

A Case of Multiple Cardiac Myxomas

A. J. NAQVI
T. HABIBA
AZHAR FARUQI
A. H. KHAN
M.A. NAJEEB

SUMMARY

The case of a 28 years old male with vague symptoms and suspected mitral stenosis is presented. 2-D echocardiography demonstrated multiple solid masses in all four cardiac chambers. At operation five myxomatous tumors were excised from the left and right atria and the left and right ventricles. This is a rare case of multiple cardiac myxoma.

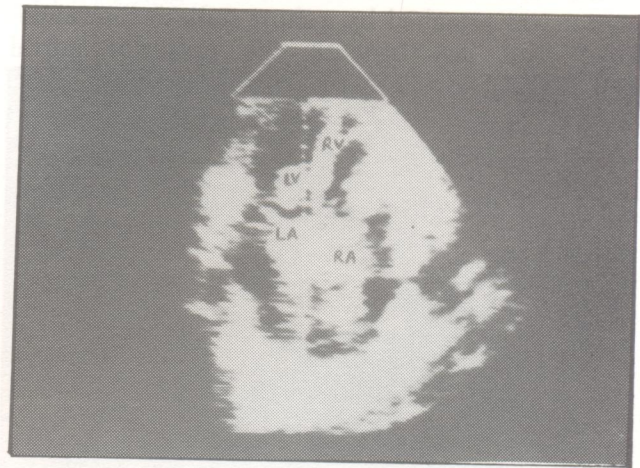
INTRODUCTION

Intra-cardiac myxomas are the most common primary tumours of the heart (8, 10, 15). Most are located in left or right atrium. Although tumours arising from multiple sites in the heart have been reported their location in all four cardiac chambers is extremely rare (1, 2). The purpose of this presentation is to report a case where five separate tumors were excised from both the atria and both the ventricles.

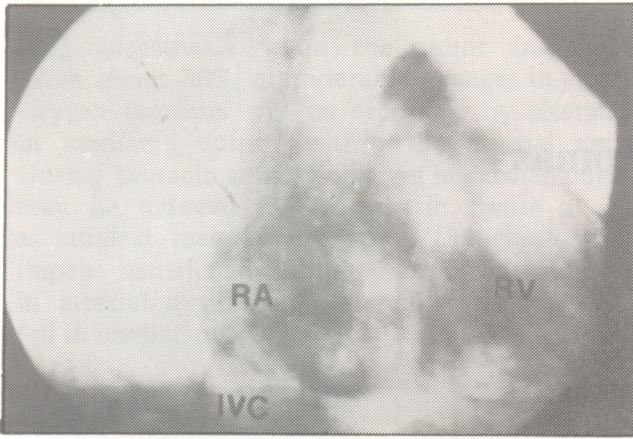
CASE REPORT

A 28 years old male presented with complaint of fatigue, exertional dyspnoea and a low grade fever of two months duration. He looked pale. His pulse rate was 130 beats/min. and blood pressure was 120/80 mmHg. A grade 3/6 systolic murmur was audible over the left sternal edge and grade 2/6 diastolic murmur at the apex. The liver was smooth and palpable 3 cm below the right costal margin. The rest of the physical examination was within normal limits. His haemoglobin on admission was 9.2 G%, and erythrocyte sedimentation rate 105 mm/hr. Blood count and urinalysis were within normal limits. Chest X-ray showed moderate cardiac enlargement, predominantly of the right atrium and the right ventricle. The

pulmonary vascularity was slightly increased. The electrocardiogram showed right axis deviation and an incomplete right bundle branch block. 2-D echocardiography showed multiple echogenic masses in all the four cardiac chambers and the inferior vena cava (Fig. 1). Calcification was noted in the left ventricular outflow tract. Right heart angiocardiology demonstrated large filling defects moving in systole from right ventricle and right atrium into inferior vena cava (Fig. 2). Levophase follow through of the Angiogram showed the filling defect moving from the left atrium into the left ventricular outflow tract.

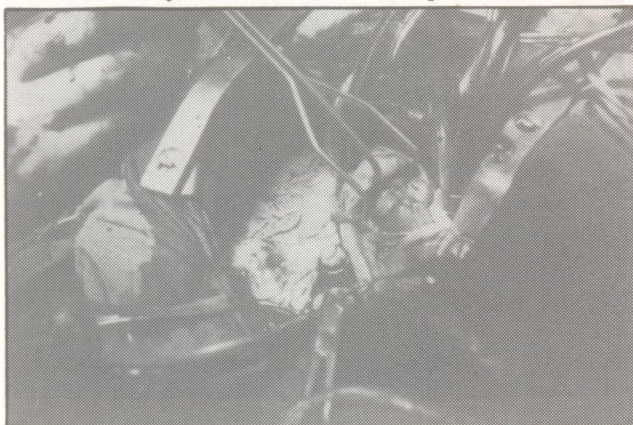


1. 2-D Echocardiography showing solid tumor masses in all 4 cardiac chambers.



2. A right heart angiogram showing the filling defects in the right atrium and right ventricle caused by the tumor.

The cardiac operation was performed through median sternotomy, with cardio-pulmonary bypass, moderate hypothermia and cold cardioplegic arrest. To prevent pulmonary embolization of the tumor the main pulmonary artery was occluded immediately prior to right atrial cannulation and going on bypass. When the right atrium was opened a large friable myxomatous mass was seen to occupy whole of the right atrium and protruding in and out of the tricuspid valve and inferior vena cava (Fig. 3). It was attached to the inter-atrial septum in the fossa ovalis area by a broad base. In the right ventricle, one large firm tumor with a smooth and haemorrhagic surface was attached by a short stalk to the endocardium of the free wall. A second smaller polypoid tumor was attached to the apical endocardium. The left atrial tumor was susage shaped, firm with a smooth surface and had calcific deposits. It originated from the inter-atrial septum in the Fossa ovalis area and protruded in and out of the mitral valve. In the left ventricle a small polypoid tumor was attached by a short stalk to the ventricular wall just beneath the posterior mitral



3. Operative photograph showing one of the tumor being delivered out of the right atriotomy.

leaflet near the postero medial commissure. The right and left atrial tumors were excised together with a portion of the inter-atrial septum. In the ventricles the tumor pedicles were excised along with the underlying endocardium. Access to the left ventricle was gained through an incision at the base of the posterior mitral leaflet which was later repaired with interrupted 5/0 prolene sutures. The defect in the inter-atrial septum was closed with a patch of Dacron. At the conclusion of the procedure the cardiac chambers were flushed with copious amounts of saline solution.

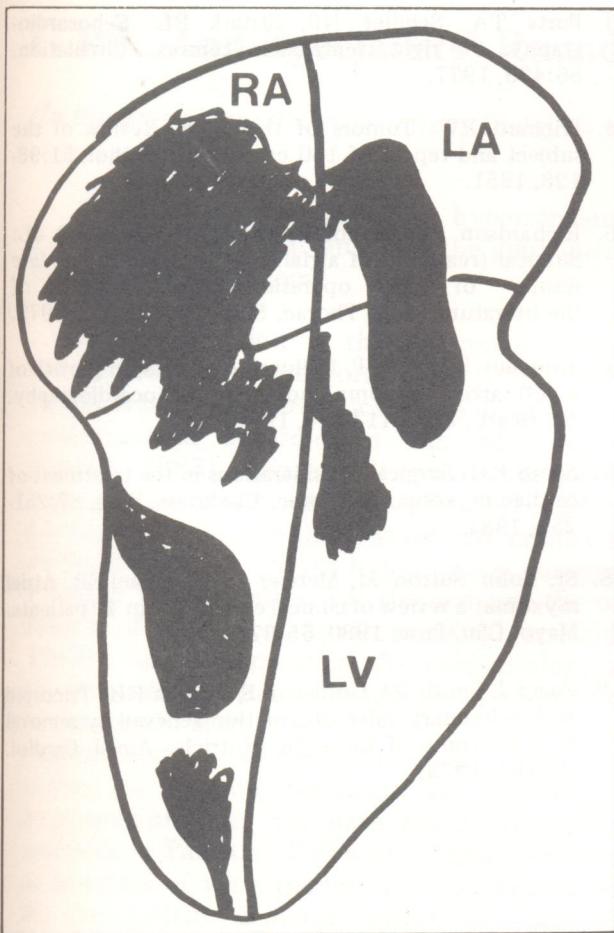
The post-operative course was uncomplicated. The tumor masses weighed 250 G. (Fig. 4). Histological examination showed myxoid stroma with variably dilated vascular channels. There was focal stromal haemorrhage and necrosis.



4. The tumor masses removed from all 4 Cardiac Chambers.

DISCUSSION

Primary tumors of the heart are uncommon with myxomas being the most frequent (9), usually they are lobulated single masses presenting as pedunculated left or right atrial tumor developing from Fossa Ovalis area of the inter-atrial septum (14). Occasionally these neoplasms are multiple in one or more cardiac chambers (1,2). Their location in all four cardiac chambers is quite unusual (Fig. 5). These tumors are generally very friable polypoid masses of pale, soft gelatinous tissue. Areas of calcification and haemorrhage are frequently encountered. Microscopically, the tumor consists of fibroblasts, multi-nucleated cells and round or polygonal cells within a polysaccharide rich myxoid stroma. Blood vessels are



5. Diagrammatic representation of the 5 tumor sites in the 4 Cardiac Chambers.

prominent at the base whereas peripherally the tumor is relatively avascular and lined by endothelium. Although pathologically, cardiac myxomas are benign they may grow rapidly and recur after resection (4, 6, 8, 16, 17). Systemic reactions such as fever, weight loss, anaemia and elevated erythrocyte sedimentation rate constitute the major clinical manifestations before the tumor is of significant size to cause mechanical obstruction. Patients may also present with symptoms of peripheral embolism or atrioventricular valvular obstructions (11, 12, 17). The presence of variable cardiac murmurs and atypical symptoms in a patient suspected of having valvular disease should arouse suspicion of myxoma. In our experience with cardiac myxomas the tumor has been a chance finding in patients undergoing evaluation for valvular disease.

Electrocardiogram and chest X-ray are often non-diagnostic although sometimes incomplete

right bundle branch block may be present. While 2-D echocardiography is usually diagnostic (3, 5, 7, 13) confirmatory angiography is still done in most cases especially if other lesions or coronary disease is to be excluded. In the case presented, angiography and 2-D Echo. beautifully demonstrated the tumors as intracardiac filling defects floating in and out of the cardiac chambers. Once diagnosis of cardiac myxoma is made, early operation is recommended to prevent complications. While standard cardio-pulmonary bypass techniques are employed, special care is taken to prevent tumor fragmentation and embolization (17). To this end, in our case the main pulmonary artery was occluded prior to right atrial cannulation. In the relaxed heart access to the cardiac chambers is gained through the right atriotomy and when needed, through the inter-atrial septum which in our case was excised together with the tumor. While the left ventricular cavity can be visualized through aortic or mitral orifice, the location of left ventricular tumor in our case necessitated an incision at the base of the posterior mitral leaflet. A large defect in the inter-atrial septum as in our case is usually closed with a patch of Dacron.

In conclusion, multiple cardiac myxomas are very rare. Early operation must be performed to prevent complications. The tumor must be completely excised to reduce chances of recurrence.

We thank Mr. Fasihuddin Alvi, Librarian, N.I.C.V.D., Karachi and Mr. Mohammad Arif, for their help in preparation of this presentation.

REFERENCES

1. Anderson ST, Pitt A, et al: A case of biatrial myxomas with successful surgical removal. *J. Thorac. Cardiovas. Surg.* 59:786, 1970.
2. Balk, A. H. M., Wagenaar, S.S., and Brusckhe, A.V.G.: Bilateral Cardiac Myxomas and Peripheral Myxomas in a Patient with Recent Myocardial Infarction. *Am. J. Cardiol.* 44:767, 1979.
3. Chandraratna PAN, San Pedro S, Elkins RC, Grantham N: Echocardiographic Angiocardiographic, and Surgical correlations in right Ventricular Myxoma simulating Valvular Pulmonic Stenosis. *Circulation* 5: 619, 1977.
4. Dang CR, Hurley FJ: Contra lateral recurrent myxoma of the heart, *Ann. Thorac, Surg.* 21: 59-62, 1971.

5. Dashkoff, N., Boersma, R.B., Nanda, N. C., Gramiask, R., Anderson, M.N., and Subramanian, S.: Bilateral Atrial Myxomas: Echocardiographic considerations. *Am. J. Med.* 65:361, 1978.
6. Desousa A. L., Muller, J., Campbell, R., Batnitzky, S., and Rankin, L.: Atrial Myxoma: A review of the Neurological complications, Metastases, and Recurrence. *J. Neurol. Neurosurg. Psych*, 41:1119, 1979.
7. Fitteret, J. D., Spicer, M.J., and Nelson, W.P.: Echocardiographic demonstration of Bilateral Atrial Myxoma, *Chest*, 70:281, 1976.
8. Gerbode F, Kerth JW, Hill DJ: Surgical management of tumors of the heart. *Surgery* 61:94, 1967.
9. Griffith CG: A review of primary tumors of the heart: *Prog. Cardiovas. Dis.* 7:465-479, 1965.
10. Larrieu AJ, Jamieson WRE, et al: Primary cardiac tumors, experience with 25 cases. *J. Thorac. Cardiovasc. Surg.* 83:339-348, 1982.
11. MacGregor GA, Gullen RA: The syndrome of feve, anaemia and high sedimentation rate with an atrial myxoma. *Br. Med. J.* 2:991, 1959.
12. Macrac FR, Galea M: Myxoma of the right ventricle of heart. *Can. Med. Asso. J.* 85:435-436, 1961.
13. Ports TA, Schiller NB, Strunk BL: Echocardiography of right ventricular tumors. *Circulation.* 56:439, 1977.
14. Prichard RW: Tumors of the heart. Review of the subject and report of 150 cases. *Arch. Pathol.* 51:98-128, 1951.
15. Richardson JV, Brandt B, Doty DB, Ehrenhaft JL: Surgical treatment of atrial myxomas. Early and late results of 11 operations and review of the literature. *Ann. Thorac. Surg.* 28:354-358, 1979.
16. Roudaut R, Gosse P, Dallochio M.: Rapid growth of a left atrial myxoma shown by Echocardiography. *Br. Heart . J.* 58:413-416, 1987.
17. Semb KH: Surgical considerations in the treatment of cardiac myxoma. *J. Thorac. Cardiovas. Surg.* 87:251-259, 1984.
18. St. John Sutton M, Mercier LA, Giuliani ER. Atrial myxoma: a review of clinical experience in 40 patients. *Mayo. Clin. Proc.* 1980, 55:371-176.
19. Zager J, Smith JO, Goldstein S, French RH: Tricuspid and pulmonary valve obstruction relieved by removal of a myxoma of the right ventricle. *Am.J. Cardiol.* 32:101, 1973.

