ANGIOMYXOMA OF THE LEFT AURICLE

Report of a Case

By R. H. B. PROTHEROE, M.A., M.D.

Senior Registrar and Research Fellow in Pathology, Westminster Hospital

Primary tumours of the heart are uncommon. Various authors estimated their frequency at autopsy as follows: Rand and Sachs (1943), 0.5 per cent.; Straus and Merliss (1945), 0.0017 per cent.; Lymburner (1934), 0.61 per cent. About three-quarters are benign and about half of the benign tumours are the so-called myxomas. Approximately 70 per cent. of these myxomas arise in the left auricle in the neighbourhood of the fossa ovalis, the majority of the remainder arising in the same situation in the right auricle. Occasional examples have been seen on the cusps of valves.

The term myxoma which only indicates a peculiar feature of collagenous stroma probably in part degenerative (Raeburn, 1952) has little merit and has led to needless confusion. Better terms are hamartoblastoma or angiomyxoma as they arise from elements of pre-existing intramural hamartomas. They may present at any age but the majority do so in the third to sixth decade.

The establishment of an accurate diagnosis of primary neoplasms as forecast by Mahaim (1945) has been assisted by the development of angiocardiology, and the intracavitary position of the primary angiomyxomas means that this technique is especially useful in establishing a clinical diagnosis. Both Brock (1956) and Scannell *et al.* (1956) have stressed the desirability of making a clinical diagnosis as the preparation of the patient and the surgical approach to these tumours when in the left auricle is different from that for mitral valvulotomy.

Pritchard (1951) reviewed 127 primary myxomas of the heart. Ante-mortem diagnosis was not made in one case. Since then over 30 cases have been reported and in 14 cases an ante-mortem diagnosis was made. Successful removal has been performed in three.

The case reported here illustrates the rapidity with which the clinical manifestations of a tumour may develop.

Clinical Details

54.2100. H.G.S. Aged 59 years. First attended the Gordon Hospital Outpatients in December, 1954, complaining of constipation together with intermittent rectal bleeding. At this time he complained of no symptoms referable to the cardio-vascular and respiratory systems. B.P. 150/95. Physical examination of the lower distal colon and rectum revealed no abnormality and a barium enema examination was within normal limits.

The patient was admitted as an emergency on April 16, 1955. On admission he was very dyspnoeic, orthopnoeic, restless and unco-operative. A history from relatives revealed that he had suffered from progressive shortness of breath, asthma and restlessness of sudden onset 48 hours before admission.

On examination he looked very ill and was dyspnoeic and orthopnoeic. There was cyanosis of finger tips and ears. The accessory respiratory muscles were prominent but the respiratory excursion was poor with expiratory wheezing. The apex beat was 160/min. with a pulse rate of 84/min. Owing to the tachycardia the heart sounds could not be heard well. There was no thrill or murmur. A clinical diagnosis of auricular fibrillation was confirmed by an electrocardiograph. The jugular venous pressure was raised, the liver was enlarged and tender and there was slight pitting oedema of the ankles.

A clinical diagnosis of congestive heart failure with auricular fibrillation was made and the patient was digitalised, given diuretics and sedated. However, the condition of the patient gradually deteriorated and he died on April 19, 1955, three days after admission. Owing to the severity of the patient's condition further investigation of the cardio-vascular system could not be carried out.

Necropsy Report

The relevant findings were as follows:

The body was of an elderly man of good

FIG. 1.—View of dilated left auricle with angiomyxoma arising near the fossa ovalis. The mitral valve appears normal.

nourishment. There was pitting oedema of ankles and sacrum. The facies were congested and cyanosed. The heart weighed 380 g. and there was approximately 200 ml. of straw-coloured fluid in the pericardial sac. The left atrium was dilated to nearly twice its normal size and its wall somewhat thickened. There was a sessile tumour 6.5 cm. x 3.5 cm. greatest diameter attached to the inter atrial septum in the neighbourhood of the fossa ovalis (see Fig. 1). The tumour was bluish-yellow in colour with red areas and evidence of recent haemorrhage. The surface was smooth and glistening and there was no evidence of a fibrinous exudate. The tumour was projecting down into the mitral valve, the leaflets of which showed no abnormality. There was no ante-

mortem thrombus in the left auricular appendage and the pulmonary veins were *not* obstructed.

The myocardium of the left ventricle showed some hypertrophy. The coronary arteries showed patchy atheroma with no evidence of embolism or thrombosis.

The right ventricle was dilated, the tricuspid valve admitted four fingers and the right auricle was also dilated and filled with post-mortem thrombus.

Lungs. Both pleural sacs contained large effusions of approximately 1,500 ml. of straw-coloured fluid. The lungs showed gross oedema and congestion of their bases and microscopy revealed oedema of the alveoli, congestion of the alveolar capillaries with numerous macrophages

FIG. 2.—Shows base of angiomyxoma richly supplied with thin-walled blood vessels. Note continuous layer of elastic tissue across its base. Verhoeff's elastic stain x 90.

FIG. 3.—'Lepidic cells' in stroma of angiomyxoma. Note vacuolation of the matrix. Haematoxylin and eosin x 380.

of the heart failure type filled with iron in the alveolar spaces. There was no abnormality of the pulmonary arterial tree.

Microscopically the tumour in the left auricle was covered by endothelium and there was no fibrinous exudate on its surface. The base of the

tumour was continuous with the subendothelial tissue of the auricle. The superficial subendothelial elastic fibres of the auricle were reflected onto the surface of the myxoma but there was a layer of deeper elastic tissue continuous across its base. Small arterioles and veins were numerous

FIG. 4.—Focus of calcification in angiomyxoma. Haematoxylin and eosin x 100.

FIG. 5.—Recent haemorrhage into matrix of the angiomyxoma. Haematoxylin and eosin x 150.

at its base (see Fig. 2), whilst in its substance large capillaries and vascular spaces were plentiful. The bulk of the tumour was composed of a matrix of faintly blue, oedematous mucinous material in which collagen and elastic fibres ramified. Within this matrix were found the characteristic so-called lepidic or vasoformative cells. These cells were usually elongated but stellate oval or rounded

forms were also seen and frequently the matrix was vacuolated in the areas where these cells were plentiful (see Fig. 3). The nuclei of the cells varied with their shape but had prominent nucleoli, nuclear membranes and chromatin pattern. These lepidic cells were frequently linear in pattern and related to vascular channels which were clearly lined by similar cells.

The tumour matrix stained pinkish-purple with PAS reagent, reddish-purple with mucicarmine. A stain for free iron revealed deposits, the result of old haemorrhage. Some of the iron pigment was impregnated on collagen. There were also foci of calcification (see Fig. 4).

There were also large areas of recent haemorrhage into the matrix of the tumour (see Fig. 5) and these were clearly the cause of the recent onset of symptoms. The liver weighed 1,600 g. and the parenchyma had a nut-meg appearance. Microscopy revealed passive venous congestion. The kidneys showed cortical vascular congestion and except for an aberrant renal artery supplying the lower pole of the left kidney, were otherwise normal. There was no evidence of infarction of the internal organs.

Discussion

The pathology of angiomyxoma of the heart has been well described by Pritchard (1951) and Raeburn (1952). Both these authors have discussed the confusion that has arisen with mural thrombi. Their comparative rarity and the consequent difficulty of workers to amass a series has contributed towards this confusion. Also angiomyxomas may be covered with fibrin and superficially resemble the mass thrombus associated with mitral valvular disease described by Evans and Benson (1948).

However, the angiomyxomas are usually larger than thrombi, are less cellular and do not retract. Although occasional cases of cardiac myxoma are associated with mitral valvular disease probably due to trauma of the tumour (Dexter and Work, 1941), the vast majority occur in hearts free from disease. However, in over half the cases the signs and symptoms of mitral valvular disease may be mimicked by the angiomyxomas of the left atrium and this has contributed to confusion with thrombi amongst clinicians.

Further arguments to support the theory that these tumours arise from embryonic mesenchyme are as follows. First, none of these tumours is found in the auricular appendages where thrombi are most common. Second, there is no evidence of stratification so common in thrombi and third, there appears to be a predilection of these tumours for the fossa ovalis of the atria.

Pritchard (1951) sectioned the fossa ovalis routinely in normal hearts and although myxoid tissue was not found, he did demonstrate minor endocardial abnormalities in the form of collections of capillary lucunae lined by swollen endothelial cells. Raeburn (1952) has stressed the angiomatous nature of these tumours. Indeed the term angiomyxoma is preferable to myxoma which only indicates a peculiar feature of the

collagenous stroma. As the fossa ovalis is most rich in connective tissue it is perhaps not surprising that the myxomas arise most commonly in this region.

Clinically angiomyxomas of the left auricle may present in a variety of ways. The commonest is as a case of atypical mitral stenosis. A history of rheumatic fever will not be obtained. The mitral diastolic murmur may alter with posture due to the tumour intermittently obstructing the mitral valve. The obstruction, however, may be chronic and severe enough to cause pulmonary hypertension and right ventricular enlargement. The case reported by Block *et al.* (1952) illustrated this.

Episodes of Stokes Adams syncope or epileptiform attacks may be encountered due to the tumour infringing on the mitral valve. Cases illustrating these symptoms have been described by Kendall and Symonds (1952) and Mills and Philpot (1951).

Fragmentation of these tumours leads to release of embolic material into the peripheral circulation. This may involve the entire tumour as in the case described by Brewin (1951) or portions as in the cases of Gleason (1955) and Block *et al.* (1955). The villous type of tumour appears especially liable to fragment. Another source of arterial emboli is the thrombotic material on the surface of the tumour (Weinstein and Arata, 1949).

The clinical signs and symptoms of mitral valvular disease together with the manifestations of peripheral emboli will naturally suggest to the clinician the presence of subacute bacterial endocarditis. The case described by Gleason (1955) illustrates this phenomenon. Only one case has been reported of a positive blood stream infection complicating angiomyxoma. Dick and Mullin (1956) have recently described a case of myxoma complicated by blood stream infection by staph. aureus and candida parapsilosis.

Another group of cases present as a relentless, progressive heart failure unrelieved by digitalis or diuretics. The case presented is of this type. The sudden onset of symptoms two days before admission, the rapid deterioration of the patient's general condition so that when first seen in hospital his general condition precluded any hope of surgical intervention. The tumour had remained silent until haemorrhage had occurred into its parenchyma probably due to incidental trauma. The sudden increase in size had caused the rapid onset of congestive heart failure unrelieved by digitalis or diuretics. This was supported by the absence of changes in the pulmonary arterial tree.

Notwithstanding the difficulty of diagnosis, ante-mortem diagnosis has been made on 14 cases since 1951 and successful removal has been undertaken in three cases (Crafoord (1954), quoted

by Brock, one case; Scannell (1956), one case; and Bigelow *et al.* (1956), one case). Both Brock and Scannell *et al.* have stressed the necessity for diagnosis before laparotomy. The former considers that the best surgical approach is through the intra-atrial septum and right auricle, and the latter have stressed need for removal under direct vision with the circulation temporarily arrested.

Unsuccessful attempts at removal have been made in a further nine cases: Bahnson and Newman (1953), one case; Steinberg (1953) *et al.*, two cases; Clowes (1954) *et al.*, one case; Bailey (1955), a review of four cases; and Jones and Julian (1955), one case. In nearly all these cases the diagnosis was made at operation for mitral valvulotomy. In two, surgical removal was not attempted (Kirkeby and Leren (1952), one case; and Steinberg (1953), one case).

Clearly removal of these tumours is a hazardous procedure but as the prognosis in these cases is so poor, early diagnosis and surgical removal is the only hope of successful treatment.

Summary

A case of angiomyxoma of the left auricle is presented and discussed.

Death was due to congestive heart failure five days after onset of symptoms.

The results of surgical removal are discussed.

Successful removal would appear to depend on early diagnosis assisted by angiocardiography.

Acknowledgments

I wish to thank Mr. A. Lawrence Abel, M.S., F.R.C.S., for his permission to publish this case, also Dr. Peter Hansell for photography of the specimen.

BIBLIOGRAPHY

- BAHNSON, H. T., and NEWMAN, E. V. (1953), *Bull. Johns Hopk. Hosp.*, **93**, 150.
 BAILEY, C. P. (1955), 'Surgery of the Heart,' p. 901, Lee-Febringer, Philadelphia.
 BIGELOW, W. G., DOLAN, F. G., and CAMPBELL, F. W. (1956), *Bull. Soc. internat. de Chir.*, in press.
 BLOCK, W. J., PARKER, R. L., and EDWARDS, J. E. (1952), *Proc. Mayo Clin.*, **27**, 361.
 BREWIN, T. B. (1951), *Guy's Hosp. Rep.*, **100**, 278.
 BROCK, R. (1956), *ibid.*, **105**, 382.
 CLOWES, G. H. A., JR., NEVILLE, W. E., HOPKINS, A., ANGOLA, J., and SIMEONE, F. A. (1954), *Surgery*, **36**, 557.
 CRAWFORD, C. (1954), quoted by Brock, R. (1956).
 DEXTER, R., and WORK, J. L. (1941), *Arch. Path.* (Chicago), **32**, 995.
 DICK, H. J., and MULLIN, E. W. (1956), *N.Y. St. J. Med.*, **56**, 856.
 EVANS, W., and BENSON, R. (1948), *Brit. Heart J.*, **7**, 180.
 GLEASON, I. O. (1955), *Cancer*, **8**, 839.
 JONES, I. G., and JULIAN, D. G. (1955), *Brit. med. J.*, **2**, 361.
 KENDALL, D., and SYMONDS, B. (1952), *Brit. Heart J.*, **14**, 139.
 KIRKEBY, K., and LEREN, P. (1952), *Acta med. scand.*, **143**, 385.
 LYMBURNER, R. M. (1934), *Canad. med. Ass. J.*, **30**, 368.
 MAHAIM, I. (1945), 'Les Tumeurs et les Polypes du Coeur,' Masson et Cie, Paris.
 MILLS, P., and PHILPOT, M. (1951), *Brit. Heart J.*, **13**, 115.
 PRITCHARD, R. W. (1951), *Arch. Path.* (Chicago), **51**, 98.
 RAEBURN, C. (1952), *J. clin. Path.*, **5**, 339.
 RAND, J. M., and SACHS, J. (1943), *Amer. Heart J.*, **26**, 385.
 SCANNELL, J. G., BREWSTER, W. R., and BLAND, E. F. (1956), *New Engl. J. Med.*, **254**, 601.
 STEINBERG, I., DOTTER, C. T., and GLENN, F. (1953), *Dis. Chest*, **24**, 599.
 STRAUS, R., and MERLISS, R. (1945), *Arch. Path.* (Chicago), **39**, 74.
 WEINSTEIN, M. S., and ARATA, J. E. (1949), *Amer. Heart J.*, **38**, 781.

THYROID DISEASE

(*Postgraduate Medical Journal*, July 1957)

Price: 3s. 9d., post free

DIAGNOSTIC PROCEDURES IN THYROID DISEASE

Russell Fraser, M.D., F.R.C.P., D.P.M.

THE PLACE OF RADIOACTIVE IODINE IN THE TREATMENT OF THYROID DISEASE

E. E. Pochin, M.D., F.R.C.P.

ANTITHYROID DRUGS

James Crooks, M.B., M.R.C.P. (Lond. and Ed.), F.R.F.P.S.G.

SUBACUTE THYROIDITIS

Selwyn Taylor, M.Ch., F.R.C.S.

RECENT WORK ON THYROID HORMONES

J. H. Wilkinson, B.Sc., Ph.D., F.R.I.C.

SOME UNUSUAL MANIFESTATIONS OF THYROID DISEASE

W. R. Trotter, D.M., M.R.C.P.

CARCINOMA OF THE THYROID

John E. Piercy, F.R.C.S., F.R.C.S.E.

LYMPHOID GOITRES

T. Levitt, M.A., F.R.C.S.Eng., F.R.C.S.Ed., F.R.C.S.I.

Published by

THE FELLOWSHIP OF POSTGRADUATE MEDICINE

60, Portland Place, London, W.1