Malignant neurilemmoma of left atrium

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SUMMARY A 31-year-old woman, whose chief complaint was back pain, was found to have an echocardiographic abnormality suggestive of a left atrial myxoma. Angiography clearly showed a large radiolucent mass protruding from the left atrium into the left ventricle during diastole. X-rays of the sacrum disclosed the presence of a malignant tumour. Biopsy specimens taken from the tumour in the sacral plexus showed a malignant neurilemmoma. Despite extensive chemotherapy, widespread metastases developed and the patient died four months after admission. Necropsy disclosed three discrete tumours in the left atrium, which were found to be malignant neurilemmoma, shown on histology to be metastases from the primary in the sacral plexus.

Cardiac tumours, primary or secondary, are rare and their diagnosis during life is unfortunately difficult. Recently, however, echocardiography, especially M-mode and cross-sectional in combination, has proved valuable in this respect. The case presented in this paper is of special interest in that a neurilemmoma originating in the sacral region was complicated by three large secondary deposits in the left atrium, and precise diagnosis of the cardiac tumours was successfully made by echocardiographic techniques.

Case report

A 31-year-old woman remained in good health until the beginning of February 1979, when she noticed slight back pain and numbness in the left leg. These symptoms progressed rapidly and in April 1979, she was admitted with a suspected lumbar disc herniation. There was no history of syncope or of a heart murmur having been discovered. No embolic phenomenon had occurred.

On examination, the blood pressure was 96/48 mmHg and the pulse rate was 98 per minute and regular. The first heart sound was loud, there was a grade 2/6 presystolic rumble best heard at the apex, and an early diastolic sound at the left sternal border

in the fourth intercostal space, which closely resembled a mitral opening snap.

The lungs were clear. The haemoglobin was 10.4 g/100 ml, and the white blood cell count was 9400/mm.³ The erythrocyte sedimentation rate was 105 mm/h. Serum potassium was 5.1 mmol/l, alkaline phosphatase 4.5 international units, and lactic dehydrogenase 628 international units/l. The electrocardiogram showed sinus rhythm, right axis deviation, a deep S wave in the left praecardial leads suggesting right ventricular hypertrophy, and right atrial enlargement. The chest x-ray film disclosed a normal configuration of the cardiac shadow and faint oval shadows in the left upper and right middle lung fields. An M-mode echocardiogram showed a mass of echoes appearing behind the anterior mitral leaflet during diastole, highly suggestive, but not absolutely diagnostic, of a left atrial tumour. On cross-sectional echocardiography, however, a left atrial tumour could be more clearly seen (Fig. 1). Angiography disclosed a large radiolucent mass which protruded from the left atrium into the left ventricle during ventricular diastole (Fig. 2). A left atrial myxoma was diagnosed, the early diastolic sound being considered to be a tumour plop, but x-rays of the sacrum disclosed a destructive lesion at the left side of the sacrum.

End-diastolic phase

End-systolic phase

Fig. 1 M-mode (upper panel) and cross-sectional (lower panels) echocardiograms (four-chamber views). M-mode echocardiogram shows a mass of echoes appearing behind the anterior mitral leaflet during diastole. Cross-section echocardiogram shows a left atrial mass. IVS, interventricular septum; RV, right ventricle; RA, right atrium; LV, left ventricle; LA, left atrium; TV, tricuspid valve; MV, mitral valve.

Fig. 2 Left ventricular angiography showing a radiolucent mass protruding from the left atrium into the left ventricle during diastole. This finding is suggestive of the presence of a mobile left atrial mass.

indicating the presence of a malignant tumour, and biopsies from a tumour in the sacral plexus showed it to be a malignant neurilemmoma. This was not considered resectable. The patient was subsequently treated by chemotherapy, but there was no response. Widespread metastases developed and she died four months after admission.

POSTMORTEM FINDINGS

The pericardium was normal and there were no pericardial adhesions or significant fluid. The heart was slightly enlarged, weighing 300 g. The thickness of the right ventricular free wall was 3 mm. Upon opening the left atrium, three myxoid fibrous tumour masses were found (Fig. 3A). The largest one, measuring $45 \times 35 \times 30$ mm, ovoid in shape, protruded from the atrial septum on a short pedicle. The other two tumour masses, both of which were located in the anterior commissural region of the mitral valve, measured $30 \times 25 \times 20$ mm and $30 \times 20 \times 20$ mm, respectively. They were elastic, firm, fibrous but partly myxoid with whorled patterns. In

the retroperitoneal space, there was a huge tumour mass, $13 \times 10 \times 10$ cm, covered by a thin fibrous capsule. On cut section, the tumour was firm and fibrous, with haemorrhagic and necrotic areas. There were multiple metastases elsewhere, many scattered in the lungs.

HISTOLOGICAL FINDINGS

The histological findings of the tumours in the left atrium were identical to those of the primary in the retroperitoneal space. They were highly cellular and composed of plump spindle cells with ovoid or spindle-shaped nuclei. The spindle cells were arranged in wavy fascicles, simulating the Schwann cell cords. The nuclei were arranged in a parallel fashion, forming a striking palisade pattern (Fig. 3a and c) with some aberrant cells showing bizarre mitotic activity. These findings suggested a malignant neurilemmoma (Schwannoma) primarily arising from the left sciatic nerve.

Discussion

Malignant neurilemmoma is a malignant neoplasm, arising in the perineurium of large nerves, characterised by the morphological patterns of Schwann cell differentiation. Neurilemmoma of the heart has been reported before,¹⁻⁵ but its occurrence in the left atrium is extremely rare. In this case the primary retroperitoneal tumour arose in the sacral plexus and metastasised to the left atrium after passing through the pulmonary circulation, both the cardiac and the sacral tumours showing the typical naked eye and cytological pattern of a neurilemmoma. The retroperitoneal tumour was far larger than the left atrial tumours, and the latter arose from the endocardium without invasion of the underlying myocardium. This indicates that the retroperitoneal tumour was the primary and the cardiac lesions were metastatic deposits resulting from endocardial implantation of tumour emboli. This mechanism was considered by Crofts and Forbes² to be the explanation for a left atrial secondary deposit in their case of malignant neurilemmoma of the lung. The alternative, in our case, that the tumours were all separate primaries which grew simultaneously and independently is unlikely, for then the cardiac tumours would have originated from the cardiac plexus and would have occurred in the myocardium or on the external aspect of the base of the heart, not within the atrium.

The combined use of M-mode and cross-sectional echocardiography clearly identified the presence of left atrial tumours in our patient but was not able to distinguish the pathological causes. The case appears to be unique.

Fig. 3 (A) On opening the heart, three tumour masses are found occupying the left atrium, one of which protrudes from the atrial septum on a short pedicle; the other two bulge from the anterior commissure of the mitral valve like a dumb-bell. (B) The tumour in the left atrium. (H and E. stain. Original magnification $\times 100$.) (C) The tumour in the retroperitoneal space. (H and E stain. Original magnification $\times 200$.) The tumour cells are spindle or ovoid, with wavy fibre formation. Their nuclei are hyperchromatic, varying in size and shape, with giant cell formation in places.

References

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