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DIAGNOSIS OF PRIMARY CARDIAC TUMORS: REPORT OF 30 CASES

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ABSTRACT

Between 1981 and 1993, 30 cases of primary cardiac tumors were diagnosed at the Imam Khomeini Hospital. 20 patients were female and 10 were male, with an average age of 32 years (ranging from 17 days to 65 years of age). The mean duration of symptoms before diagnosis was 6 months, with dyspnea and palpitation being the most common symptoms (60%) and cardiac murmurs the most usual signs (84%). The diagnosis of cardiac tumors was made by echocardiography in all patients.

Transesophageal echocardiography and cardiac catheterization were performed in 8 and 9 patients respectively for more precise localization of the tumor and evaluation of the coronary arteries. The left atrium was the only site of tumor in 60%. 4 patients had tumors in more than one chamber. 27 patients underwent surgery without mortality while 3 patients did not undergo surgery.

Pathological examination revealed benign myxoma in 23 patients, fibroma (1 patient), round cell sarcoma (1 patient), chondrosarcoma (1 patient), liposarcoma (1 patient), and undifferentiated sarcoma (1 patient).

Keywords: Primary Cardiac Tumors, Transesophageal Echocardiography

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INTRODUCTION

Most primary cardiac tumors are histologically benign,[1] but all of them are potentially lethal as a result of valvular obstruction, embolization and disturbances of rhythm or conduction. Early diagnosis and surgical excision are important and lifesaving. Nadas and Ellison reported that primary cardiac tumors are seen in 1: 10000 routine autopsies and are found ten to twenty times less frequently than cardiac involvement by secondary growth or direct extension.[2] In reviewing 500 primary cardiac tumors, Griffiths found 50% to be myxomas. Almost all of the malignant lesions were sarcomas.[3]

This report presents the clinicopathological spectrum and diagnosis and follow-up of 30 patients with cardiac tumors in our hospital.

PATIENTS AND METHODS

Between 1981 and 1993, 30 patients with primary cardiac tumors were diagnosed at our hospital. There were 20 female and 10 male patients, from 17 days to 65 years of age (mean: 32 years).

The hospital records and charts of these patients were reviewed. Relevant information was obtained from the history, physical examination, operative notes, and pathological reports. Autopsy reports were also reviewed. Follow-up of the patients was carried out by reviewing any outpatient notes available.

27 patients had undergone cardiac surgery and 3 patients had not due to refusal (1 patient), death due to staphylococcal septicemia before operation (1 patient), and extensive involvement of all cardiac chambers causing inoperability

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(autopsy was performed in this patient).

RESULTS

Clinical Presentation

Palpitation and dyspnea were the most frequent symptoms. The manifestations of right- sided heart failure were seen in 9 patients (30%), most of them having right-sided tumors. Other important cardiac manifestations were syncope (16.7%), rhythm disturbances (16.7%) and chest pain (10%). The most frequent finding in the physical examination was a diastolic murmur (Table I). Constitutional symptoms were frequently noted. Evidence of systemic emboli with resultant hemiplegia (permanent or transient) was noted in 13.5% of patients. One of the cases with right atrial chondrosarcoma had developed a pulmonary embolus.

Rhythm disturbances of particular importance were episodes of paroxysmal atrial tachycardia with heart block in one patient with right atrial myxoma, ventricular tachycardia in one patient with right ventricular fibroma, complete heart block in one patient with liposarcoma involving all cardiac chambers, and type A Wolff-Parkinson-White with episodes of very rapid supraventricular tachycardias (300 ppm) in one patient who had tumors in both ventricles. This 17 day old patient did not undergo operation. In follow-up, the tumor regressed in both ventricles and electrocardiographic changes disappeared.

Table I. Cardiac manifestations in patients.

Cardiac Manifestations	Number (percent)
Palpitation & dyspnea	18 (60%)
Diastolic murmur	21 (70%)
Systolic murmur	12 (40%)
Symptoms & signs of right-sided heart failure	9 (30%)
Syncope	5 (16.7%)
Arrhythmia	5 (16.7%)
Chest pain	3 (10%)

Table II. Anatomical location.

Site	Number (percent)
Left atrium	18 (60%)
Right atrium	5 (16.5%)
Right ventricle	2 (6.7%)
Left ventricle	1 (3.3%)
Left & right atrium	1 (3.3%)
Left atrium & left ventricle	1 (3.3%)
Left & right ventricle	1 (3.3%)
All cardiac chambers	1 (3.3%)
Total	30 (100%)

Table III. Pathological results.

Histological reports	Number
Benign tumors	24
Myxoma	23
Fibroma (RV)	1
Malignant tumors	4
Chondrosarcoma (RA)	1
Round cell sarcoma (RA)	1
Undifferentiated sarcoma (LA)	1
Liposarcoma	1
Unknown	2

Diagnosis

The diagnosis of all 30 patients was made by echocardiography. Transesophageal echocardiography (TEE) was performed in 8 patients. The left atrium was the only site of involvement in 18 patients (60%) as shown in Table II. In 4 patients more than one chamber was involved and in one of them all cardiac chambers were involved.

Cardiac catheterization was performed in 9 patients, of whom 4 elderly patients underwent coronary angiography. The hallmark of angiography was a filling defect in the related chamber.

Operative Results

27 patients underwent surgery without mortality. Echocardiographic findings were compatible with surgical findings with no false-positive or false-negative results. In 3 patients with malignant tumors who underwent surgery, the tumors were incompletely resected.

Pathological Findings

Pathological examination revealed benign myxoma in 23 patients as shown in Table III. The tumor pathology of 2 patients is unknown. One of them died before operation due to staphylococcal septicemia, and the other refused to undergo surgery. In follow-up of the latter patient utilizing echocardiography, the tumors gradually disappeared in both ventricles and the patient had normal growth and development. Later, skin lesions of tuberous sclerosis appeared, strongly suggesting that the tumors could have been rhabdomyomas.

Follow-Up

The range of patient follow-up was from 2 months to 126 months (mean: 74 months). All 3 patients with sarcoma who underwent surgery died because of widespread metastases. The patient with liposarcoma involving all four cardiac chambers was inoperable and died. One patient with myxoma of the left atrium had recurrence of the tumor after about 38 months and underwent reoperation. All of the other patients are free of recurrence and are in class I of the New York Heart Association classification.

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DISCUSSION

All 30 patients with intracardiac tumors were diagnosed by echocardiography. The location, mobility, size and shape of these tumors can be accurately assessed with 2-D echocardiography.[4-6]

Echocardiographic features of myxoma usually consist of pedunculated, mobile and sharply-demarcated masses. A left atrial tumor must always be differentiated from a thrombus by echocardiography,[7,8] especially if the tumor is of the nonprotruding type. A left atrial thrombus is usually a nonmobile mass with attachment to the posterior atrial wall, but in myxoma the site of tumor origin is usually the interatrial septum. Aside from this, the echocardiographic appearance of a thrombus is laminated and irregular. Other intracardiac masses including cysts, vegetations, calcifications, fibrosis and artifacts must be considered in the differential diagnosis of cardiac tumors.

Transesophageal echocardiography is particularly helpful because of its much better resolution in accurately determining the site of origin, size and number of tumors[9,10] (Figs. 1 and 2). By angiography the size, location and mobility of the tumor can be characterized.[11] Its major risk is peripheral embolization of a fragment of tumor.[12,13] In our patients, cardiac catheterization and angiography failed to reveal any additional diagnostic information than noninvasive evaluation of the tumors by echocardiography.

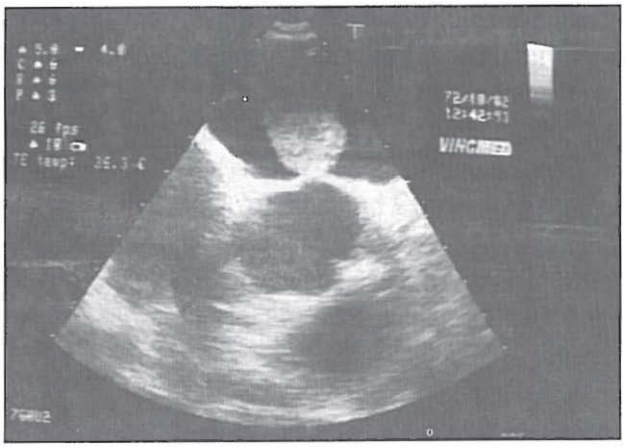

Fig. 1. Transesophageal echocardiography in a patient with left atrial myxoma.

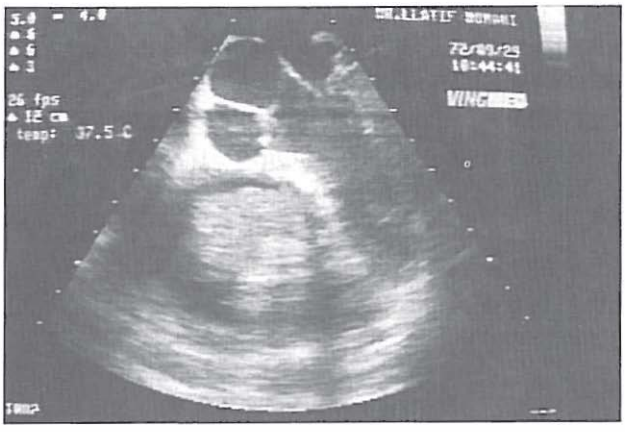

Fig. 2. Transesophageal echocardiography in a patient with a tumor in the right ventricle.

2-D echocardiography is highly sensitive for diagnosing primary cardiac tumors,[5,13-15] and transesophageal echocardiography can be used for more precise visualization of these tumors. With the availability of these noninvasive diagnostic procedures, cardiac catheterization is only justified for evaluation of coronary vessel anatomy before surgery and in many patients it is sufficiently reliable to permit operation without additional invasive studies.[4-6]

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