Primary cardiac tumors in Basrah Clinical and echocardiographic features

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Summary:

Baack ground: Primary cardiac tumors are rare disease and most of them are benign in nature, although these lesions are benign histopathologically, and they are potentially fatal because they may cause sudden hemodynmic obstruction and death of patient

The aim of this study: to report these rare cases to study their clinical, echocardiographic feature and their prognosis.

Patients and methods: Ten pts with primary cardiac tumors were studied from period of February 1994 to march 2001 clinically and by echocardiography.

Result: Eight pts were female, two pt were male. All pt were between third and sixth decade of life. Myxoma was the commonest cardiac tumors. it is more common in the left atrium (7pts) than right atrium (1pt). malignant pericardial mesothelioma was reported in one pt, and left ventricular angiosarcoma in one pt .Non of pt referred for echocardiographic study with suspicion of cardiac tumors in mind because of the non specific and different mode of presentation. Shortness of breath was the commonest presenting symptoms were occurred in 4(40%)pts follow by cerebral embolic emblization and stroke3(30%)pts. One pt present with picture mimic acute pericarditis.

Conclusion: cardiac tumours had different presentations and without complete surgical excision, these tumors whether begnin or malignant histopathologically, they were carried high morbidity and high mortality.

Introduction:

The heart can be the site of a host of primary as well as metastatic tumors. Although metastatic tumors of the heart are more frequent than previously realized. They seldom play a significant role in the patient's course [1]

Primary cardiac tumors are rare disease [1,2.3] and most of them are benign in nature, although these lesions are benign histopathologically, and they are potentially fatal because they may cause sudden hemodynmic obstruction and death of patient. [1, 2,3]

Intracavitary tumours are difficult to diagnose clinically, they can mimic valvular, pericardial or myocardial disease.
Cardiac myxoma is the most frequent and also clinically the most significant primary benign tumor of the heart, [4,5 6] for its discovery and treatment can be life saving

While the first account of left atrial myxoma diagnosed during life was not until 1951, In autopsy series, the incidence of primary tumors of the heart is evaluated at 0.0017% to 0.19%. [6]

It is now exceptional for the diagnosis to be made first at autopsy. [4] This is chiefly attributable to the wide availability of echocardiography, which has proved itself both reliable and specific tool for recognizing myxoma [4]

Cardiac myxomas are easily demonstrated by conventional transthoracic echocardiography [1-4] and it is usually the echocardiographer who first suspects and then confirms their presence; seldom has the patient been referred with this diagnosis in mind.
The echocardiographic appearance is so characteristic that angiography no longer has a role in diagnosis of myxoma. [4] The

only time to undertake it is in an older patient in whom there is fear of occult coronary artery disease. (4,5)
As a cause of left atrial obstruction, myxomas are 200 to 400 times less common than mitral stenosis. (4)
Atrial myxoma is treated by urgent surgical removal with its pedicle (2) it is important to ensure complete removal of the base by excising a full thickness button of the atrial septum. (1, 4)
The resulting defect is repaired with a small patch.

The rare recurrence after excision of the usual kind of myxoma usually occurs within the first 2 years; thereafter follow-up can safely be infrequent) .1, 4)

The aims of this study are to report these rare tumors, to study different presentation of these tumours and their prognosis. This is the first study on cardiac tumors in our city Basrah.

Patients and methods:

Ten patients (pts) with primary cardiac tumours were studied from the period of February 1994 to march 2001 prospectively. Eight pts were female, Two pts were male, their age range from 22-59years with mean age 40.4years.
Complete history was taken, full physical exam was performed, and investigation requested includes hemoglobin, erythrocyte sedimentation rate, and chest x-ray, 12 lead Electrocardiography, prothombine time &blood culture and sensitivity for all patients.
M Mode &2D echo with 2-4Mhz sector brobe with pts, in supine &left decubitus position were done, apical 4 chamber veiw, long axis, &short axis were studied. Coloured Echo and Doppler study were done when indicated
Transesophegeal echo, 3D echo were not done because of lacking facilities.CT scan of the heart was done in one pt. MRI was not done because of lacking facility. (7)

Results:

Out of ten pts two were male (20%), eight were female (80%), most pts are young age (20-40 years) and only two pts (20%) were above the age of 50 years. (See table I)

The commonest presentation was symptom was shortness of breath occurred in 4pts (40%). This was followed by cerebrovascular accident in 3pts(3o%), one of them got four attack of stroke in same year despite anticoagulant therapy and she die with last stroke because her family can't arrange for surgery for financial reason.

Severe generalized anasacra, which did not respond to heavy diuretic therapy, was the presenting finding in one pt.with right atrial myxoma. (Case no 7, table I)

An interesting observation that non-of the ten pts were referred with suspicion of primary cardiac tumours They referred with suspicion of mitral stenosis or for investigation of the cause of cardiomegally or heart failure.

Table two demonstrates the histopatholgical type of tumors. Interestingly this study reported very rare cardiac tumour (case no. 1) which is malignant pericardial mesothelioma (8,9) in 34years young pt (figure 1). This pt present with fever chest pain and pericardial rub on cardiac auscultation .He was admitted to hospital with professional diagnosis of viral pericarditis.but echocardiography show big pericardial tumour which prove later by histopatholgical exam as case of malignant pericardial mesothelioma. This pt was dead with constrictive pericarditis like picture due to expansion of tumor with possible right ventricular infiltration (figure 2,3) despite partial surgical resection and 8 session of radiotherapy postoperatively.

One patient had malignant angiosarcoma originated from left ventricle, who present with feature of congestive heart failure was reported in this study, but the commonest histopatholgical types were atrial myxoma reported in 8pts(80%), Four of pts undergo cardiac surgery with full excision of the tumors and their histopatholgical exam show atrial myxoma, The other four pts had classical clinical and echocardiographic features of myxoma (5) (See figure 4,5) but surgery can't be done for them for economic reason.

The commonest site for atrial myxoma was the left atrium occurred in 7pts(87.5%) and only one pts (12.5%) had right atrial tumour. This pt also had extension of tumor to superior and inferior vena cava. The left ventricle was the site of angiosarcoma in one pt. (case no.10).

The prognosis of pts with primary cardiac tumour whether malignant or begnin tumour were poor and carries high mortality and high morbidity. Non of them survive more than 2 years without cardiac surgery. Those who survive till time of the study they are severely disable either with neurological sequelies or the features of hemodynmic obstruction of tumors despite aggressive medical therapy with diuretic, digoxin and anticoagulant therapy. Atrial fibrillation was the commonest form of arrythmia. It was reported in 4pts (40%). Those pts whom underwent full surgical removal of the tumor, non-of them show evidence of recurrence of tumors but longer period of follow-up will be needed.

Table I show the pts charchterstic and presentation

Case	Age	Sex	presentation
Case 1	35	Male	Fever, Chest pain, and pericardial rub on auscultation (picture mimic viral pericarditis).
Case2	56	Female	Dyspnea orthopnea,with rapid AF
Case 3	27	Female	Dyspnea ,orthopnea,with diastolic murmur ,AF
Case 4	59	Female	Recurrent 4 stroke in one year,AF
Case5	37	Female	L. sided hemiplegia, AF& diastolic murmur? mitral stenosis
Case 6	34	Female	Referred with suspicion of mitral stenosis because dyspnoea and

			diastolic murmur
Case 7	36	Male	Generalized edema ,rise JVP,ascites & hepatomegally
Case8	50	Female	Fever wt lost,myalgia rise ESR,anemia
Case 9	30	Female	R. Sided hemiplegia
Case10	40	Female	Feature of congestive heart failure

Table II shows the site, Histopathological diagnosis and the prognosis of pts

Case	Site of lesion	Histopathological diagnosis	Prognosis
Case1	Pericardium	Malignant mesothelioma	Die after 18month despite partial resection and 8 session of radiotherapy
Case 2	Left atrium	Not done	Sudden death after 8 month of diagnosis
Case 3	Left atrium	myxoma	Free of recurrence 3month after surgery
Case 4	Left atrium	Not done	Die after 12 month of diagnosis
Case5	Left atrium	myxoma	Survive for last 2 years free from recurrence
Case 6	left atrium	Not done	Survive, Hemplegic ,for one year she cannot do surgery
Case 7	Right atrium	Not done	Dead within 8 month of diagnosis

Case8	**Left atrium**	**myxoma**	**Free from recurrence since 1994date of surgical resection**
Case9	**Left atrium**	**Not done**	**Hemplegic for last one year**
Case 10	**Left ventricle**	**Angiosarcoma**	**Die after 2years despite partial resection with CHF like state**

Discussion:

Primary cardiac tumors were more common in females (80%) than in males (20%) in this study, this is consistent with most of reports [1- 4] All cases occurred in age group range between 22to 55 years, and non was reported in this study in older age group or in childhood, this is consistent with most reports [1,2,3,4,10,11] . Jardine-DL reported atrial myxoma in 69 years old man but this case was only diagnosed on post mortem exam, this pt was managed clinically as case of recurrent pulmonary embolism. This case was missed because echocardiography was not done [11] Sobolewski-P; report left atrial myxoma in 7 years old child presented with cerebral embolization [10]

Shortness of breath was the commonest presenting symptom ,was seen in 4pts (40%), this was consistent with most of the studies. [1-5] Cerebral emblization was the second commonest presenting feature which occurred in 3pts(30%), one of them get 4 event in same year despite anticoagulant therapy, and this were frequently reported with Intracavitary tumors [1- 4, 6,10,15], Kassab R, also report recurrent strokes in pt with left atrial myxoma in 30 years old man [16]

Trailil stated that in left atrial myxoma , there were 40% incidence of stroke [4]

Benbouazza administered that myxoma represent 0.4% of stroke etiology [14]

Generalized edema was presenting feature of pt with R. atrial myxoma (case no 7), this resistant sever edema due to extension of tumour to superior and inferior vena cava this is again very

rare presentation. James H. reports similar case with involvement of both vena cava.
Pt with R.atrial myxoma rarely show systemic immobilization but it may be the source of recurrent pulmonary emboli, some cases of R.atrial tumour may be miss diagnosed as cases of recurrent pulmonary embolism, [11] some time Intracavitary thrombus may be misdiagnosed as myxoma (Figure 6)especially if this associated with antiphospholipid syndrome. [17- 19] Myxoma may trigger the formation of large thrombus by development of antiphospholipid syndrome
Quintanilla-S; Ferrer-S& Bravo-M postulate that interleukin-6 produced by the myxoma could trigger an immunological reaction leading to the primary antiphospholipid syndrome. [20]
Panos A report rare case of multiple embolisation to right coronary artery in pt with atrial myxoma causing acute myocardial infarction in 53 years old man. (21)
This study show that begnin tumors were more frequent than malignant tumors, this is consistent with most of the study (1,2,3,4,9)
The left atrium was the most frequent site of tumours7pts (70%) in this study, this also consistent with most of the study. (1,2,3,4,9)
Malignant mesothelioma is very rare condition. (8,12) It was reported in this study, this is the 1st time this case reported in our city, and I did not come across any report of this condition in our country.this case presented with picture mimic pericarditis. (See table 1) Oreoboulos report similar case on October 1997 (8)
The finding of high mortality and morbidity of these tumors if left without resection in this study is consistent with most other study which make surgery is mandatory for all these pts. (1,2,3,49,10)

Conclusion:

Although most cardiac tumors are histologically benign, they may be lethal because of their strategic position. Patients with an Intracavitary mass may present with protean symptoms, which may include one or more of a triad of obstructive
Constitutional or embolic symptoms.because of high mortality and morbidity of these tumors without surgery.this make early

diagnosis (by increase index of suspicion and echocardiographic exam) and surgical resection is only hope for that pts.

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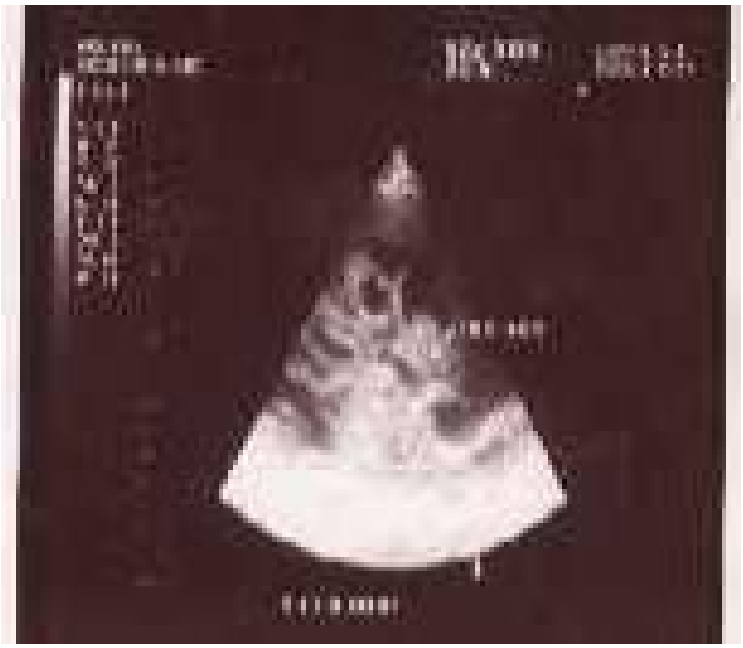

Figure 2-show ,two masses on this veiw during follow up of pt with malignant mesothelioma

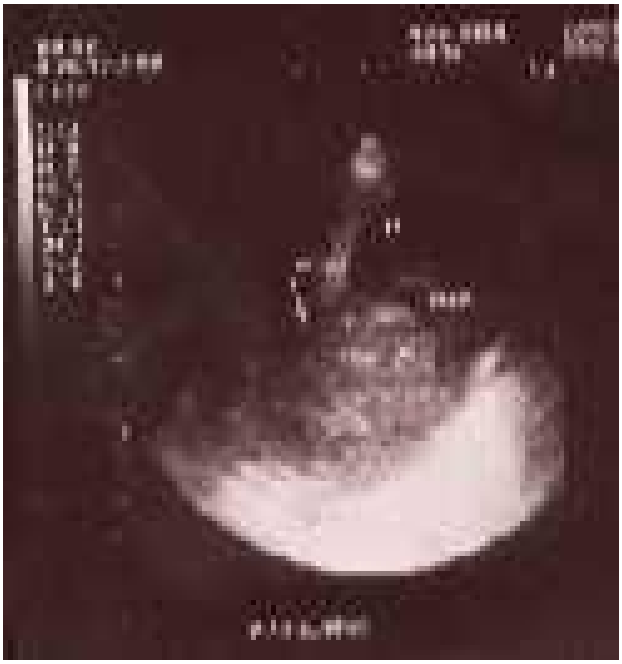

Figure 1 shows big pericardial mass (malignant mesothelioma

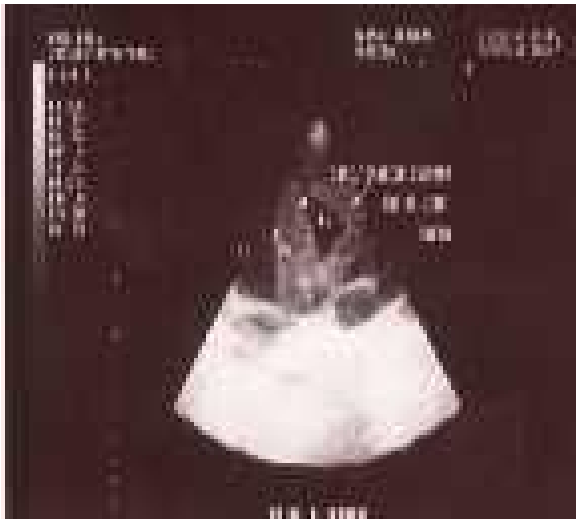

Figure 3 show increase thickness of right ventricle with possible infiltration with tumor pt with mesothelioma

Figure 5 show the Mode and 2D-echo pt with left atrial myxoma

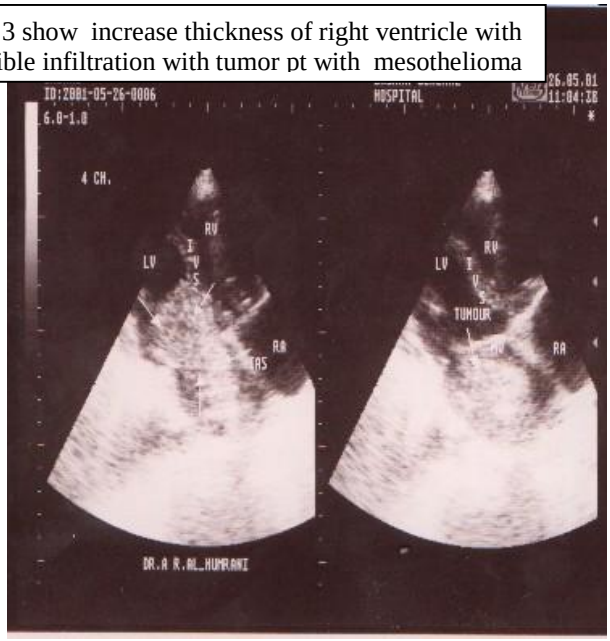

Figure 4 show classical echo feature of myxoma: big left atrial mass, which prolapse in diastole to left ventricle though mitral valve orifice

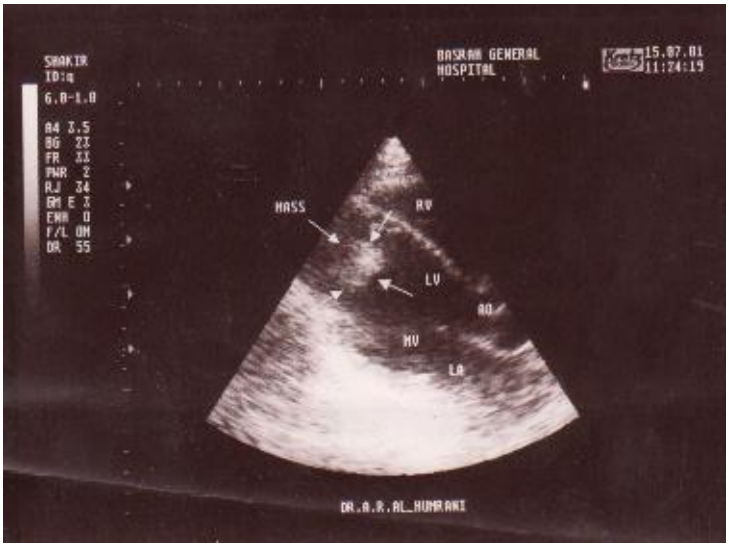

Figure no 6 show left ventricular mass due to thrombosis it is important to differentiate from cardiac