L E F T A T R I A L M Y X O M A

THESIS

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C O N T E N T S

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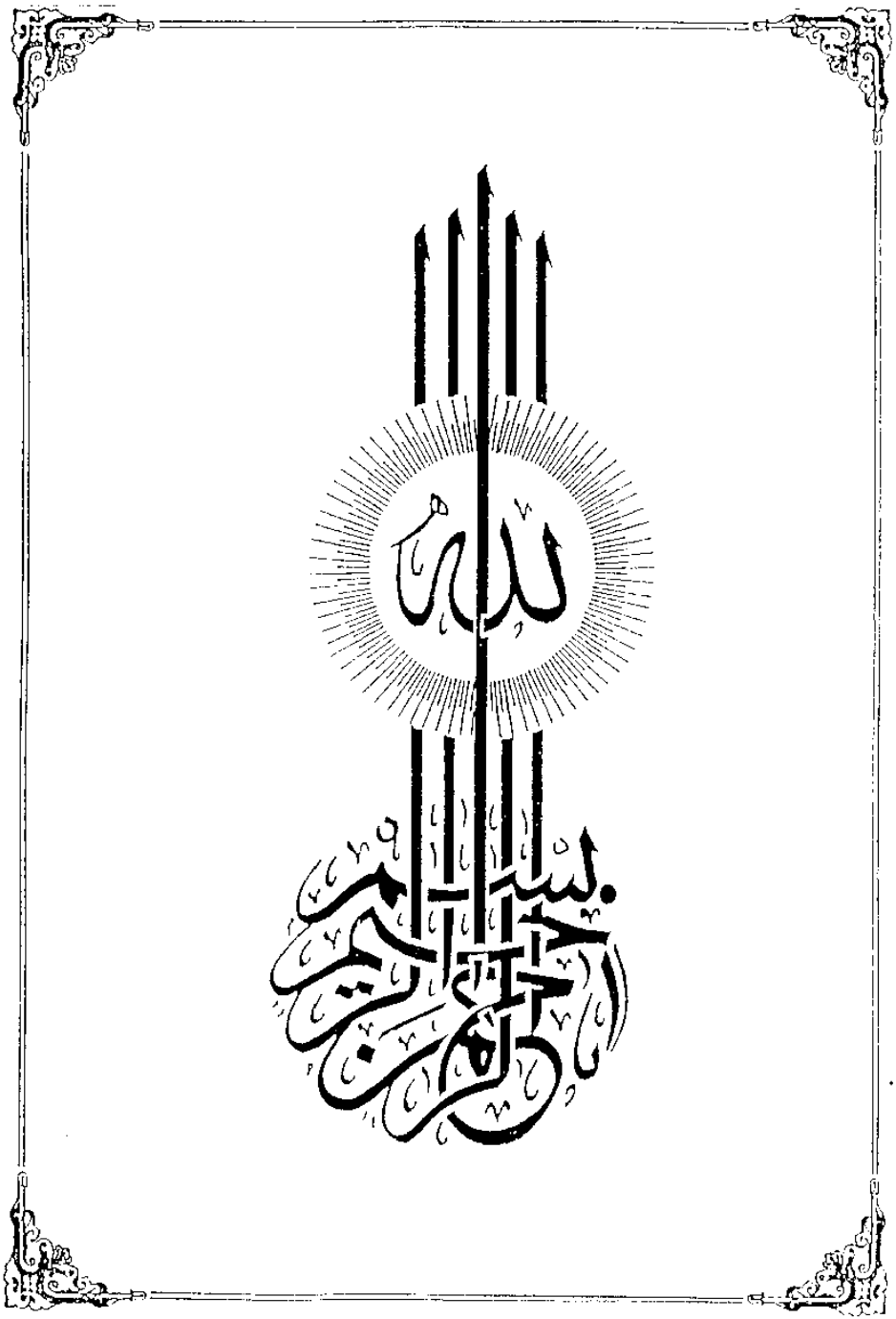

A C K N O W L E D G E M E N T

" THANKS TO GOD "

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INTRODUCTION

HISTORY AND INTRODUCTION

" The heart is too noble an organ to be attacked by a primary tumor " De Senac

Primary cardiac tumors are uncommon lesions that can mimic almost any type of cardiac disease, they have the potential for causing life-threatening complications and are frequently fatal as a consequence of their local growth. Antemortem diagnosis of intracardiac tumors was largely an academic matter since effective treatment was not possible. As recently as 1951, many believed that " the diagnosis of cardiac tumors is either impossible or a matter of chance " (Bulkley and Hutchins, 1979).

Although primary tumors of the heart have been recognized since as early as the 16th. Century, a correct antemortem diagnosis was not recorded until 1934.(Colucci and Braunwald, 1980). Only five patients, all of whom had primary sarcomas, were diagnosed during life before 1950. The first diagnosis, aided by angiography, and attempted surgical removal of an intracardiac myxoma was reported in 1952. (Goldberg, 1952).

Before the development of modern open heart surgical techniques, there were only rare reports of the successful removal of cardiac tumors, mostly on the

epicardial surface of the heart. Attempts to remove intracardiac tumors prior to the use of cardiopulmonary bypass were unsuccessful. (Castaneda, 1968).

In 1954 Crafoord performed the first successful excision of an intracardiac tumor, a left atrial myxoma, utilizing total cardiopulmonary bypass under direct vision. (Colucci and Braunwald, 1980). Since then, there have been many reports of the successful surgical excision of a wide variety of cardiac tumors, and in many instances a "complete" cure apparently has been achieved. (Castaneda, 1968).

The introduction of echocardiography in the diagnosis of cardiac tumors was a revolutionary step that changed the natural history of cardiac tumors. Since its first application in the diagnosis of intracavitary tumors in 1959, subsequently, increased clinical awareness coupled with the other non-invasive and angiographic techniques have led to more frequent correct diagnosis. (Hurst, 1982).

Since the incidence of cardiac tumors is extremely low, of the order of 0.0017 to 0.28 percent in the general population, the most important elements in arriving at the correct diagnosis are a high index of suspician and an appreciation of the necessary diagnostic procedures. (Mc Allister and Fenoglio, 1978).

RELATIVE INCIDENCE OF TUMORS OF THE HEART

(Table I)

Type	Number	Per Cent
Benign :		
Myxoma	130	30.5 %
Lipoma	45	10.5 %
Papillary fibroelastoma	42	9.9 %
Rhabdomyoma	36	8.5 %
Fibroma	17	4.0 %
Hemangioma	15	3.5 %
Teratoma	14	3.3 %
Mesothelioma of the AV-node	12	2.8 %
Granular cell tumor	3	-
Neurofibroma	3	-
Lymphangioma	2	-
Subtotal	319	75.1 %
Malignant : (1ry. & 2ry.)		
Angiosarcoma	39	9.2 %
Rhabdomyosarcoma	26	6.1 %
Fibrosarcoma	14	3.3 %
Malignant lymphoma	7	1.6 %
Extraskeletal osteosarcoma	5	-
Neurogenic sarcoma	4	-
Malignant teratoma	4	-
Thymoma	4	-
Leiomyosarcoma	1	-
Liposarcoma	1	-
Synovial sarcoma	1	-
Subtotal	106	24.9 %
Total	425	100.0 %

(Quoted from Tumors of the cardiovascular system from the Atlas of Tumor Pathology. Washington, D.C., Armed Forces Institute of Pathology, 1978.).

Intracardiac myxoma is the most frequent benign tumor of the heart. While most (75 %) are located in the left atrium, myxomas are also found in the right atrium (18 %), right ventricle (4 %), and left ventricle (4 %). (Hall and Cooley, 1982).

The aim of this work is to study retrospectively the six cases of left atrial myxomas removed at the Cardio-Thoracic Surgery Department of Ain Shams University Hospitals during the period between Jan. 1978 to Dec. 1983.

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REVIEW OF LITERATURE

PATHOLOGY OF LEFT ATRIAL MYXOMA

Macroscopic features and nature:-

The left atrial myxoma is usually found attached to the endocardium of the area of fossa ovalis by a braod base, it may be pedunculated, polypoid and friable, although some may have a smooth surface and be rounded. Sessile myxomas are uncommon. It appears as a soft, gelatinous, mucoid, usually grey-white mass, often with areas of haemorrhages or thrombosis. (Mc Allister and Fenoglio, 1978), (Colucci and Braunwald, 1980) and (Hall and Cooley, 1982).

Myxomas may arise rarely from sites other than the fossa ovalis. It has been known to arise rarely from the posterior wall of the left atrium near the left atrial appendage. (Friedberg, 1966). Sandrasagra et al., (1979) reported a case of left atrial myxoma having a pedicle attached to the anterior cusp of the mitral valve close to the lateral commissure.

Myxomas may be found single or rarely multiple, in more than one chamber of the heart commonly the right atrium. Atrial myxoma may pass through the foramen ovale and present in both atria. The tumor is usually shaped like a dumbell with the common stalk attached to

the margin of the fossa ovalis. (Hall and Cooley, 1982). Multichambered cardiac myxomas occasionally involve other chambers than the usual biatrial combination. A combination of a left ventricular and left atrial myxomas has been reported by Morgan et al.,1977. A combination of a left atrial and right ventricular myxomas was reported by Balk et al., (1979).

Multiple myxomas are more frequent in familial occurrences. (Liebler et al., 1976).

Left atrial myxomas vary from 1 - 15 cm. in diameter with most of them are measuring from 5 to 6 centimeters (Mc Allister and Fenoglio, 1978), (Bulkley and Hutchins, 1979) and (Colucci and Braunwald, 1980).

Considerable controversy exists about the neoplastic nature of cardiac myxomas, as its pathologic feature are very similar to those of an organized thrombus. Sayler and his colleagues (1975), proposed that myxomas are merely organized thrombi.

The most compelling arguements standing against the theory of a thrombotic origin are based upon the ultrastructural characteristics of the tumor cells which closely resemble multipotential mesenchymal cells rather than fibroblasts. (Frishman et al., 1979). In addition, the ability of myxomas to recur and their occurance in

multiple sites and in families all favour a neoplastic nature. (Siltanen et al., 1976),(Dang et al., 1976) and (Balk et al., 1979).

Atrial myxoma is not always benign. Reed et al., (1974), discussed the malignant potential of these neoplasms and recommended radical excision of the atrial septum to avoid local recurrence. Dang and Hurley, (1976), also discussed the local recurrence of atrial myxomas and reported one case of possible multicentric origin. Hannah et al., (1982), reported a case of left atrial myxoma with malignant behaviour which invaded the atrial septum and the conductive system which necessitated a pacemaker insertion during operation and a palliative subtotal excision of the tumor.

Light microscopy:-

The cells of left atrial myxoma (Lepidic cells) are small, polygonal in shape with rounded or oval nuclei and a moderate amount of cytoplasm. The cells are surrounded by myxomatous stroma composed predominantly of an eosinophilic matrix which appears to be composed of an acid mucopolysaccharide similar to chondroitin-C. (Heath, 1968).

Other cellular elements include lymphocytes, plasma cells, mast cells, histiocytes, and, rarely, fibrocytes. Thin-walled channels simulating primitive capillaries are present; these channels communicate from the surface to deep within the tumor and are lined by endothelial-like cells resembling multipurpose mesenchymal cells, from which the tumor is purported to arise. (Hall and Cooley, 1982).

The surface of the tumor consists of the typical myxoma cells and may show areas of organized thrombi . (Colucci and Braunwald, 1980).

Atypical glandular formations were described in a case of right atrial myxoma the cells of the glands resembled those of myxoma cells, this finding supports the theory that atrial myxomas are true neoplasms and that the cell of origin of that tumor is the multipotent mesenchymal cells. (Frishman et al., 1979).

Electron Microscopy:-

The myxoma cell contains a single large, irregulrly indented nucleus having a prominant nucleolus, few cytoplasmic organelles, a large number of thin filaments coursing irregularly through the cytosol. The plasma membrane shows many thin projections; the projections

of adjacent cells may interdigitate and become attached by desmosomes. The extracellular space contains collagen fibrils and electron dense granular precipitate which is focally condensed along the plasma membrane. No basement membrane is observed around the myxoma cells. (Frishman et al., 1979), (Colucci and Braunwald, 1980) and (Bortolotti et al., 1982).

Infected left atrial myxoma:

Left atrial myxoma may become rarely infected, and blood cultures may demonstrate a variety of organisms. Graham et al., (1976) reported a case of infected left atrial myxoma that was erroneously diagnosed as subacute bacterial endocarditis. The patient died before surgical intervention due to ruptured mycotic aneurysm.

Powers et al., (1979) reported a case of an infected right ventricular myxoma and reviewed the previous reports of seven cases of infected myxomas (six of them were left atrial) and they noticed that all the cases of infected left atrial myxomas presented with major neurological events and, interestingly, in all these cases there was no significant damage to the mitral valve, which was spared by both the tumor and infective process.