Unusual Aspects of Atrial Myxoma

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Cardiac myxomas are infrequently encountered lesions. Certain aspects of these tumors are unusual and include right atrial location, calcification, valve destruction, hematologic abnormalities, occurrence in children, and familial occurrence. A 12-year-old girl manifesting all of these unusual aspects of myxoma is described and each of the unusual aspects is discussed.

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THE HALLMARK OF ATRIAL MYXOMA is an enigmatic presentation of intermittent and variable cardiac signs frequently combined with vague systemic and constitiutional abnormalities. The less common right atrial myxoma is an especially elusive diagnosis, and may present as right heart failure in the absence of demonstrable pulmonary disease or apparent underlying cardiac pathology. Such lesions are rare in children, and familial occurrence is especially unusual. Calcification of myxomas occurs infrequently but is often associated with destructive lesions of the tricuspid valve. Hemolytic anemia and thrombocytopenia have rarely been noted to occur with calcified right atrial myxoma.

A 12-year-old girl was recently treated with advanced right heart failure who had a mobile, calcified, intracardiac mass noted at fluoroscopy. The child's mother had previously died from complications of a left atrial myxoma. Tricuspid valve destruction necessitated prosthetic replacement. Hemolytic anemia and thrombocytopenia resolved after removal of the mass. It is unique that most of the uncommon aspects of myxoma (childhood occurrence, right atrial location, calcification, familial incidence, hematologic abnormalities, and valve destruction) are combined in a single patient.

Case Report

D. S., a 12-year-old girl, was initially seen at the University of Mississippi Medical Center at age 8 because of suspected rheumatic heart disease. The patient's mother had died several years previously and a left atrial myxoma was found at autopsy. Physical examination demonstrated a grade II/VI systolic murmur, loudest at the lower left sternal border. No hepatomegaly or peripheral edema was noted. On return visit several months later, the patient described diminishing exercise tolerance and occasional peripheral edema. A grade IV/VI systolic murmur and a grade II/VI mid-diastolic murmur were present at the lower left sternal border. Electrocardiogram showed sinus rhythm, right atrial and right ventricular hypertrophy. Cardiomegaly was noted on chest x-ray. No laboratory abnormalities were noted. Cardiac catheterization in August, 1972, revealed essentially normal hemodynamic data. Right ventricular cineangiocardiography demonstrated an enlarged, poorly contracting right ventricle and a dilated tricuspid annulus with mild tricuspid insufficiency. "Right ventricular myopathy" was listed as the discharge diagnosis.

The patient was lost to follow-up until October, 1976, when she returned with a history of recent increased congestive heart failure manifested by dyspnea, chronic fatigue, peripheral edema, and severely limited activity. She had experienced intermittent chest pain, right upper quadrant discomfort and fever. On physical examination, she had icterus, ascites, and edematous lower extremities. Venous pulsations in the neck were prominent. Lung fields were clear. A grade IV/VI systolic murmur and thrill at the left lower sternal border were noted, as was a soft grade II/VI diastolic murmur. The liver edge was palpable at the level of the umbilicus. Chest x-ray demonstrated extreme cardiomegaly (Fig. 1a). Electrocardiogram showed sinus tachycardia with right atrial hypertrophy and right bundle branch block. Pertinent laboratory findings are outlined in Tables 1 and 2. Admission hemoglobin was 11.1 and peripheral blood smear revealed fragmented and distorted RBC's. Urine was positive for hemoglobin and bilirubin. Coagulation studies were normal except for mild thrombocytopenia. Coomb's test and blood cultures were negative. Treatment with digitalis and furosemide was started, but she failed to improve. A drop in hemoglobin to 8.4 occurred. Subsequent hematologic data are noted in Table 2. Echocardiography showed a mass of dense echos moving into the right ventricle in diastole. The right ventricular cavity was greatly enlarged, measuring 4.0 cm at the onset of QRS inscription in the plane of the posterior mitral leaflet. Fluoroscopy demonstrated a calcified mass moving through the tricuspid orifice from right atrium to right ventricle and back with each cardiac cycle (Figs. 2a and b). Cardiac catheterization demonstrated severe tricuspid insufficiency and angiograms again showed the mass moving freely within the right atrium and ventricle.

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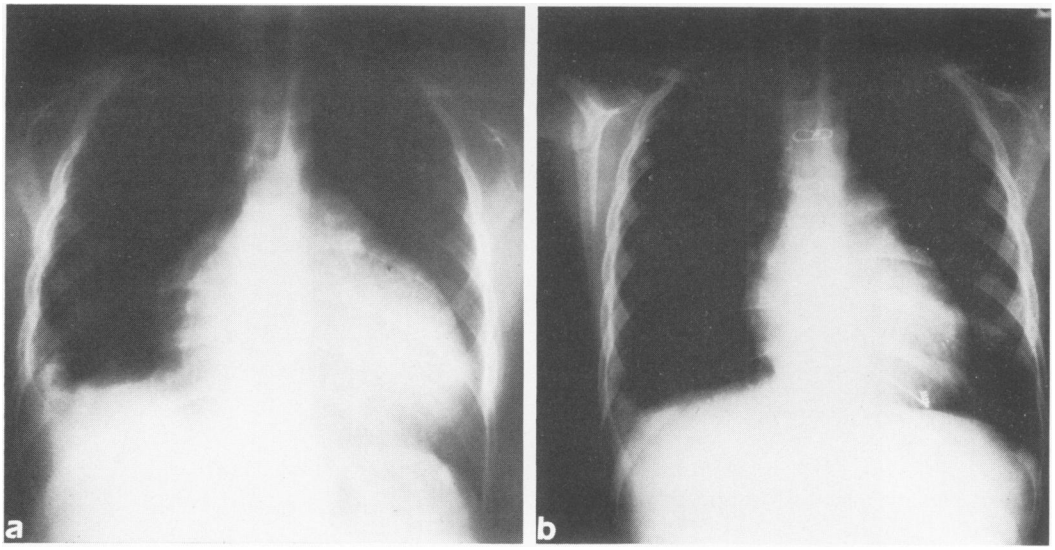
Submitted for publication December 27, 1977.

0003-4932-78-0800-0240-0075



FIGS. 1a and b. (a) Preoperative chest x-ray. (b) Postoperative chest x-ray.

At surgery on November 4, the right atrium and ventricle were enormously dilated. Cardiopulmonary bypass was established. Right atriotomy revealed the tumor lying free within the atrium with a portion projecting through the tricuspid valve. The tumor was spherical, approximately six centimeters in diameter, and had a hard cobbled surface. It had no cardiac attachments and could be lifted out of the heart intact (Fig. 3). The point of origin, consisting of a stalk measuring 1 cm in diameter and projecting into the right atrium, was identified just superior to the fossa ovalis. There was no connection to the tumor. This apparent origin of the tumor from the atrial septum was removed by excising the septum in this area. The left atrium and ventricle were inspected throughout this atrial septal defect and no tumor was apparent within these chambers. The atrial septal defect was closed with a patch of Dacron.® The tricuspid valve was found to be virtually destroyed, presumably by the to-and-fro motion of the calcified tumor through the orifice, and the valve annulus was markedly dilated. The remaining fragments of the tricuspid valve were excised and a 33 mm porcine heterograft was sutured into place. Immediately after defibrillation, atrioventricular dissociation was noted which then converted to a junctional rhythm. Because the patient had intermittent periods of third degree block after cessation of bypass, a permanent pacing electrode was placed onto the left ventricle. The patient's postoperative course was essentially uncomplicated. Hematologic values and liver function tests rapidly returned to normal (Tables 1 and 2). However, on several occasions, she demonstrated complete heart block and as a precautionary measure, the pacing electrode was connected to a permanent pacemaker on the tenth postoperative day. At the time of discharge, the patient was in sinus rhythm as she has been on all follow-up clinic visits. The child remains asymptomatic one year following surgery. Cardiac size has decreased remarkably on chest x-ray (Fig. 1b).

TABLE 1. *Laboratory Data*

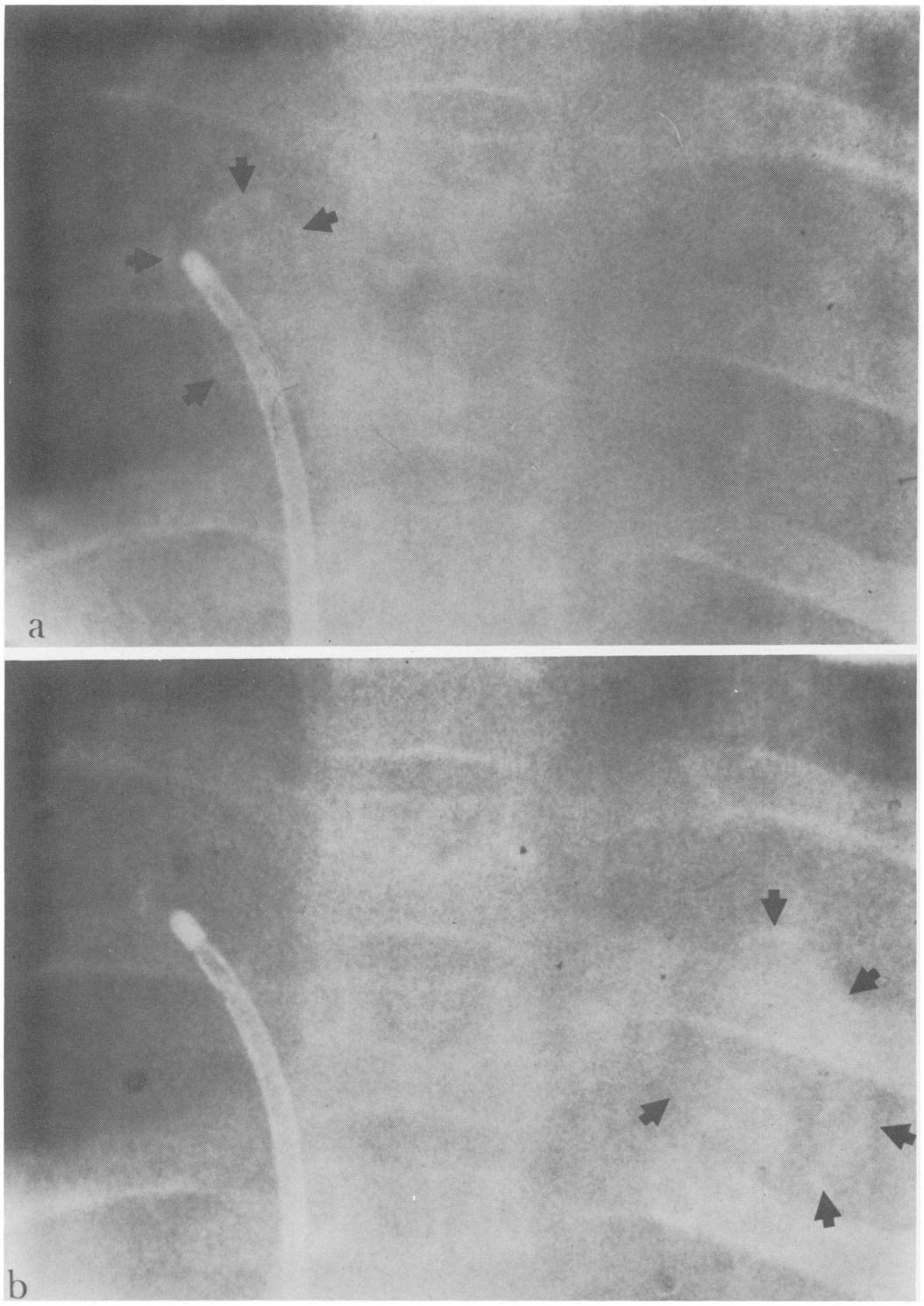
Date	Bilirubin (Total/ Direct)	SGOT	LDH	CPK	Alkaline Phosphatase
10/28 (Adm.)	6.2/2.5	92	170	2950	45
11/1 (Preop)	12.2/5.1	206	172	7040	—
11/14 (Postop)	1.4/1.0	29	130	550	25

Discussion

Cardiac myxomas have been found in all chambers of the heart but most frequently in the left atrium. Approximately one fourth occur in the right atrium.[6] Symptoms of myxoma are most frequently related to intermittent valvular obstruction, embolic episodes, or constitutional systemic abnormalities. The septal area of the fossa ovalis is the most common site of origin.[5] Endothelial or subendocardial reserve cell has been postulated as the origin. Syndromes suggestive of subacute bacterial endocarditis, rheumatic fever, or collagen vascular disorders may also occur. Pulmonary

TABLE 2. *Hematologic Data*

Date	Hgb/Hct	WBC	Reticulocytes	Platelets	Fibrin Degradation Products	RBC Morphology
10/29 (Adm.)	11.0/36	5,800	7.0%	150,000	1:5	Fragmented
11/1 (Preop)	8.4/32	10,000	11.4%	125,000	1:5	Marked fragmentation
11/5 (Postop)	13.4/41	15,000	10.8%	139,000	1:5	—
11/10 (Postop)	13.8/43	7,500	3.4%	365,000	Neg.	Essentially normal



FIGS. 2a and b. (a) Fluoroscopy in systole. Tip of catheter in right atrium (RA). Note the calcified mass (outlined by arrows completely within RA. (b) Fluoroscopy in diastole. Note that the calcified mass is now within the right ventricle. Catheter tip in same location as (2a).

tumor emboli are rare[15] and such myxomatous embolic fragments rarely grow into secondary deposits. Excision of the tumor and involved area of the atrial septum is usually curative.

Although right atrial myxoma may present in bizarre ways, almost all patients have right heart failure which may be unresponsive to conventional therapy. Fatigue, dyspnea on exertion, hepatomegaly, and peripheral

edema, are commonly present. A variable presystolic murmur may occur with tumor movement. Other murmurs are likely related to tricuspid valve abnormalities.[5,12,13] The course is progressive with unexplained obstruction of the venous circulation in the conspicuous absence of pulmonary disease or left heart failure. Right bundle branch block has been recorded in as many as one third of reported cases.[3,13,15] Cardiac catheterization is most likely to show elevation of right atrial pressures and a diastolic gradient across the tricuspid valve. A right to left atrial shunt may be noted in the presence of a patent foramen ovale or atrial septal defect.[5,14,16] Cineangiocardiogram demonstrating a mobile filling defect is the ultimate means for diagnosis.[11] However, failure to inject dye directly into the right atrium may result in the myxoma being missed as was probably the case with our patient at the first catheterization. Careful echocardiography has been shown to accurately diagnose the presence of both left and right atrial myxoma and should prevent the misdiagnosis of this lesion.

Cardiac myxoma in childhood is rare, and very few right atrial myxomas have been recorded in this age group. Sanyal noted only two right atrial myxomas recorded in children by 1967.[16] At least one has since been added to literature.[1] Interestingly, this most recently recorded childhood right atrial myxoma was also characterized by hemolytic anemia with iron depletion. The patient responded poorly to steroid therapy and poorly to conventional therapy of heart failure. Diagnostic findings were subsequently noted on cineangiography and the tumor was removed 16 months after the patient's first admission. The diagnosis of myxoma in children is typically delayed by the rarity of its occurrence in this setting, by the lack of specificity of signs and symptoms, and by the misleading constitutional signs.

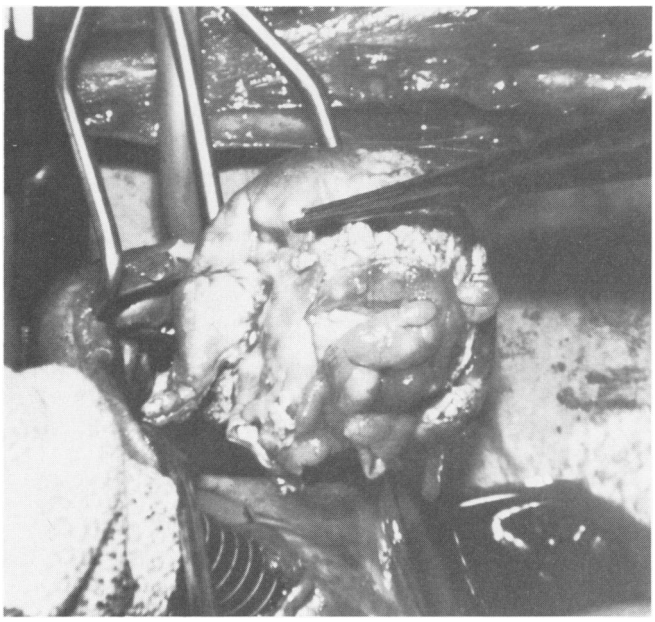


FIG. 3. Right atrial myxoma being lifted intact from right atrium. Note calcification just beneath forceps.

TABLE 3. *Reports of Familial Occurrence of Atrial Myxoma*

Author	Patients	Anatomic Location
1. Krause, 1971[10]	25 y/o male	Left atrium
	35 y/o brother	Right ventricle outflow tract
2. Kleid[9] and Heydorn, 1973[7]	14 y/o male	Left atrium
	16 y/o brother	Right atrium
3. Farah, 1975[2]	21 y/o male	Right atrium
	30 y/o sister	Left atrium
4. Siltanen, 1976[17]	mother	Biatrial
	12 y/o son	Left atrium
	16 y/o son	Right atrium
	18 y/o son	Left atrium
5. Liebler, 1976[11]	25 y/o male	Left atrium
	34 y/o brother	Left atrium, right atrium (2), right ventricle
	42 y/o brother	Left atrium, and right ventricle
	46 y/o brother	Left atrium and right ventricle
6. Crawford, 1978	mother	Left atrium
	12 y/o daughter	Right atrium

A familial tendency is noted in textbook descriptions of myxoma, but surprisingly few instances of such occurrence are well documented. Krause reported the first such occurrence in the English language literature in a report of two brothers.[10] Subsequent reports have added four families, two with several involved members (Table 3).[2,7,9–11,17] One interesting aspect is that in spite of the usual 3:1 prevalence of left atrial myxoma, each of these familial occurrences involves opposite atria in the affected members. Although several modes of genetic transmission have been postulated, none is well substantiated.

While calcified myxomas are rare, they do occur more often in the right atrium. They are infrequently identified on standard x-rays but may be with special short exposure films. They are readily seen at fluoroscopy and at the time of cardiac catheterization. Calcific debris has been noted within the pulmonary vascular tree in at least one case.[3] The duration of symptoms in patients with calcified tumors has been noted to be longer than in those with noncalcified tumors.[13]

Tricuspid insufficiency in association with right atrial myxoma has been reported in several patients and seems to occur most often with calcified tumors.[8] At least one instance of an unattached myxoma traveling

through the valve orifice is recorded prior to our experience.[3] In these patients, a holosystolic murmur of tricuspid insufficiency is frequently noted in addition to other variable murmurs typical of myxoma. Because pressure tracings in the superior vena cava, right atrium, and right ventricle may be similar when valve destruction is complete, Ebstein's anomaly has been suspected clinically (as occurred early in the evaluation of our patient). Attempts to do valvuloplasty or avoid valve replacement in instances of severe valve destruction by right atrial myxoma have resulted in clinically severe valvular insufficiency or even death following successful removal of the tumor.[12] In at least one patient, a second operation for valve replacement was necessary because of disability following removal of the calcified myxoma.[4]

Hemolytic anemia is apparently also more likely to occur with calcified myxoma, though anemia has been recorded in over one third of atrial myxomas.[18] Thrombocytopenia has also been noted in several cases and is readily reversed by removal of the tumor.[1,19] The stigmata of hemolytic anemia have been well documented with initial findings on peripheral blood smear of fragmented red blood cells, burr cells, and elevated reticulocytes. Total serum iron may be depleted by urinary excretion of free hemoglobin and siderocytes. Bone marrow erythropoiesis is intensive and megarcaryocytes are typically normal. A shortened life span of transfused labeled red blood cells has been documented.[18] The turbulent flow induced by the tumbling and projection of the myxoma into the atrium and tricuspid valve area is postulated as the etiology of these hematologic abnormalities although the calcium spicules and rough edges of the tumor probably contribute. Removal of the myxoma usually results in a complete return to normal of all such hematologic abnormalities as demonstrated by our patient.[1,20]

Cardiac myxomas are infrequently encountered lesions. Certain aspects of myxomas such as right atrial location, calcification, occurrence in childhood, familial occurrence, and hematologic abnormalities are particularly unusual. We have successfully treated a young girl with a calcified right atrial myxoma who particularly well illustrates most of these unusual aspects.

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