a 33-year-old white farmer from South West Africa (SWA)/Namibia. He was a non-smoker and had no other risk factors for ischaemic heart disease (IHD). In February 1984 he began to experience classic effort-induced angina pectoris. Early in September 1984 the angina pectoris began to come on with minimal effort as well as at rest; dyspnoea was associated with the episodes of chest pain. Because of this he decided at last to approach his general practitioner, who carried out an exercise electrocardiogram (ECG) on 16 September 1984. The resting ECG was entirely normal but minimal effort caused chest pain and dyspnoea accompanied by 1 mm downward-sloping ST-segment depression in the anterolateral leads. He was promptly given sublingual nitroglycerine with rapid relief and return of the ECG tracing to normal. A diagnosis of IHD was made and he was given nifedipine which provided some symptomatic relief, although nitrate therapy and nifedipine 20 mg 3 times daily. He responded quite dramatically to treatment and acute myocardial infarction was excluded by serial serum enzyme estimation and ECGs. A few days after discharge he asked a friend to drive him from SWA down to the Western Cape; during the journey he had two further episodes of severe angina pectoris at rest accompanied by dyspnoea and haemoptysis characteristic of acute pulmonary oedema.

On admission to the Intensive Coronary Care Unit at Tygerberg Hospital on 17 October 1984 there was no evidence of cardiac failure and both the resting ECG and chest radiograph were still normal. Serum enzyme levels were also within the normal range.

Cardiac catheterization

This investigation was undertaken on 17 October 1984. Left ventricular cine angiography in the right anterior oblique projection revealed no abnormality. Selective coronary angiography revealed a dominant right coronary artery with no evidence of any atherosclerosis (Fig. 1) or of right-to-left collateralization, but there was a long and severe 80-90% internal luminal stenosis of the left mainstem coronary artery (Figs 2 and 3). However, the left anterior descending and left circumflex coronary arteries were angiographically free of disease. During angiography of the left coronary artery the patient experienced recurrent shortlived chest pain which responded briskly to sublingual nitroglycerine. Nitrate and sublingual nifedipine administration to exclude possible coronary artery spasm caused no change in the stenosis. The catheterization was completed without complication. In view of the coronary angiographic findings, highly suggestive of fibromuscular hyperplasia, i.e. non-atheromatous coronary artery disease, urgent cardiac surgery was carried out on 19 October 1984.

The cardiac operation

At operation with cardiopulmonary bypass no evidence of any significant atherosclerosis could be found in the coronary arteries. After successful creation of three saphenous vein coronary bypasses the left mainstem coronary artery was explored. This vessel was seen to lie posteriorly in fibrotic tissue posterior to the main pulmonary artery. Biopsy of the left mainstem coronary artery would have required transection of the main pulmonary artery, and this procedure was considered too risky; small biopsy specimens of the ascending aorta were therefore taken. These specimens only showed minimal atherosclerosis, in keeping with the patient's age. The operation was completed without complication.

Postoperative course

Special investigations excluded postoperative acute myocardial infarction. The patient made an uneventful recovery and was discharged approximately 1 week later. At discharge his medication results of side-room tests, chest radiography and hepatic and renal function tests were also normal.
Fig. 1. Right coronary cine angiograms in the (a) left anterior oblique and (b) right anterior oblique projections. The coronary artery is angiographically free of atherosclerosis and there is no right-to-left collateralization.

Fig. 2. Left coronary cine angiograms in the (a) shallow left anterior oblique view with cranial angulation and (b) shallow right anterior oblique view with caudal angulation. A severe 80-90% long-segment lesion (arrowed) is visualized in the left mainstem coronary artery. Both the left anterior descending (lad) and left circumflex (lc) coronary arteries appear normal.

Consisted of dipyridamole 200 mg 3 times daily and 1 tablet of aspirin daily. His maintenance nitrates and nifedipine therapy were discontinued. Eight weeks after surgery an effort ECG was negative. He is now entirely free of symptoms and is leading a normal life.

Discussion

This patient had no known risk factors for IHD, and the cine angiographic features of a single localized and severe left mainstem stenosis were against atherosclerosis since it has been shown repeatedly that atherosclerotic left mainstem disease is usually associated with significant narrowings in the other coronary arteries, although a few cases of isolated atherosclerotic left mainstem disease have been documented. Thus, non-atherosclerotic coronary artery disease must be seen as a cause of this young man's lesion.

Our patient did not have non-atheromatous left coronary ostial stenosis known to follow healing of aortitis in diseases such as syphilis, Takayasu's disease and rheumatoid arthritis. Diseases known to affect the left mainstem coronary artery (without ostial involvement) in isolation, such as polyarteritis nodosa, systemic lupus erythematosus, and acute rheumatic fever, are rare. Congenital left mainstem stenosis, atresia, and hypoplasia occur but these conditions present at a much earlier age.
The most attractive aetiological possibility is fibromuscular hyperplasia of the left mainstem coronary artery, a very rare condition more common in females and invariably associated with similar involvement of the renal arteries.\(^{20}\) Brill et al.\(^{21}\) documented acute myocardial infarctions secondary to this lesion in 2 young sisters, but it is debatable whether their cases were not of 'coronary intimal fibrous stenosis', an early stage of coronary atherosclerosis.\(^{22}\) The chronic use of methysergide is a most unusual cause of coronary artery fibromuscular hyperplasia.\(^{23}\)

A most intriguing complication was the recurrent pulmonary oedema after the onset of severe angina pectoris at rest. The precipitation of acute pulmonary oedema by global myocardial ischaemia due to increased myocardial oxygen consumption superimposed on fixed narrowing of the subtotal left mainstem is understandable, but the fact that these episodes took place at rest must raise the possibility of superadded functional obstruction by coronary artery vasospasm. If the patient did have fibromuscular hyperplasia, spasm of such a coronary artery could be powerful and probably more effective in diminishing coronary blood flow than spasm in an atherosclerotic artery. Left mainstem coronary artery spasm is rare and potentially lethal,\(^{24-28}\) it was probably excluded by the failure of administration of nitrates and nifedipine during coronary arteriography to dilate the narrowed segment.

The choice of management in isolated left mainstem obstruction is not very wide, since medical therapy has been shown to be inferior to surgery.\(^{27-31}\) Some 20 years ago various 'coronary plasty' operations using saphenous vein patches were attempted, but the high complication rate led to their discontinuation.\(^{32,33}\) Nevertheless, Hitchcock et al.\(^{36,37}\) recently reintroduced this technique in isolated left mainstem and left anterior descending coronary artery stenosis and obtained excellent results, which they mainly attributed to drastically improved intra-operative myocardial protection methods. We have attempted this technique in a few patients at Tygerberg Hospital but have so far been disappointed with our results. Because of this limited experience, routine saphenous coronary artery bypass grafting was carried out in this young man.

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REFERENCES

Hyponatraemia complicating the treatment of myxoedema coma

A case report

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Summary

Hyponatraemia is a common complication of severe hypothyroidism, but is usually readily corrected by administering L-thyroxine. A case of myxoedema coma in which the serum sodium level dropped precipitously when therapy was started is described and it is suggested that this was due to a state of relative hypocortisolism. Serum sodium values should be closely monitored in myxoedema treated with L-thyroxine; their decrease may signal the onset of an Addisonian-like crisis requiring urgent corticosteroid supplementation.


Hyponatraemia commonly accompanies severe myxoedema. The probable causes include dilutional hyponatraemia due to impeded water excretion, inappropriate secretion of antidiuretic hormone, and corticosteroid deficiency. If the serum sodium level is initially low the first change to occur when thyroid replacement therapy is started is a rise in the sodium level. Restoration of the euthyroid state may occasionally exaggerate a state of relative hypocortisolism; a clue to this is a drop in the serum sodium level, as illustrated by the following case.

Case report

A 61-year-old woman presented with mental confusion of recent onset. The patient was semicomaose and unable to give an account of her illness. Physical examination revealed the typical features of myxoedema with coarse skin, puffy facial features and loss of eyebrows. The pulse rate was 72/min and the blood pressure 210/90 mmHg with hypertensive changes in the ocular fundi. Initially, the serum sodium value was normal (Fig. 1), the potassium level low and the blood sugar level elevated to the diabetic range. The serum tri-iodothyronine level at that time was 2.6 pmol/l (normal 3-8 pmol/l) and the thyroxine level 9.9 pmol/l (normal 8-22 pmol/l) with a high thyroid-stimulating hormone (TSH) level of 7.5 IU/l (normal 0.7-1.7 IU/l). L-thyroxine 0.1 mg daily was given by mouth. No hypoglycaemic agents were given except for a low-energy diet. After a few days the patient’s mental state improved markedly until she was able to respond to questions. Within 6 days of starting L-thyroxine, however, the serum sodium level had begun to decline, reaching its nadir on day 9 at 125 mmol/l, with potassium 7.6 mmol/l, at which time the patient was confused again. Random measurements of serum cortisol on two occasions during this period gave normal results (512 and 464 nmol/l respectively). Oral administration of sodium polystyrene and prednisolone was started, with normal

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