Primary Sarcoma of Left Atrium

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Tumours of the left atrium are usually first discovered at necropsy. When the diagnosis has been made during life the finding has often been a fortuitous one during an operation for mitral valve disease. Campeau and David (1960) reported 45 left atrial myxomas, of which 4 were suspected clinically, 3 were confirmed by angiography, 16 were discovered during operation for mitral stenosis, and 22 were found at necropsy. Rheumatic heart disease, subacute bacterial endocarditis, and disseminated lupus erythematosus are the diagnoses most commonly made in these patients before death. Four cases described by Goodwin *et al.* (1962) had all been suspected on clinical grounds before surgery. Three of the four had first been thought to have an inflammatory endocarditis.

We report a case of primary sarcoma of the left atrium because it presented features which allowed a diagnosis of a left atrial tumour to be made on clinical grounds alone. The malignant nature of the lesion was not appreciated before operation, but in retrospect the relentless rather than fluctuating changes in the physical signs relating to the mitral valve with the comparatively slight constitutional upset might have made us suspect, before the operation, that this was a sarcoma and not a myxoma.

CASE REPORT

In December 1964 a housewife aged 47 presented with the sudden onset of aphasia and right facial weakness. No murmurs were audible at this time. She was admitted to hospital and a carotid angiogram confirmed a left middle cerebral artery occlusion. She made a good recovery and was discharged. Further episodes of right-sided weakness with aphasia occurred. There was no evidence of embolism affecting other parts of the brain or the body, and the attacks usually occurred when postural hypotension, jeopardizing a collateral circulation, could have been incriminated. A pansystolic

murmur was heard in May 1965, but no diastolic murmur was audible at that time.

Acute pulmonary oedema recurred in 1965. A definite diastolic murmur was heard but was not always present. A striking feature was the normal heart size on the chest x-ray films despite the severe attacks of pulmonary oedema. She was readmitted.

Investigations.—Haemoglobin 76% ; W.B.C. 12,000 ; E.S.R. 19 mm. in one hour. No L.E. cells. Serum proteins and electrophoretic strip normal. Chest x-ray film was within normal limits. Electrocardiogram showed sinus rhythm and P mitrale. Phonocardiogram: mitral diastolic and systolic murmurs and third heart sound recorded at the apex.

A diagnosis of left atrial myxoma was made and cardiac catheterization was performed. This showed a large filling defect above the mitral valve and extending through it into the left ventricle. It was assumed that these findings confirmed the clinical diagnosis of left atrial myxoma.

Surgical removal of the tumour was undertaken. An obviously malignant tumour fungating through the left atrial wall was present. On opening the left atrium an extensive polypoidal and necrotic tumour appeared to arise from just above the interatrial septum. The mitral valve was extensively infiltrated throughout its circumference, and a small area of tumour extended into the posterior cusp of the mitral valve. The tumour filled the left atrium from just below the inferior pulmonary veins and almost completely obstructed the mitral valve. As much of the tumour as possible was removed. A paraffin section showed a "spindle cell" sarcoma.

The patient recovered slowly from the operation, but she then deteriorated, dying six weeks later.

Post-mortem Findings.—The visceral and parietal pericardium were lightly adherent. Enlargement of the heart was present without hypertrophy. A grey-white soft granular tumour mass (see Fig.) filled the mitral orifice, extending in the way described at operation. Metastatic deposits were present in the left middle cerebral artery, omentum, and adrenal glands.

COMMENT

The three effects of left atrial tumours are obstructive, embolic, and constitutional. Goodwin rightly stressed the constitutional effects of benign myxomas. Sarcoma of the left atrium behaves differently. The embolic effects were predominant in our patient and occurred before there was any evidence of heart disease or constitutional disturbance. When obstruction of the mitral valve developed, changing heart murmurs were present. Of equal importance, however, were the attacks of pulmonary oedema, with a normal-sized heart on the chest x-ray film. This case simulated mitral incompetence initially but later went on to develop a mitral diastolic murmur as obstruction developed.

Examination of the tumour at operation provided the probable explanation for this sequence of events. The tumour first infiltrated the posterior cusp of the mitral valve, interfering with its closure and causing incompetence. Later the growing tumour obstructed the mitral orifice, causing the diastolic murmur.

Malignant cardiac tumours usually present with pericardial effusion. Diagnosis is often made by examination of the pericardial aspirate (Johnson and Stokes, 1964). This case was also unusual in having no clinical evidence of pericardial involvement.

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Post-mortem specimen showing the relation of the tumour to the left atrial cavity and the mitral valve.