Secondary leiomyosarcoma of the right ventricle

A surgical report

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SUMMARY A 48-year-old woman developed symptoms related to a tumour attached to the outflow tract of the right ventricle. Eleven years earlier she had undergone hysterectomy for leiomyosarcoma, and 4 years later a retroperitoneal mass was removed. One year later a further retroperitoneal mass was removed, and after a further 4 years a cutaneous tumour was removed from her back. Histologically these three tumours were identical with the leiomyosarcoma removed with the uterus at the primary operation.

A confident preoperative diagnosis of recurrent leiomyosarcoma within the pericardium was made, and it was possible to remove this tumour using cardiopulmonary bypass with relief of symptoms. The patient remains well 15 months after surgery.

Nicol and McAndrew (1968) reported a cardiac leiomyosarcoma causing death almost 10 years after removal of a uterine leiomyosarcoma. The diagnosis was made after death and they and others were unable to determine whether the myocardial neoplasm was a further primary or a solitary secondary deposit.

Rosenblatt and Featherston (1960) reported a patient with metastatic leiomyosarcoma involving the heart, and arising from a primary uterine tumour removed 3 years before the onset of cardiac manifestations.

We report a third case in which the diagnosis of secondary cardiac leiomyosarcoma was made and successful excision was carried out.

Case report

A 48-year-old woman was admitted in July 1976 to the Royal United Hospital, Bath, complaining of pain in her left shoulder for 5 days, radiating downwards into the left chest wall. It was pleuritic in nature and was relieved by lying flat. For some weeks she had noted a continuous dull aching pain in the left shoulder, progressive dyspnoea on exertion, and tiredness. There were no other significant symptoms.

In 1965, at the age of 37, she had undergone subtotal hysterectomy for a tumour, which at operation and examination was found to be a leiomyosarcoma. Postoperatively she remained well

until 1969, when she developed a mass in the abdomen, which at laparotomy was found to be a well-encapsulated retroperitoneal mass lying medial to the left kidney and ureter. It was removed completely and histology was similar to the previous specimen. A further abdominal mass appeared in 1970, lying in a retroperitoneal position on the left pelvic brim, and was removed completely. Histological examination again revealed metastatic leiomyosarcoma, and at this time she was treated with Provera (medroxyprogesterone acetate) 100 mg t.d.s., which she continued to take until her admission. A further cutaneous recurrence was removed from her back in 1973.

On admission she was a well-nourished woman with no pallor, cyanosis, or lymphadenopathy. There were no abnormal signs on general examination. Heart rate sinus rhythm 80 a minute, low volume, not paradoxical, blood pressure 130/90 mm Hg. Jugular venous pressure not raised initially, but venous paradox was noted subsequently. The apex beat was not palpable and auscultation revealed faint normal first and second heart sounds with no added sounds, murmurs, or rubs. Percussion of the cardiac outline suggested cardiac enlargement. No oedema was present, and the lungs were clear. Well healed abdominal scars were present from her previous operations; there was no hepatic enlargement or tenderness and no other abdominal abnormalities. The central nervous system was normal.

INVESTIGATIONS

Haemoglobin 14.7 g/dl, white cell count 6700 mm³, plasma viscosity 1.77. Blood urea, electrolytes, glucose, proteins, bilirubin, alkaline phosphatase, aspartate transaminase—all within normal limits. Electrocardiogram showed sinus rhythm, very low voltages but otherwise normal complexes, compatible with pericardial effusion. Chest x-ray film showed gross enlargement of the cardiac shadow which on screening indicated virtually no movement of its borders. There was no evidence of pulmonary venous hypertension and the appearances were compatible with those of a large pericardial effusion. Ultrasonic scan disclosed a very large pericardial effusion but no localised nodule or tumour mass within the pericardial sac.

PROGRESS

The effusion was aspirated and 670 ml straw coloured fluid removed without significant change in the radiological appearances. Bacteriological studies of the fluid were negative and cytological examination disclosed no neoplastic cells. It was considered that the patient had a further secondary deposit within the pericardium and exploratory surgery was advised as repeated attempts at aspiration were unsuccessful. She was transferred to Bristol Royal Infirmary, Cardiac Surgery Unit.

OPERATIVE FINDINGS

Median sternotomy confirmed the presence of a large pericardial effusion and on opening the pericardium nearly 2 litres of straw coloured fluid were aspirated. The heart was tiny and tucked behind the

tumour mass which was the size of a grapefruit, loculated, cystic, and solid in parts, and arising from the surface of the outflow tract of the right ventricle 1 cm below the pulmonary valve and between the right coronary artery and the left anterior descending coronary artery. The whole tumour was mobile except where it was attached by a pedicle 2 cm in diameter to the myocardium. There was no evidence of other metastases within the pericardial sac.

Using full cardiopulmonary bypass the right ventricle was opened just proximal to the tumour which was removed together with a large disc of right ventricular wall, incising the muscle as closely as possible to the coronary arteries. It was apparent that a thin rim of tumour remained in the myocardium but this was extensively diathermied and it was hoped that no residual tumour was left viable. The defect in the right ventricle which was about 5 cm in diameter was repaired with a large Dacron patch. The pulmonary valve was undamaged. Bypass was readily discontinued, the pericardium was closed, and she made an uneventful post-operative recovery.

HISTOLOGY

Macroscopically the specimen was a lobulated tumour (18 × 12 × 9 cm) weighing 225 g, with a smooth surface. At the margin of resection there was some attached muscle tissue. On section it had a grey/brown colour with areas of haemorrhage and necrosis and an ill-defined trabecular pattern. Sections showed a fairly well differentiated smooth muscle tumour in which there were only occasional

Fig. 1 Operative view which shows the heart to be completely obscured from the tumour arising from the outflow tract of the right ventricle.

Fig. 2 Excised specimen.

mitotic figures and foci of cellular atypicity. The histological appearances were consistent with metastasis from a uterine leiomyosarcoma.

FOLLOW-UP

When seen one year after operation she was asymptomatic. No abnormal signs were present, and the chest radiograph showed no evidence of cardiac enlargement.

Discussion

In view of the preceding history in this patient, there is little doubt that the myocardial tumour was a secondary deposit from the primary uterine lesion removed 11 years previously, since 3 similar lesions had been removed in the intervening period.

This is the third report of uterine leiomyosarcoma metastasising to the heart (Rosenblatt and Feather-

ston, 1960; Nicol and McAndrew, 1968) but the first in which successful surgical excision was carried out and the diagnosis established before operation. The need to excise an isolated myocardial secondary deposit will not occur frequently but should not be overlooked. Clearly this patient's history and survival are related to the very low malignancy of her tumour.

References

- Nicol, A. G., and McAndrew, G. M. (1968). Cardiac leiomyosarcoma: primary or secondary? *British Heart Journal*, **30**, 432-435.
- Rosenblatt, W. H., and Featherston, W. P. (1960). Metastatic leiomyosarcoma of the heart. *American Journal of Cardiology*, **6**, 672-675.

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