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Surgical Progress

Primary Cardiac Tumors

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Cardiac tumors are a rare, but potentially curably form of heart disease. A high index of clinical suspicion is necessary for diagnosis as these tumors have protean manifestations that mimic a variety of other cardiac and noncardiac diseases. Presently, M-mode and two-dimensional echocardiography are utilized as safe, reliable, and noninvasive imaging modalities. Seventy-five per cent of these tumors are benign, with myxoma accounting for 50% and rhabdomyoma comprising 20% of lesions. Various histologic types of sarcoma are the predominant malignant cardiac neoplasms. With strict attention to avoiding perioperative tumor embolization, surgical resection of these lesions can be accomplished with minimal morbidity and mortality. Sixteen consecutive primary tumors of the heart have been surgically treated at Duke University Medical Center since 1966 with no perioperative deaths and no late recurrences.

RECENT ADVANCES IN DIAGNOSTIC imaging modalities and cardiac surgical techniques have altered the clinical importance of cardiac tumors. No longer are these rare neoplasms only diagnosed by the pathologist in the autopsy room, but these tumors have become an often surgically curably form of heart disease. The purpose of this review is to discuss the many clinical presentations of cardiac tumors in general, then focus upon certain specific neoplasms, in particular on myxomas, and finally to present the surgical experience with cardiac tumors accrued at Duke University Medical Center since 1966.

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Historical Aspects

Although cardiac tumors have been recognized since Columbus' report in 1559,[10] these lesions were pathologic curiosities until Barnes' initial clinical diagnosis of a primary sarcoma of the heart in 1934.[3] Further progress accompanied the development of more sophisticated diagnostic and surgical techniques. Whereas Beck removed an intrapericardial teratoma in 1942,[4] the first successful resection of a tumor arising from the heart was by Maurer in 1951.[38] Goldberg, et al., in 1952 made the first preoperative diagnosis of an intracavitary myxoma using angiocardiography.[23] A myxoma was then successfully resected in 1954 by Craaford,[11] a feat duplicated by Bigelow one year later.[6] The most recent significant milestone was the introduction by Shattenberg in 1968 of echocardiography as a reliable imaging modality for cardiac tumors.[49]

Incidence and Classification

Primary cardiac tumors are rare. In the largest consecutive autopsy series, Straus and Merliss reviewed 480,331 cases from 1938–1942 and found the incidence of primary cardiac tumors to be only 0.0017%.[56] Benjamin found primary tumors in 0.03% of 40,000 necrosies.[5] In contrast, tumors metastatic to the heart are significantly more common, being

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TABLE 1. *Classification of Primary Cardiac Tumors*

Classification
Benign (75%)
myxoma (50%)
rhabdomyoma (20%)
Lambl's excrescence
fibroma
lipoma
hemangioma
lymphangioma
mesothelioma
teratoma
thyroid adenoma
chemodectoma
neurilemmoma
ganglioneuroma
valve cyst
granular cell myoblastoma
Malignant (25%)
sarcoma (20%)
angiosarcoma
hemangioendotheliosarcoma
Kaposi's sarcoma
rhabdomyosarcoma
leiomyosarcoma
osteosarcoma
chondrosarcoma
neurogenic sarcoma
lymphoma
plasmacytoma
mesenchymoma

found 20–30 times more often than primary lesions in various series.[44]

Primary tumors can be categorized as benign or malignant (Table 1). Seventy-five per cent of primary tumors are benign, with myxomas comprising 50% of the lesions in most series. Rhabdomyoma is the second most common benign tumor, and is the most frequent neoplasm occurring in childhood. Lambl's excrescence, although thought by some to be a variant of myxoma,[47] is considered by most authorities to be a distinct entity.[7] Fibroma, lipoma, hemangioma and lymphangioma are tumors of mesodermal origin that infrequently arise from the heart. Mesothelioma, teratoma and thyroid tumors are lesions of heterotropic tissue that can arise from cardiac structures because of the close relationship in embryogenesis between the endodermal and gonadal germ cells and the primitive myocardium.[7] Chemoreceptor and sympathetic nervous tissue can rarely give rise to cardiac chemodectoma,[25] neurilemmoma, and ganglioneuroma.[15] Valve cysts[35] and granular cell myoblastoma[7] have also been reported.

Malignant tumors account for approximately 25% of primary cardiac neoplasms, and these are predominantly various forms of sarcoma. Cardiac sarcomas are usually poorly differentiated and this often makes precise histologic classification extremely difficult. The most common sarcomas are tumors of vascular origin, in particular, angiosarcoma.[22,52] However, all varieties of bony, neurogenic, and soft tissue sarcomas have been reported to arise from cardiac tissue.[48] In addition, anecdotal reports of primary lymphoma,[55] plasmacytoma,[59] and malignant mesenchymoma[7] of the heart have appeared in the literature.

Clinical Signs and Symptoms

The clinical manifestations of cardiac tumors are protean.[51] They produce symptoms by their mass effect, local invasion, embolization, or sytemic constitutional manifestations. Primary cardiac tumors must be in the differential diagnosis of any patient who presents with one or a combination of the following symptoms complexes:[29]

Pericardial Involvement

Neoplastic infiltration of the pericardium can produce pericarditis with resultant effusion and tamponade. In addition, the pericardium can become noncompliant resulting in a clinical presentation indistinguishable from constrictive pericarditis of infectious or idiopathic origin.

Congestive Heart Failure

The mass effect of an intracavitary tumor can obstruct the transfer of blood through the cardiac chambers or can interfere with the normal coaptation and opening of the cardiac valves. Depending which chambers or valves are involved, the patient may present with syncope, angina, dyspnea, edema, ascites, or any combination of murmurs indicative of valvar stenosis or incompetence. In addition, neoplastic involvement of the myocardium may impede myocardial pump function resulting in a presentation of congestive heart failure similar to a cardiomyopathy.

Pulmonary Hypertension

Elevated pulmonary artery pressures can be caused by obstruction of the pulmonary vessels by tumor emboli, but more commonly is a result of pulmonary venous hypertension due to obstruction to left heart filling.

Embolization

Systemic emboli from left heart lesions, in particular left atrial myxoma, are much more frequently seen than pulmonary tumor emboli from right heart lesions.

Arrhythmia

Certain patients with cardiac tumor manifest only recurrent supraventricular or ventricular arrhythmias,

most likely due to the irritative effect of tumor invading cardiac muscle. More rarely, the conduction system is injured resulting in heart block and Stokes-Adams attacks.

Chest Pain

Pain is a common manifestation of malignant tumors, but rare instances of ischemic cardiac pain due to tumor embolization to the coronary arteries[18] or extrinsic compression of myocardial vessels have been reported.[31]

Constitutional Symptoms

Fever, malaise, weight loss, polymyositis, hepatic dysfunction, Raynaud's phenomenon, hyperglobulinemia and an elevated erythrocyte sedimentation rate have all been described in both benign and malignant lesions but most frequently are associated with left atrial myxomas. The etiology of these phenomena is speculative. The leading theories postulate either an autoimmune or a systemic response to altered serum proteins damaged by tumor movement and to tumor breakdown products released into the circulation because of necrosis or hemorrhage. The hyperglobulinemia is always polyclonal and is completely reversible after tumor resection.[30] There are no qualitative, just quantitative serum protein abnormalities. The elevated globulins probably explain the concomitant elevated erythrocyte sedimentation rate.

Hematologic Abnormalities

The hemolytic anemia and thrombocytopenia, again often noted in left atrial myxomas, are felt to be due to the mechanical destruction of these blood elements by a mobile intraluminal tumor.[61] Polycythemia is seen on occasion with right atrial tumors and most likely represents the response to hypoxia when the tricuspid valve is obstructed with subsequent elevation of right atrial pressure, stretching of the foramen ovale, and right to left shunting.[24]

Diagnostic Modalities

The diagnostic modalities available are more or less specific for demonstrating a cardiac mass lesion. The plain chest x-ray merely reflects the anatomic changes which have occurred because of the hemodynamic consequences of the tumor. Direct evidence of a cardiac tumor is manifest only in the infrequent instances when tumor calcification is noted. Right atrial myxomas are the most common tumors that demonstrate this calcification. Similarly, the electrocardiogram gives only indirect evidence of cardiac tumors by showing chamber enlargement or rhythm disturbances. Heart scans using radioiodinated serum albumin may demonstrate intracavitary defects consistent with a tumor. Gated radionuclide cardiac imaging has increased the accuracy of this technique, but these defects may also represent thrombus or an infected vegetation.[42]

Phonocardiography is a useful technique for detecting a mobile atrial mass.[65] The characteristic phonocardiographic clue to a left atrial myxoma is an increased intensity of the first heart sound with subsequent vibrations extending throughout the isovolumic contraction period (Fig. 1). In addition, the murmurs generated by valve dysfunction can be recorded as well as an early diastolic sound which is felt to represent prolapse of the tumor into the ventricle. The notched upstroke in the apex cardiogram, once thought to be of diagnostic significance, is now felt to occur often in normal subjects.

Cardiac catheterization and angiography may be hazardous in patients with cardiac tumors because of the risk of tumor embolization. In particular, transseptal puncture of the left atrium is contraindicated.[41] The intracavitary filling defects noted on angiography may represent a primary cardiac tumor, however, false-positive angiograms have been caused by abscess, thrombus, hematoma, lipomatous hypertrophy of the atrial septum, metastatic tumor, and aortic aneurysm. Pressure measurements at the time of catheterization may be of diagnostic help.[8] A mobile atrial tumor can cause large spontaneous variations in the end diastolic pressure gradient across the atrioventricular valve. This occurs independently of heart rate or rhythm and presumably reflects tumor movement resulting in various degrees of valvar obstruction (Fig. 2). Additionally, large changes in the diastolic gradient varying with respiration have been noted with atrial tumors.

Echocardiography is perhaps the most important modality available today for imaging cardiac tumors.[15] Furthermore, it is noninvasive, thus avoiding the risks of catheterization and possible embolization. In a recent review, the advent of echocardiography was cited as a major contributing factor to the increasing present day accuracy in the preoperative diagnosis of cardiac tumors.[9] Standard M-mode echocardiography is dependent on tumor movement for visualization. The classic imaging of a left atrial myxoma is a mass of reflected echos behind but separate from the anterior mitral leaflet that prolapses into the ventricle during diastole (Fig. 3). Using different beam projections, right-sided tumors have also been accurately visualized. Intracavitary tumors are more echogenic

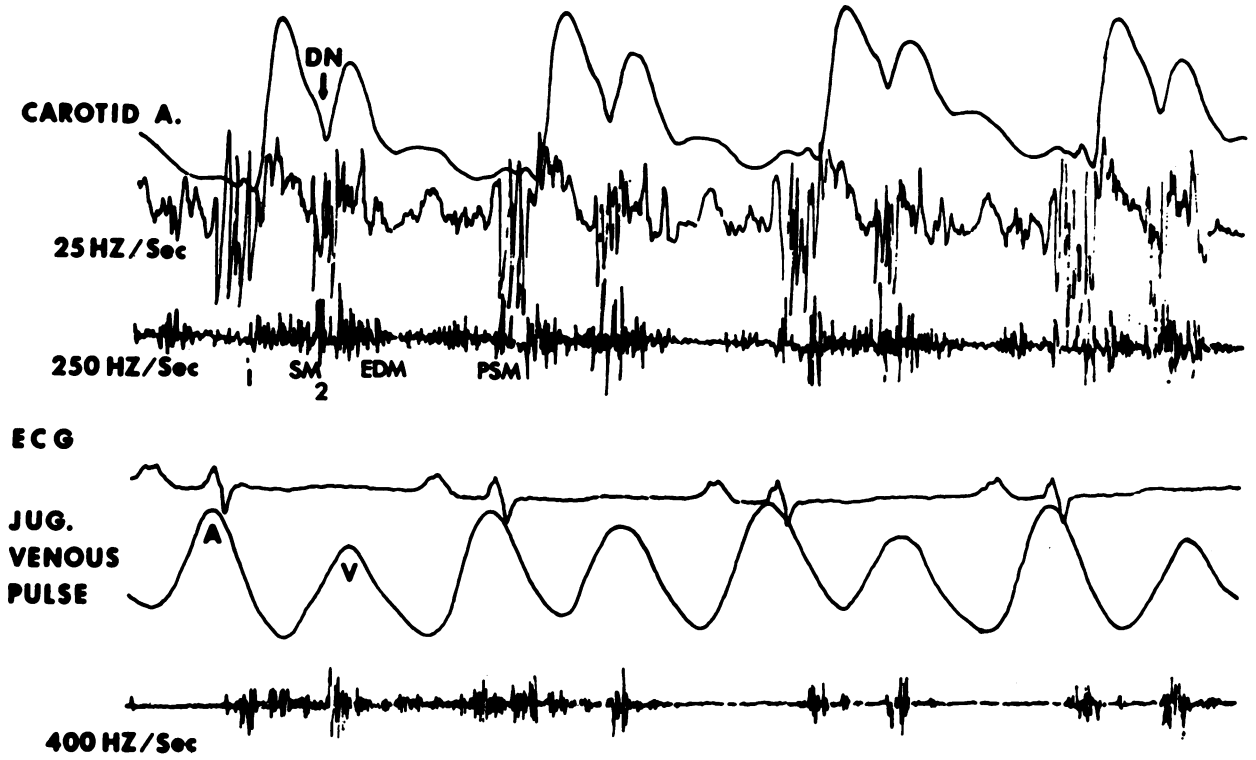
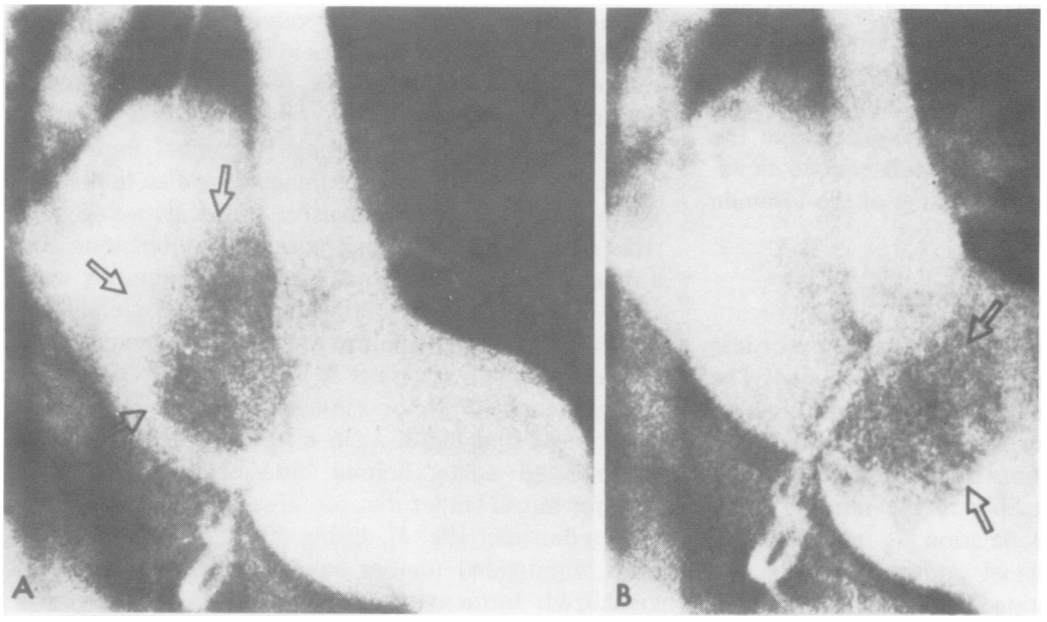


FIG. 1. Carotid artery pulse tracing and phonocardiograms of a patient with a left atrial myxoma. The vibrations of the first heart sound extend through the isovolumic contraction period. A varying systolic and diastolic murmur are also recorded. DN = dicrotic notch; EDM = early diastolic murmur. PSM = presystolic murmur; SM = systolic murmur (Reprinted with permission from Bower PJ, Ritter DG, Callahan JA, et al. Unusual hemodynamic findings of diagnostic value in a case of left atrial myxoma. Am J Cardiol 1969; 23:592.

than intramural tumors, however, ultrasound techniques have depicted an interventricular sarcoma that clinically and echocardiographically resembled asymmetrical septal hypertrophy.[31]

A more recent major advance has been the application of two-dimensional echocardiography for visualizing cardiac tumors (Fig. 4).[34] Two-dimensional echocardiography has many advantages over standard M-mode studies. The tumor size, shape, and mobility can be easily quantitated. Cardiac function can be



FIGS. 2A and B. (A) Myxoma is outlined in left atrium during systole. (B) During diastole, myxoma prolapses across mitral valve into left ventricle (Reprinted with permission from Selzer A, Sakai FJ, Popper RW. Protean manifestations of primary tumors of the heart. Am J Med 1972; 52:9).

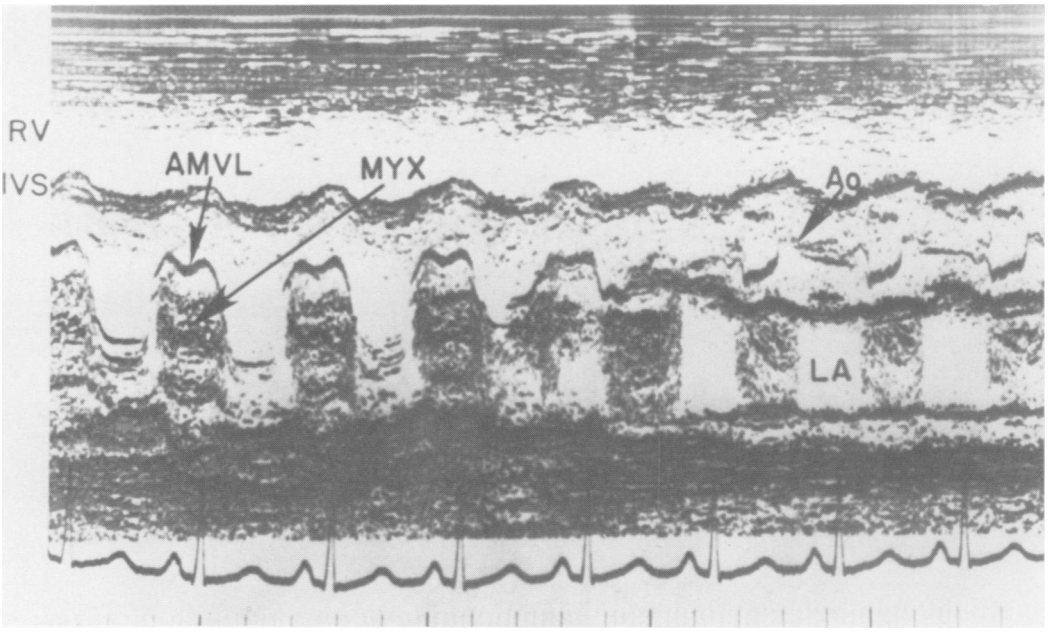


FIG. 3. M-mode echocardiogram demonstrating tumor prolapsing into left ventricle in diastole and appearing in left atrium during systole. AMVL = anterior mitral valve leaflet; MYX = myxoma; LA = left atrium; Ao = aorta; RV = right ventricle; IVS = ventricular septum (Reprinted with permission from Lappe DL, Bulkley BH, Weiss JL. Two-dimensional echocardiographic diagnosis of left atrial myxoma. Chest 1978; 74:55).

assessed. All chambers can be visualized simultaneously, and finally, there is better imaging of the right heart chambers. Recent evidence suggests that two-dimensional studies provide sufficient information to preempt the need, risk, and expense of preoperative cardiac catheterization in patients with cardiac tumors.[14]

Benign Tumors

Myxoma

These are the most common primary cardiac tumors and have been reported in patients ranging from three to 83 years of age, but are most frequently found in adult females.[27,64] Seventy-five per cent of myxomas originate in the left atrium, 20% occur in the right atrium, whereas ventricular tumors are rare. A familial incidence has been described, but the location of the tumor can vary among affected members.[12,43] Multiple chamber involvement can also occur, most often both atria being involved.[63] In rare instances, a myxoma has become secondarily infected.[26] This is an extremely lethal occurrence which can cause rupture of a cerebral mycotic aneurysm as the terminal event.

Grossly, myxomas are globular, or slightly lobulated, 4–8 cm in diameter, and have a soft, gelatinous consistency. These tumors are pale grey or yellow-brown, and on cut section often show areas of hemorrhage and necrosis (Fig. 5A). Myxomas are usually pedunculated with attachment to the endocardial surface near the margin of the fossa ovalis by a fibrovascular stalk. The length of the stalk determines the degree of tumor mobility during the cardiac cycle. Microscopically, polyhedral cells with a small, round nucleus occur in clusters separated by an afibrillar, eosinophilic myxomatous stroma. The predominant substance in the stroma is an acid mucopolysaccharide. Also present are other cellular elements, such as lymphocytes, histiocytes, plasma and mast cells (Fig. 5B). Of surgical significance is the finding that myxomas rarely extend deeper than the endocardium, but grow as polypoid tumors to fill the cardiac chamber.

There has been much debate over the origin of myxomas. One view has held that myxomas are not neoplasms, but represent only one of three possible entities into which an endocardial thrombus may evolve.[47] The other two are a mural thrombus and a Lambl's excrescence. However, most authors believe myxomas are neoplastic.[16,57] Supporting evidence for this concept include first, the location of myxomas. Most occur in the atria, whereas mural thrombi occur with equal frequency in the atria and ventricles. Also, when present in the atria, mural thrombi are usually found in the appendage whereas myxomas arise from the fossa ovalis. Secondly, myxomas lack lamination, contain little hemosiderin, are relatively acellular, are completely endothelialized, and contain acid mucopolysaccharides, unlike thrombi. Finally, some myxomas have associated constitutional symptoms, have metastatic potential, and their tumor emboli may invade blood vessel walls, unlike mural thrombi.

Clinically, left atrial myxoma may simulate many other diseases.[37] No physical finding is pathognomonic. The "tumor plop" is often indistinguishable from an opening snap. Although a varying murmur is considered diagnostic, this is seen infrequently. When the tumor obstructs the left heart and produces sys-

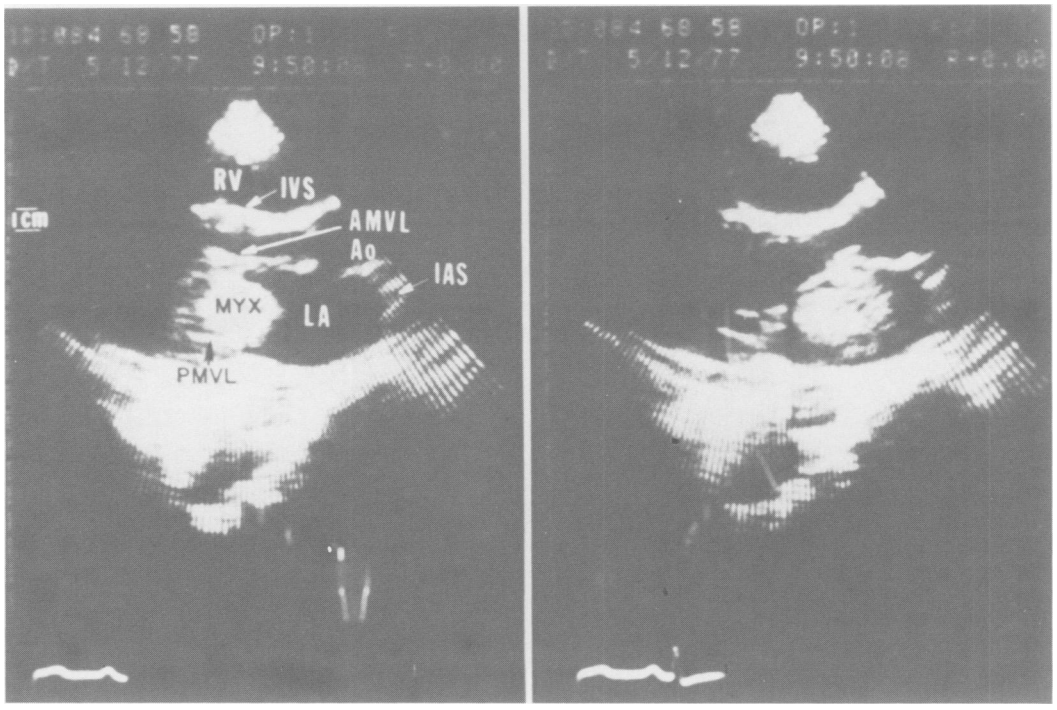


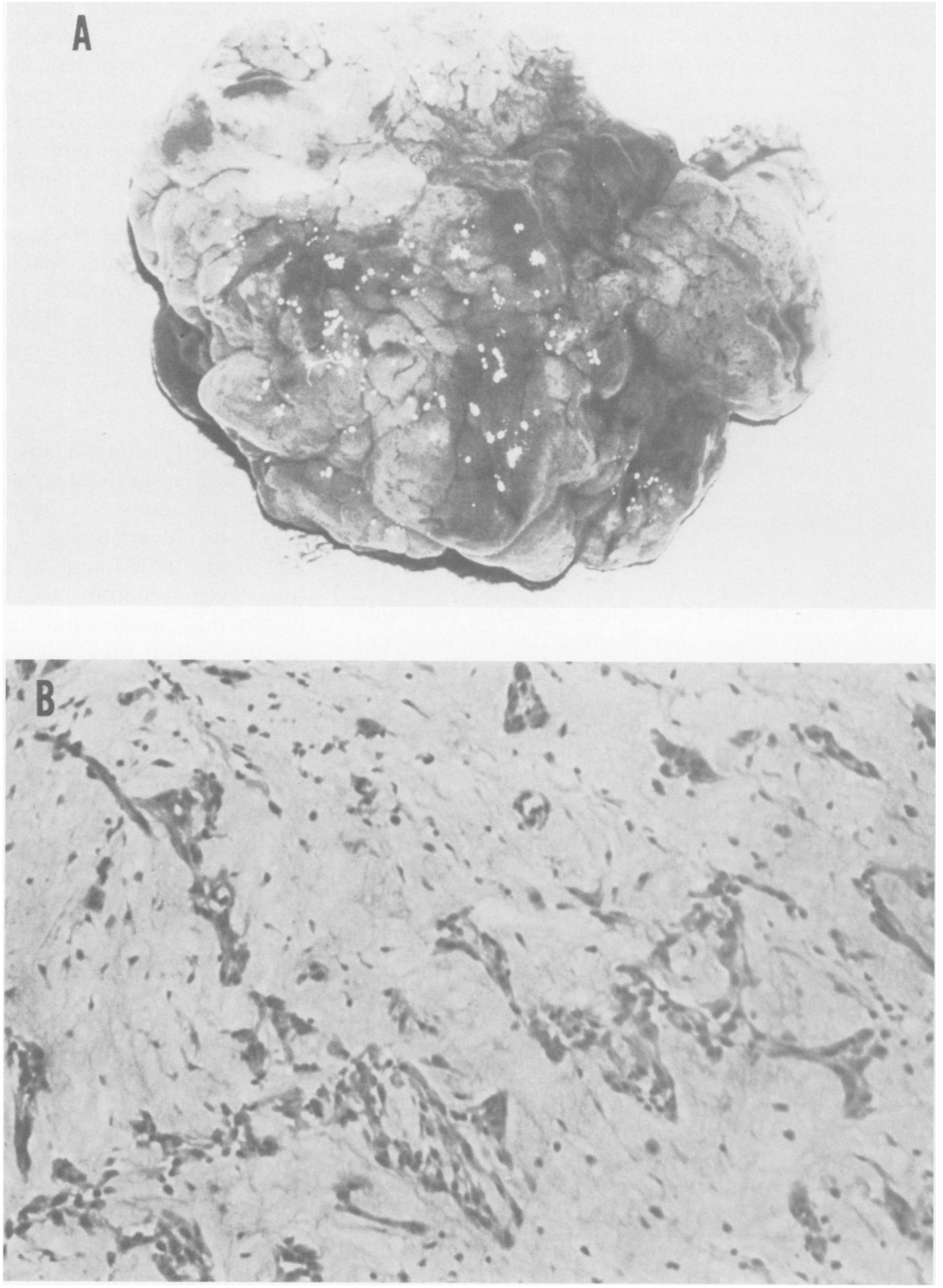
FIG. 4. Two-dimensional sagittal echocardiogram showing mobile tumor prolapsing into left ventricle during diastole (left) and returning to left atrium during systole (right). RV = right ventricle; IVS = interventricular septum; AMVL = anterior mitral valve leaflet; Ao = aorta; IAS = atrial septum; MYX = myxoma; PMVL = posterior mitral valve leaflet (Reprinted with permission from Lappe DL, Bulkley BH, Weiss JL. Two-dimensional echocardiographic diagnosis of left atrial myxoma. Chest 1978; 74:55.

temic embolization, mitral valve disease must be considered. Systemic embolization occurs in approximately 40% of patients and may be fragments of tumor or thrombus from the surface of the tumor. Left atrial myxomas frequently embolize to the brain, kidneys, aortic bifurcation and the lower extremities. Any material surgically extracted from the arterial system should be sent to the department of pathology as peripheral embolization is often the presenting manifestation of this tumor. When constitutional symptoms predominate, subacute bacterial endocarditis, collagen vascular disease and occult malignancy or infection enter the differential diagnosis.

The right atrium is the second most common location for myxoma.[60] These tumors produce signs and symptoms of right heart failure due to obstruction to vena caval inflow or to the tricuspid valve. Although occurring infrequently, pulmonary tumor emboli can produce chest pain, cough and dyspnea. On occasion, systemic constitutional symptoms can occur, as well as polycythemia and digital clubbing. The differential diagnosis includes rheumatic tricuspid valve disease, constrictive pericarditis, myocardiopathy, Epstein's anomaly, carcinoid heart disease, and other malignant or infectious causes of vena caval obstruction.

Ventricular myxomas are rare. Right ventricular myxomas occur in a younger age group, arise from the right ventricular outflow tract producing obstruction to ventricular ejection, and must be differentiated from pulmonary valvular and infundibular stenoses.[54] The most infrequent location for myxoma is in the left ventricle. Sixty per cent of patients have systemic emboli perhaps due to the vigorous contraction of the left ventricle. Unlike left atrial myxomas, left ventricular tumors rarely cause constitutional symptoms. These tumors are often attached to the interventricular septum along the left ventricular outflow tract. Systolic excursion of the tumor obstructs the aortic valve producing findings mimicking aortic stenosis, or idiopathic hypertrophic subaortic stenosis.[34]

Surgical resection is facilitated by utilizing cardioplegic arrest to provide a dry, motionless operative field to optimize complete excision and minimize perioperative tumor dislodgement and embolization. The extent of endocardial resection needed around the tumor stalk has been a controversial matter since intracardiac recurrences have been reported.[13,46] To obviate this, some authors have advocated a more extensive resection necessitating patch reconstruction of the atrial septum.[21,40] However, many believe an adequate resection encompasses a small "button" of normal endocardium around the tumor stalk and in these series recurrences are rare.[14,30] Recurrence of a myxoma raises the possibility of malignant degeneration. Although myxomas have been reported to metastasize to the brain, meninges, skin, bone, soft tissue and lung,[45,46] some authors believe these lesions are tumor emboli.[16] Others believe that these malignant tumors are not myxomas, but should be categorized as chondrosarcomas.[28]



FIGS. 5A and B (A) Typical lobulated, gelatinous gross appearance of a myxoma. (B) Histologically, polyhedral cells appear in clusters surrounded by an afibrillar stroma (original magnification ×150).

Lambl's Excrescence

This papillary tumor involves the aortic valve and adjacent endocardium in adults and more often the tricuspid valve in children. Histologically, it resembles a myxoma with a papillary lesion having a central core of fibrous connective tissue covered by endothelial cells contiguous with the endocardium of the valve cusp. This lesion is usually an asymptomatic incidental autopsy finding, but rarely can cause valve dysfunction.[7]

Rhabdomyoma

These hamartomas are the most common cardiac tumor of childhood.[17] Eighty-five per cent occur in

children less than 15 years of age, and one half of the children have other manifestations of tuberous sclerosis. These tumors have a poor prognosis, with a majority of the patients diagnosed during infancy not surviving until their first birthday. The tumors are usually multiple and involve the ventricles more often than the atria. On occasion, a lesion can enlarge to encroach upon the ventricular cavity and produce symptoms of ventricular outflow tract obstruction. More often they produce recurrent tachyarrhythmias. Histologically, the classic spider cells are seen as groups of dilated spaces containing a central nucleus with a narrow rim of cytoplasm and fine radiating fibrillar processes. Although rhabdomyomas are benign, their multiplicity, poor encapsulation and deep myocardial location make surgical resection difficult. However, there are reports of large solitary lesions producing symptoms that have been successfully removed.[53]

Fibroma

This rare tumor is usually a solitary, ventricular mass. Seventy per cent of these lesions have produced symptoms and all have been reported in children less than 10 years of age. Grossly, fibromas are firm, nonencapsulated and 3–7 cm in diameter. Microscopically, fibrous tissue of varying cellularity replaces cardiac muscle. Occasional calcification and foci of ossification can occur. Angiographic diagnosis has been made in the evaluation of children with symptoms of chest pain or congestive heart failure. These children are at a high risk for sudden death. Successful surgical resection of symptomatic tumors has been reported.[19]

Lipoma

Cardiac lipomas have been reported in patients ranging from neonates to 77 years of age.[50] Most patients are evaluated for cardiomegaly of unexplained etiology. One-half of these tumors originate from the subendocardium, the rest equally distributed in the myocardium and subepicardium. The left ventricle and right atrium are the most frequent locations. Histologically, these tumors resemble lipomas situated elsewhere in the body. Those that arise from the free left ventricular wall can be easily excised, but intramural atrial lesions require the use of cardiopulmonary bypass for resection. A lipoma arising from the mitral valve has been cured by valve excision and prosthetic valve replacement.[2]

Mesothelioma

This congenital tumor occurs most often in adult females. Mesothelioma has been called the smallest tumor that causes sudden death because of the frequent discovery at autopsy of this lesion in patients who rapidly succumbed with complete heart block. These tumors involve selectively the atrioventricular node and histologically are formed by nodules lined by cuboidal epithelium forming cysts, tubules and gland-like structures. Surgical resection has not been attempted as this would unavoidably produce heart block because of the tumor's location and thus necessitate permanent pacing. Electronic ventricular pacing in these patients has also produced sudden death from ventricular fibrillation. The explanation for this electrophysiological instability is unknown.[32]

Hemangioma

The most common vascular tumor arising from the heart is the hemangioma. These tumors occur mostly in adults and usually are small, clustered sessile or polypoid subendocardial excrescences that are incidentally found at autopsy. However, large tumors can infiltrate the myocardium and grossly distort the cardiac silouette. Extensive leisons often are surgically irresectable and unfortunately, radiation therapy has not been efficacious. Microscopically, the most common type of lesion is the cavernous hemangioma.[58]

Cysts

Blood cysts are usually asymptomatic and located on the heart valves.[7] They are 1 or 2 ml in size and most frequently involve the mitral or tricuspid valve. These cysts are comprised of a single blood-filled space lined by a layer of endothelial cells. Larger cystic lesions have been found on the mitral and pulmonic valves causing valve dysfunction necessitating surgical excision with prosthetic valve replacement.[35]

Malignant Tumors

Sarcoma

These are the most frequently encountered malignant primary heart tumors accounting for approximately 20% of all primary cardiac neoplasms.[48] They occur in adults and have no predilection for either sex. The right heart, particularly the right atrium, is the most frequent site of origin. Although arising from the endocardium or pericardium more often than from the myocardium, these highly malignant tumors rapidly infiltrate all layers of the heart, invade adjacent mediastinal structures and widely metastasize. Systemic metastases are present in 80% of cases when first diagnosed. The lungs and mediastinal lymph nodes are particularly prone to metastatic deposits.

This growth pattern makes cardiac sarcomas often irresectable and thus the prognosis is grave. Survival is measured in weeks or months.

A characteristic clinical presentation is progressive, unrelenting congestive heart failure, cardiomegaly, chest pain, fever, hemopericardium, arrhythmias, or sudden death. The rapid onset of superior or inferior vena caval obstruction is also common (Fig. 6). The precise histologic classification of sarcomas is difficult and often academic as the clinical picture and prognosis are similar. Angiosarcoma is the most commonly reported histologic variety. When diagnosed during life, palliative resections have been attempted, but with limited success. Irradiation and chemotherapy have also yielded only anecdotal partial remissions.[22,52] Rhabdomyosarcomas arise *de novo*, not from malignant degeneration of a rhabdomyoma. Both rhabdomyosarcomas and fibrosarcomas have been resected, but the palliative benefits may not justify the operative risk. Early recurrences have suggested that adjuvant therapies should be employed.[1] Extremely rare, but reported, are neurogenic sarcoma,[20] leiomyosarcoma,[33] osteosarcoma[36] and chondrosarcoma[62] arising as primary cardiac tumors.

Other Malignant Tumors

Although frequently the heart is involved by disseminated lymphoma, rare cases of lymphoma arising as a primary cardiac tumor have been noted.[55] In addition, primary plasmacytoma[59] and malignant mesenchymoma[7] of the heart have also been reported.

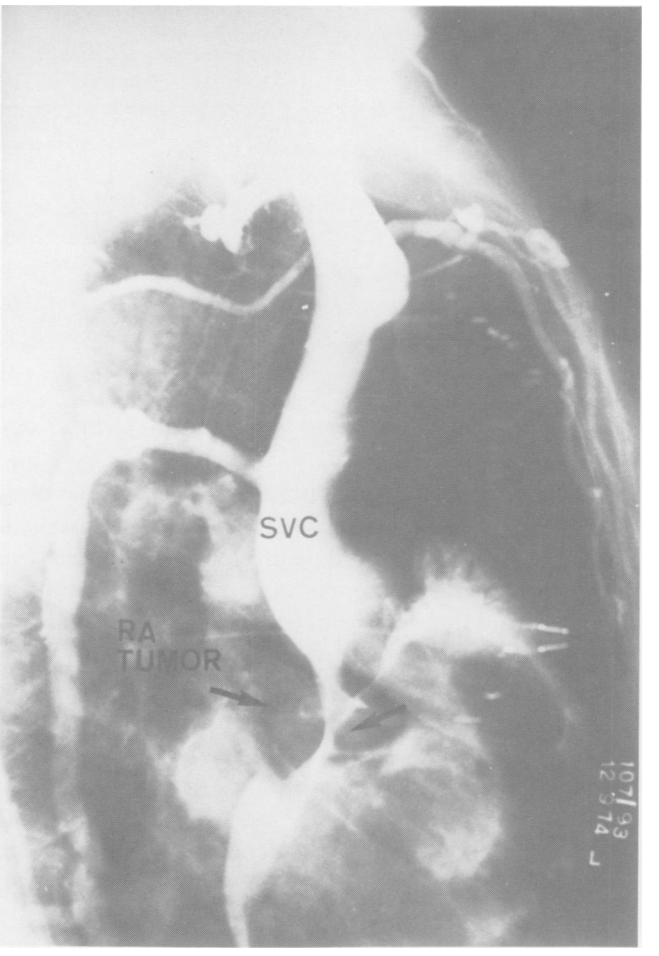


FIG. 6. Right atrial tumor obstructing cavo atrial junction with retrograde filling of azygos vein (Reprinted with permission from Torstveit JR, Bennett WA, Hinchcliffe WA, et al. Primary plasmacytoma of the atrium. J Thorac Cardiovasc Surg 1977; 74:563).

Surgical Experience at Duke University Medical Center

At Duke University Medical Center from 1966–1978, 16 patients with primary cardiac tumors and two patients with tumors metastatic to the heart have undergone surgical resection (Table 2). Fifteen of these tumors were myxomas. There was one patient with a primary lymphocytic lymphoma of the heart that after extensive staging was found to involve no other organ. Also treated were an osteosarcoma of the humerus that metastasized to the heart 19 years after a shoulder disarticulation and a hypernephroma that extended from the renal vein into the right atrium. Ten of the patients were females and 8 were male. They ranged in age from 30 to 71 years of age with an equal distribution in each decade.

Twelve myxomas were located in the left atrium and two were located in the right atrium. All except two of these myxomas arose from the foramen ovale. One tumor arose from the posterior left atrial wall and another was located in the right atrium between the coronary sinus and the tricuspid valve annulus. The left ventricular myxoma arose from the ventricular septum along the left ventricular outflow tract. The primary lymphoma obliterated the right ventricular cavity causing tricuspid and pulmonic valve obstruction. The osteosarcoma was metastatic to the free right ventricular wall and caused severe outflow tract obstruction. The hypernephroma lay free in the right

TABLE 2. *Cardiac Tumors Treated Surgically at Duke University Medical Center*

Type of Tumor	Number
Primary tumors	
myxoma	15
lymphoma	1
Metastatic tumors	
osteosarcoma	1
hypernephroma	1
	18

TABLE 3. *Location of Cardiac Tumors Treated at Duke University Medical Center*

Location	Number
Right atrium	
hypernephroma	1
myxoma	2
Left atrium	
myxoma	12
Right ventricle	
osteosarcoma	1
lymphoma	1
Left ventricle	
myxoma	1

atrium just cephalad to the inferior vena cava (Table 3).

This intracardiac renal tumor was the only asymptomatic leison. All other right heart tumors produced symptoms of congestive heart failure. Two patients with right atrial myxomas concomitantly manifested constitutional symptoms of fever, weight loss, anorexia and lassitude. Obstruction to left heart filling was the presenting symptom in six patients with left atrial myxomas. Two individuals had in addition to left heart failure manifestations of systemic emboli to the brain, kidneys, extremities, or coronary arteries. One patient with a left atrial myxoma was diagnosed by pathologic examination of a surgical embolectomy specimen extracted from an acutely occluded common femoral artery. Two left atrial myxomas manifested systemic complaints of arthralgias, fever and anemia in addition to left heart obstruction and one patient presented solely with constitutional complaints and no symptoms of cardiac dysfunction. The left ventricular myxoma obstructed the left ventricular outflow tract causing syncope, palpitations and angina pectoris. This woman had remote symptoms of transient ischemic attacks, presumed to be embolic in origin (Table 4).

Cardiac catheterization was performed in 13 patients and in each instance an intracavitary mass was demonstrated. There were no catheterization induced emboli. M-mode echocardiography was diagnostic of an intracardiac tumor in 14 of 15 patients. The one false negative occurred in a 56-year-old woman presenting with a clinical picture of constrictive pericarditis. The study was interpreted as showing a large pericardial effusion with paradoxical septal motion. However, subsequent cardiac catheterization and surgical resection confirmed the diagnosis of right atrial myxoma.

TABLE 4. *Manifestations of Cardiac Tumors Treated at Duke University Medical Center*

Manifestation	Number
Right atrium	
asymptomatic	1
O&C	2
Left atrium	
E	1
O	6
C	1
O&C	2
O&E	2
Right ventricle	
O	2
Left ventricle	
O&E	1

C: constitutional. O: obstruction. E: embolism.

Two-dimensional echocardiography in nine patients was 100% accurate in diagnosing the presence and location of an intracardiac mass lesion. Of particular interest are five patients (three with left atrial myxoma, one with a left ventricular myxoma and one with a right ventricular lymphoma) who had two-dimensional echocardiograms diagnostic for cardiac tumors, but in whom surgical resection was undertaken without prior angiography or catheterization.

All patients with atrial myxomas underwent tumor resection inclusive of a small margin of normal endocardium around the fibrovascular stalk. One patient with a large left atrial myxoma required in addition prosthetic mitral valve replacement because the tumor mass had destroyed the leaflets resulting in free mitral insufficiency. The left ventricular myxoma was resected through an apical left ventriculotomy. A right ventriculotomy was used to excise the primary lymphoma and metastatic osteosarcoma. The hypernephroma ws extracted from the right atrium through an atriotomy concomitant with a radical nephrectomy.

There was no operative mortality. Supraventricular arrhythmias occurred in 28% of patients, but all were discharged in sinus rhythm. Other complications were one renal artery embolus, one case of postoperative low cardiac output, and one minor wound infection. No primary tumor has recurred in follow-up ranging from eight months to 13 years. At present, echocardiography is being used to screen for early recurrence. However, both metastatic tumors recurred within three months of surgical resection, emphasizing the palliative nature of these procedures and the necessity for careful preoperative patient selection.

This series of patients emphasizes that the successful management of cardiac tumors begins with a high index of clinical suspicion as the signs and symptoms of these lesions are often perplexing. Echocardiography is a safe, reliable, and noninvasive diagnostic modality that may obviate the need for cardiac catheterization. The morbidity and mortality of surgical resection are minimal if strict attention is paid to avoidance of perioperative tumor embolization. And

finally, with atrial myxomas, excision of a small margin of normal tissue around the base of the tumor is a sufficient resection to avoid later recurrence and prevents more extensive procedures requiring atrial septal reconstruction.

Selected Annotated Bibliography

1. Bloor CM: Tumors of the endocardium and myocardium. *In* Bloor CM, (ed.) Cardiac Pathology. Philadelphia, J Lippincott Co., 1978, pp. 391–410. The chapter on cardiac neoplasms in this excellent text is a comprehensive review of the gross and histologic morphology of the most frequently encountered benign and malignant tumors. It is well illustrated and gives appropriate clinical correlations.
2. Donahoo JS, Weiss JL, Gardner TJ, et al. Current management of atrial myxoma with emphasis on a new diagnostic technique. Ann Surg 1979; 189–763. This review summarizes the excellent surgical results that can be obtained in treating cardiac tumors. Current operative techniques are clearly outlined. The role of two-dimensional echocardiography as a reliable diagnostic modality is emphasized.
3. Harvey WP: Clinical aspects of cardiac tumors. Am J Cardiol 1968; 21:328. This is one article in an excellent symposium on cardiac tumors. Emphasis is made on the signs and symptoms that should alert the clinician to the presence of a cardiac neoplasm. The illustrative cases and figures are particularly informative.
4. Selzer A, Sakai FJ, Popper RW: Protean clinical manifestations of primary tumors of the heart. Am J Med 1972; 52:9. This article is a detailed discussion of 13 cases of cardiac tumors treated by the authors. The pathophysiologic changes caused by these tumors is extensively reviewed. Emphasis is placed on earlier diagnosis as this is a curable form of serious heart disease.
5. Zitnik RS, Giuliani ER: Clinical recognition of atrial myxoma. Am Heart J 1970; 80:689. This is an excellent review of the most common primary cardiac tumor. The sections on laboratory diagnosis utilizing standard roentgenography, electrocardiography, phonocardiography and cardiac catheterization clearly outlines the advantages and limitations of each modality.

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