An unusual case of recurrent left atrial myxoma

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Summary: In a patient who had a calcified left atrial myxoma resected, recurrence developed 31 months later. Although complete radical resection of the recurrent tumour presented a special problem, the patient survived the second operation. The tumour recurred again and the patient had two episodes of cerebral embolism 1½ and 2 years later, respectively, and died 3½ years after the second operation. The erythrocyte sedimentation rate correlated with the size of the tumour, and the recurrent tumour seemed to grow more rapidly than the primary tumour. Experience with this case and a review of the nine reported cases of recurrent left atrial myxoma suggest that a radical approach is necessary at the primary operation.

Résumé: Après la résection d'un myxome calcifié de l'oreillette gauche chez un malade, un récidive surgit 31 mois plus tard. Quoique la résection radicale et complète de la tumeur récidivante présenta un problème particulier, le patient survit la deuxième opération. Le myxome récidiva encore une fois et fut signalé par deux instances d'embolie cérébrale 1½ et 2 ans plus tard, respectivement. Le patient est mort 3½ ans après la deuxième intervention. La vitesse sédimentaire des érythrocytes montra un rapport avec la grosseur de la tumeur, et la récidive sembla croître plus rapidement que la tumeur primitive. L'expérience de ce cas et une revue des neuf cas décrits de récidive de myxome de l'oreillette gauche indiquent qu'une approche radicale est nécessaire lors de la première intervention.

Of the nine patients with symptomatic recurrent left atrial myxoma reported in the English and French literature[1-10] seven were successfully treated by radical resection under cardiopulmonary bypass and two died postoperatively[3,10] In five patients a patch was used to repair the atrial septal defect created; the two deaths were in this group.

We report the 10th case of recurrent left atrial myxoma. Complete radical resection presented a special problem. The tumour recurred and the patient died 3½ years after the second operation.

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Case report

A 46-year-old white man was seen in January 1968 because investigation for syncope revealed evidence of a left atrial tumour. Although he had had mild dyspnea on effort since childhood, there was no history of rheumatic fever or of a heart murmur. Severe pain developed in 1960 in the left leg; it recurred a year later, and because there was evidence of vascular insufficiency, a lumbar sympathectomy was carried out, with some relief of his calf claudication. A syncopal episode occurred in December 1967, followed by transient loss of vision, dysphasia and weakness of the left hand, and was investigated at another institution.

He had coarse features and large, spade-like hands. On his face and neck there were several sebaceous cysts and pedunculated skin lesions, which were familial. Arterial pulses were present in both arms and legs but those distal to the femoral pulses were reduced in volume. His blood pressure was 125/80 mm Hg and both the jugular venous pressure and pulse were normal. There was no right ventricular overactivity. Auscultation at the apex revealed a loud, split first heart sound, a grade 2/6 blowing pansystolic murmur, which became louder on expiration, and a third heart sound. There was neither a diastolic murmur nor an opening snap. At the base the second sound was split, the pulmonary component being normal. There were no aortic or pulmonic murmurs. The chest and abdomen were normal.

The electrocardiogram (ECG) revealed evidence of sinus rhythm and left atrial hypertrophy, with notched P waves. Chest radiographs with barium in the esophagus showed prominence of the pulmonary vessels, with a suggestion of right heart and left atrial enlargement. Cardiac fluoroscopy revealed a calcified density moving backwards and forwards in the region of the mitral valve.

The hemoglobin value was 13.3 g/dl and the erythrocyte sedimentation rate (ESR) was 46 mm/h. Plasma protein electrophoresis revealed the total protein value to be 8.4 g/dl, with 41% albumin and 59% globulin. All other laboratory findings were normal and the patient remained afebrile.

At cardiac catheterization done before referral the pulmonary capillary wedge pressure was mildly elevated (14 mm Hg) and a left atrial cineangiogram revealed a large mass travelling freely in and out of the left atrium and left ventricle.

Operation was performed under cardiopulmonary bypass in January 1968. The left atrium contained a large, multilobular, myxomatous tumour attached to two stalks arising from the interatrial septum near the fossa ovalis. The mitral valve was normal. The soft and friable tumour (weight, 80 g) and stalks were excised. Postoperatively tracheostomy and respiratory support were required for several days but the later hospital course was uneventful. The grade 2/6 systolic murmur at the apex persisted.

In March 1969, 14 months after surgery, poor progression of the R waves in leads V_1 to V_3 and T-wave flattening in leads V_5 to V_6 were noted on his ECG but an exercise tolerance test was negative. The ESR was 3 mm/h and the chest radiographs were normal. Cardiac catheterization revealed a moderate increase in mean pulmonary capillary wedge pressure to 18 mm Hg. In the levophase of the pulmonary angiogram the left atrium was outlined; a filling defect was not demonstrated.

In July 1970 he was readmitted because of left-sided chest pain of recent onset, and dizziness, general malaise and fatigue of 2 months' duration. The ESR was 54 mm/h and the hemoglobin value 11.1 g/dl. The ECG revealed definite evidence of inferior wall damage and posterolateral ischemia. Cardiac fluoroscopy disclosed enlargement of the left atrium; cardiac catheterization demonstrated an elevated mean wedge pressure at rest (9 mm Hg), which increased further with exercise (23 mm Hg). Pulmonary cineangiography visualized the left atrium, with a large polypoid tumour obstructing the mitral valve in diastole. Radiographs of the skull, made because of the acromegalic features, showed a normal sella turcica; growth hormone activity was found to be normal.

Subsequently a second operation was carried out under cardiopulmonary bypass. The recurrent polypoid mass (dimensions, 6.5 x 3 x 2 cm; weight, 30 g) was excised together with the broad base and other smaller tumours resembling local secondary deposits on the posterior wall of the left atrium (Fig. 1). The interatrial septum was not excised because of the numerous "deposits" on the remaining left atrial wall. The cut surface of the mass resembled old blood clot, with grey

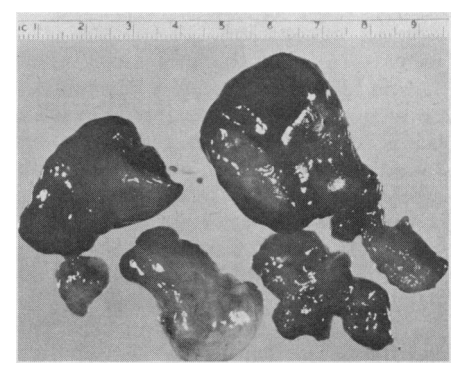

FIG. 1—Recurrent left atrial myxoma removed in July 1970.

gelatinous material extending proximally towards the stalk; microscopy revealed laminated thrombus with myxomatous areas. Atrial flutter developed postoperatively and the apical systolic murmur persisted. Radiotherapy was considered but was not used.

In September 1970 he was again seen because of recurrence of dizziness, but there were no new findings and the ESR was only 9 mm/h. In January 1972 the ESR was 23 mm/h when he was admitted because of the sudden development of a right hemiplegia. He underwent physical rehabilitation and speech therapy and made a partial recovery. By July 1972 his heart size had increased, the ESR was 40 mm/h and paroxysmal atrial flutter was documented. In September 1972 he was admitted to hospital with grand mal seizures and aspiration pneumonia. His cardiac rhythm was unstable, with episodes of atrial fibrillation, atrial flutter and sinus rhythm. The apical systolic murmur was still present. Findings on brain scan were consistent with occlusion of the right middle cerebral artery. He underwent partial rehabilitation but died suddenly in January 1974.

At autopsy multiple myxomas were found in the left atrium, with six sites of origin and extension to the right atrium through the interatrial septum. Multiple old infarcts were present in the brain, kidneys and spleen, and an old scar was present on the inferior surface of the left ventricular myocardium. Other findings included pulmonary edema, dilatation of all cardiac chambers, atherosclerosis of the aorta and left external iliac artery, liver congestion with hemorrhagic necrosis, left upper lobe pneumonia and extramedullary hematopoiesis in the spleen.

Discussion

The protean manifestations[11,12] of a left atrial myxoma are well illustrated in the case described in this report. Recurrent major systemic embolization dominated the clinical picture. The sudden appearance of symptoms of peripheral vascular insufficiency could well have resulted from a saddle embolus to the bifurcation of the iliac artery. The sudden appearance of evidence of inferior wall infarction and posterolateral ischemia on the ECG in 1970 was presumably the result of coronary embolism.

The ESR correlated with tumour size. In our patient, 14 months after primary resection the ESR was 3 mm/h and no left atrial tumour was visualized on angiography; 16 months later, when the ESR was 54 mm/h and the patient frankly symptomatic, angiography demonstrated a large left atrial tumour. Again, 2 months after the second operation the ESR was only 9 mm/h, whereas 16 months later, when the patient suffered a major cerebral embolic episode, the ESR was 23 mm/h. This illustrates the usefulness of the ESR in follow-up and the fact that a small tumour can manifest itself by systemic emboli before mechanical factors interfere with cardiac function.

The rate of growth of the recurrent tumour was more rapid than that of the primary tumour. Possible embolic evidence of the primary tumour dated back to 1960; 8 years later, at the first operation, the tumour removed weighed 80 g. However, only 31 months later a recurrent tumour, which had not been demonstrated on angiography 16 months before, was removed; it weighed 30 grams. This observation supports the contention of Walton, Kahn and Willis[3] that, although the primary tumour grows slowly, the recurrent lesions tend to grow rapidly. The average interval in the nine cases of recurrent left atrial myxoma between the primary operation and the second operation was about 30 months.[1-10]

On the basis of their experience with three patients without symptomatic recurrence after operation, followed for 5 to 10 years, Firor, Aldridge and Bigelow[13] concluded that simple excision was adequate and that resection of the adjacent interatrial septum or wall was unnecessary. However, in 1967 Gerbode, Kerth and Hill[1] reported recurrence in a patient 4 years after simple excision with cauterization of the base, and advocated excision of the septum in each case and replacement with a Dacron patch. Although Castaneda and Varco[14] cast doubt on the need for this radical approach, they favoured excision of the tumour and the broad pedicles on the interatrial septum and closure of the resulting defect by direct suture. They described a patient in whom this approach was adopted,[14] but histologic study did not reveal evidence of septal invasion. Recently Walton and colleagues[3] described a similar case in which the tumour and its broad stalk were excised with a cuff of the atrial septum and the defect repaired, but despite no histologic evidence for extension beyond the subendocardial layer into deeper atrial muscle, recurrence from the septum occurred only 6 months later and the patient did not survive a second operation.

In five of the nine cases of recurrent left atrial myxoma in the literature, radical resection of part of the interatrial septum and repair of the defect with a Dacron patch was carried out.[1-3,5,10] Recently Hardin and associates[9] reported their experience with primary tumours of the heart between 1951 and 1972; 8 of the 12 patients with left atrial myxomas operated on with cardiopulmonary bypass underwent radical resection of the tumour and the septal attachment; "in some instances a patch was used to repair the defect in the septum". In view of the finding of Newman, Condell and Pritchard[15] of a 26% overall mortality in primary resection of atrial myxomas, Maranhao and colleagues[6] have suggested a more radical approach to avoid reoperation. Although iatrogenic heart block can be a problem,[14] heart block after reoperation was successfully treated with a temporary transvenous pacemaker in the patient of Maranhao and colleagues.[5,6]

Multiple tumours were present in our patient, although growth from "pre-tumor cells"[3] in the interatrial septum cannot be excluded as the cause of recurrence. Two stalks were seen to arise from the septum at the primary operation, and small tumours were found on the posterior left atrial wall at the second operation. At autopsy, six sites of origin were found in the left atrium alone, besides invasion of the right atrium via the septum, but no histologic evidence of frank malignant tumour was seen. A truly radical resection at the second operation in our patient would have meant a formidable procedure, with the removal of the entire interatrial septum and posterior left atrial wall. However, a more radical resection at the primary operation might have prevented recurrence, as illustrated in the series of Hardin *et al*,[9] in which one patient survived 9 years after primary resection and a second patient survived for more than 2 years after reoperation for recurrence.

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