Leiomyosarcoma Of The Left Atrium Case Report And Review

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SUMMARY

Primary myocardial tumors are very rarely encountered. We describe a case of primary leiomyosarcoma arising in left atrium, which was partially resected. A brief review of the subject is also offered.

Key Words:

Cardiac Masses, Cardiac Tumor, leiomyosarcoma.

Case Report

A 54 years old, thin, lean and emaciated lady was admitted to the AFIC/NIHD, Rawalpindi with complaints of dyspnoea NYHA-III and extreme lethargy for last few months. She had no significant past medical history. Examination of CVS revealed a compensated heart with normal heart sounds and a ¼ diastolic murmur at apex without any postural variation. Rest of the physical examination was unremarkable. Her chest X-ray revealed prominent upper lobe vessels, heart was enlarged and left atrium dilated. Her Transthoracic echocardiography revealed an effective mitral valve area 2.9 cm^2 by Doppler, normal looking mitral valve apparatus, and no MR, TR or AR. A large, fixed LA mass arising from LA appendage was seen which was partially occluding mitral valve orifice. At this stage a diagnosis of LA thrombus due, possibly to mitral stenosis was entertained and patient started on anticoagulant therapy and preparations for surgery started. Meanwhile certain atypical features alerted the clinician and a repeat Transthoracic echo was performed followed by transesophageal study. ***It showed a large globular mass in the left atrium, almost completely occupying it, with beautiful spongiform consistency, with a brilliant sparkle, and scattered areas of hypolucency, which gave it an ornamental look. It had connection to the left atrial appendage, (Figures 1 & 2).*** At this stage the mass was thought to be a cardiac tumor. Detailed systemic examination revealed no possible primary site, but her abdominal sonogram revealed enlarged para-aortic lymph nodes.

She was operated through a median sternotomy. There were adhesions all around the heart. Left atrium was approached trans-septally, there was no clot in the left atrium, and mitral valve was perfectly normal. A big mass of a grayish-white solid tumor arising from the left atrial wall was lying inside the left atrium almost occupying it and obstructing the mitral valve orifice. Extra-cardiac extent of the tumor could not be ascertained but major vessels seemed to be unaffected. Radical resection was not possible but the accessable portion was excised which had a very tough and gritty feeling. Her post operative course was very rough due to low cardiac output and bleeding from the adhesions. She left the ICU on 5th post operative day. Histopathology of the tumor showed it to be a leiomyosarcoma. She was referred to oncologist for staging and management and is receiving chemotherapy presently.

Discussion

Intracardiac masses are encountered not infrequently in busy cardiac centers. Most common

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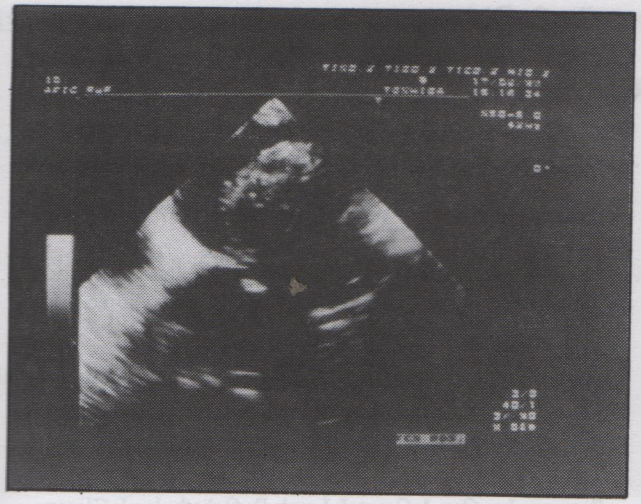
cause is thrombus in a cardiac chamber due to stasis, infarction or infective process. Alternate pathologies are comparatively so rare that these are seldom considered as the primary diagnoses unless the aforementioned causes have been excluded. This was duly highlighted in our case by the fact that initial impression of the clinician was LA clot secondary to mitral stenosis. However, certain atypical features of the case altered him to consider other possibilities; normal mitral valve apparatus, sinus rhythm, relatively mild atrial enlargement, absence of spontaneous contrast in LA, absence of factors predisposing to thrombogenesis, late onset of disease and debiliated condition of the patient. It was consideration of these factors that prompted for diagnosis of a cardiac tumor.



The heart may be the site of a primary tumor or invaded secondarily by malignancies arising in adjacent or remote organs. Primary tumors of heart and pericardium are rare, occurring with a frequency ranging from 0.001 to 0.28% various reported post-mortem series[1,2]. More than 75% are benign, approximately 50% being myxomas. Lieomyosarcoma is amongst the rarest tumors of the heart with a reported incidence of 0.02% of cardiac tumors[3]. Segesser-Lv et al. could find only 15 cases of primary cardiac literature reported in literature[4].

Being a tumor of smooth muscles, leiomyosarcoma may arise in any location. Usually reteroperitoneal, intra-abdominal tumors are encountered; however leiomyosarcomas arising from veins are also seen[5]. Leiomyosarcoma of the heart is usually listed as primary tumor of heart. Because of scarcity of tumor, exact cytologic origin has not been mentioned in literature. It is conceivable that the tumor may arise from veins within the heart or from in-coming veins like IVC and pulmonary veins. As most of the case reports describe atrial tumors, it would lend support to the second postulate, i.e., origin of tumor in incoming veins with extension into the heart. Unfortunately, the literature fails to mention whether any attempt was made to find primary site of tumor origin.

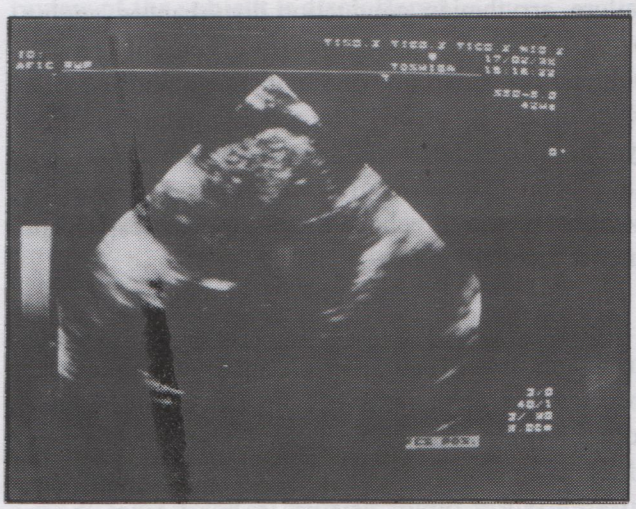
Clinical manifestations of cardiac tumors may be protean; pericardial involvement leading to pain, effusion, tamponade, arrythmias or constriction, myocardial involvement with cardiomegaly, heart failure, arrythmias, conduction disturbances, angina or infarction by coronary involvement, cavity obliteration, valve obstruction/damage and embolic phenomena[6]. Relatively non specific nature of these symptoms coupled with rarity of the condition may have been the cause of fewer reports. However, widespread use of echocardiography has resulted in greater recognition rate[7].

Echocardiography has assumed the role of primary investigation and may provide most of the desired information non-invasively. However, it can not accurately delineate pericardial involvement and extent of extra-cardiac involvement. Also, it can not differentiate a tumor arising in heart from those invading the heart from surrounding structures. Generally speaking, trans-thoracic echocardiography can detect most cases with high sensitivity. However, transesophageal echocardiography is superior in correct delineation of tumor due to its superb resolution and ability to negotiate right sided lesions and in those with poor quality Transthoracic echocardiographs[8-11].

CT scan of the chest is another investigation of immense value; not only can it define intracardiac mass, it can accurately detect pericardial involvement and extra-cardiac extent of tumor[12]. Ultra fast or cine CT is superior because of its less examination time and lack of motion artifact induced by cardiac movement[13]. MRI may provide additional information concerning heart, mediastinum, thorax and pleura, which may prove invaluable in surgical decision making and planning management of malignant tumors[14,15,16]. Contrast enhanced MRI with Gd-DTPA can be used for metastatic malignant tumors[17].

Recently, immunochemistry using cyto-chemical markers is being used extensively for diagnosis of these tumors.

Management and prognosis of benign and malignant cardiac tumors are understandably very different. Benign tumors are generally easily resectable and have good long term prognosis. On the contrary, malignant tumors are rarely resectable completely and have poor prognosis in spite of surgical resection and other adjuvant modes of therapy. A mean survival ranging from 3 months to 9 months after surgery has been reported[18,19], although occasional reports of survival up to 29 months of follow up have been seen[20].



Optimal management of the condition has not been decided upon. Surgical resection is usually first attempted, even though tumor is seldom completely resectable and post-surgical survival is far from satisfactory. There is even a view that if histological diagnosis is available before surgery, surgery should only be contemplated for palliation. Adjuvant therapies like radiotherapy or chemotherapy may be offered in face of incomplete resection. Because of rarity of the condition, few comparative trials are available on merits of surgery alone or in combination with chemotherapy or radiotherapy, though these are known to prolong survival[21]. Cardiac transplantation has been utilized to completely resect an inoperable tumor[22]. Cardiac explantation and autotransplantation may facilitate resection of some cardiac tumors[23].

In conclusion, cardiac tumor should be considered in differential diagnosis of all cardiac masses. If a tumor is suspected, all attempts should be made to define site, extent and origin of the tumor before making final management decisions. Transthoracic and transesophageal echocardiography, CT Scan and MRI should be obtained if available. A thorough search should be made for finding an extra-cardiac primary site or distant metastasis. If accessible, endomyocardial biopsy may give histological diagnosis before surgery. If malignant nature of the tumor has become known, risks and expected benefits of surgery should be carefully weighed.

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