MULTIPLE LEFT VENTRICULAR MYXOMA: CASE REPORT
AND REVIEW OF THE LITERATURE

ABDULELAH F. MOBEIREK, FRCP(C); MANSOUR AL-NOZHA, FRCP

A CASE OF MULTIPLE left ventricular myxoma is presented. The myxoma originated from the annulus of the mitral valve with involvement of the posterior mitral leaflet. The posterior leaflet was encased by the tumor which was slowly moving to the atrial surface of that leaflet. Another tumor mass, originating in the left ventricular outflow tract, was near the annulus of the anterior mitral leaflet. This tumor mass was pedunculated and prolapsed into the left ventricular outflow tract (L VOT) during the cardiac cycle. The patient presented with stroke. His echocardiographic features, both transthoracic and transesophageal, are described. The patient underwent resection of the tumor and mitral valve replacement. A review of the literature of this uncommon tumor is presented with emphasis on the role of transesophageal echocardiography in the preoperative evaluation.

Case Report

A 31-year-old male was referred to King Khalid University Hospital (KKUH) with history of sudden weakness of the right arm and leg and aphasia. He was completely healthy prior to this illness which occurred after engaging in heavy exercise. There was no history of dyspnea, fever, or syncope. Physical examination revealed motor aphasia and right-sided weakness. There was a soft ejection murmur at the apex. CT scan revealed a large, low, attenuating lesion in the left temporo-parietal region involving the left internal capsule with minimal enhancement after contrast injection. Carotid angiogram revealed reduction of vascularity in the middle cerebral artery. There was evidence of mild anemia (Hb = 10.9 g/dL) and leukocytosis (WBC = 16300/L). The ESR was 25/first hour.

Transthoracic echocardiography (TIE) revealed normal-sized cardiac chambers and two relatively large cardiac masses within the left ventricular cavity that were mobile and pedunculated. They were attached near the mitral annulus on both sides. There was no evidence of mitral valve obstruction and no significant gradient across the left ventricular outflow tract (L VOT) by continuous wave doppler.

Transesophageal echocardiogram (TEE) was performed to assess the attachment of the mass, to detect involvement of the mitral valve and its associated structures, and to look for additional tumors. TEE showed two large masses with excessive motion with two separate origins (Figure 1 A). One was attached to the ventricular surface of the posterior mitral valve leaflet; this was a sessile mass. The tumor was seen to be extended to the atrial side of the posterior leaflet through the mitral valve orifice with tumor involvement on both sides of the posterior leaflet. The second tumor mass was pedunculated with a long stalk. This mass was attached to the annulus of the anterior mitral leaflet and was prolapsing in the L VOT with each systole (Figure 1B).

The patient underwent removal of both tumors. They were large and friable with a gelatinous appearance (Figure 2). Because of tumor involvement, it was necessary to replace the mitral valve with a carbomedic size 29-mm prosthesis. The histopathology report confirmed that it was...
Primary cardiac tumors are rare with an estimated incidence in unselected autopsy series from 0.001 % to 0.28%. Metastatic tumors are more common than primary tumors of the heart (10 to 40 times more frequent); of the primary tumors involving the heart, approximately 75% are benign and 25% are malignant.1,2

This patient demonstrates unusual features - the rare development in atypical location, being multiple with multicentric origin and involvement of the mitral valve.3 The location within the ventricle is rare. About 95% of cardiac myxomas originate in the atria (approximately 75% in the left atrium and 20% in the right atrium) and about 5% originate in the ventricle with equal incidence.2,4 Thus, left ventricular myxoma is a rare condition, accounting for only 2.5% of myxomas.5-7 The most common tumor in the left ventricle is fibroma. Another rare location is within the pulmonary artery (0.1 %).

Furthermore, the majority of myxomas are solitary and only a few are multiple. Only 3% of all myxomas in general are multiple. Soma et al noted that among the 30 cases reported in the literature of primary left ventricular (LV) myxoma, there were only 2 cases that had multiple tumors.5 Multicentric tumors are more likely to recur than solitary tumors and may be associated with "syndrome myxoma."

In a review of the literature, in 1977, Meller et al noted the low prevalence of LV myxoma.8 At that time, only 15 cases had been reported in the English and French literature. Meller reported the first case of LV myxoma diagnosed using M-mode echocardiography.8 In the same year, Morgan et al subsequently reported the first case of myxoma with separate LV and left atrial (LA) origins detected by M-mode echocardiography.9

In 1982, Mazer and Harrigan reported the first case of LV myxoma diagnosed by 2-D echocardiography.10 Abramowitz et al were the first to report 2-D echocardiographic diagnoses of separate myxomas of both LV and LA in 1984.11 In a recent report, Panady et al reported a case of single LV myxoma. A review of English literature revealed only 22 cases of LV myxoma reported.12 In a subsequent report, Vargas-Barron and associates reported a case with myxoma in the four cardiac cavities.13 More recently, Soma et ai, in 1992, noted there were about 30 cases reported with LV myxoma in the literature.5
The clinical features of myxoma are protean, mimicking a variety of clinical entities. The presenting symptoms often resemble those of primary mitral valve disease, endocarditis, cerebrovascular accident, connective tissue disease, or primary pulmonary hypertension.1,2,14

There are some important pathological and clinical differences between LV and LA myxomas. While LA myxomas arise mostly from the septum in the vicinity of the fossa ovalis, LV myxomas tend not to arise from the septum. The tumor usually arises from the free wall or from the annulus. Ventricular myxomas tend to occur at a younger age and with similar sex predilection.4,10 However, some authors noted a female predominance that is similar to atrial myxoma.2,7,15 Some authors have reported a female predominance that is similar to atrial myxoma.2,7,15 They may produce obstruction of LVOT or RVOT with hemodynamic consequences of obstruction.

LV myxomas present with symptoms caused by embolization and obstruction. They may cause obstruction to LVOT producing dyspnea, syncope, chest pain, or may cause sudden death. These symptoms may be positional. They may also cause arrhythmias, conduction disturbances, and LV dysfunction.2,6,15,16 Clinically, they may simulate aortic stenosis17 and hypertrophic cardiomyopathy.

Embolic phenomenon is noted to occur more frequently with LV myxoma compared to LA myxomas, occurring in 64% of patients with LV myxoma.6 Syncopal attacks from LV outflow obstruction occur in up to 50% of cases.6,15 Constitutional symptoms have been reported to occur frequently with atrial myxomas and less frequently with ventricular myxomas. Of the 16 patients with LV myxoma described by Meller, seven had died of embolic complications while one suffered permanent neurologic deficit.8

This case also emphasizes the importance of TEE in the preoperative evaluation of patients with myxoma. It is only recently that the usefulness of TEE is being described. TEE allows superior visualization of the atria and atrial septum. Because of the proximity of the posterior structures to the probe and the high frequency of the transducer, TEE provides further information regarding tumor mobility, surface characteristics and, importantly, clarification of the tumor’s site of attachment, as well as verification of tumor involvement and functional integrity of the atrioventricular valves. The other cardiac chambers should also be inspected for the presence of the uncommon multicentric tumors (about 3% to 5% of myxoma) which may be small and beyond the resolution of TTE. In this patient, TTE was not helpful in clearly establishing the tumor attachment size. We believe that the information gained by TEE aids in planning the type of surgery. TEE was used to plan the operative approach. TEE is important for clarifying the tumor’s site of attachment and to check on possible involvement of other structures that might influence the operative procedure. The superior imaging of TEE compared to TTE should enhance preoperative assessment.

In most of these patients, echocardiography may eliminate the need for cardiac catheterization and its attendant risks, which include myxoma disruption and subsequent systemic embolization. Thus, one can proceed to excise the tumor without further diagnostic testing.1,2,14

The TEE modality used in this case was the monoplane probe (Hewlett Packard, 5.0 MHz, phased array imaging transducer). It is likely that the use of biplane and multiplane probes, which incorporate additional imaging planes, will add to the diagnostic ability of TEE. Thus, tumor attachments will be better delineated.

Recurrent myxomas have been reported in about 5% of patients after resection of the initial tumor with a higher recurrence rate in patients with multiple tumors (33%), patients with familial myxoma (10%), and when patients have other abnormalities like Cushing's syndrome, testicular tumors, or pituitary edema (myxoma complex) (21%) than in sporadic myxomas (1% to 3%).17,19 Therefore, regular follow-up is advised after surgical resection.

References


