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Imaging Procedures in the Detection of Cardiac Tumors, with Emphasis on Echocardiography: A Review

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Abstract. Although cardiac tumors are relatively rare, their diagnosis is important because successful treatment is usually feasible if the diagnosis is made preoperatively. An analysis of 219 reports of cardiac tumors described in the English literature from 1972 through 1977 demonstrated the predominance of benign tumors, in particular myxoma, which is in agreement with past reviews. The methods of diagnosis employed included plain chest films, echocardiography, cardiac catheterization, angiocardiography, and cardiac scintigraphy. Conventional x-ray examination of the chest was abnormal in 83% of cardiac tumors but non-specific and should lead to further evaluation, first by echocardiography. Echocardiography, the most efficient diagnostic procedure for screening possible cardiac tumors, was abnormal in 94% of the cases. Cardiac catheterization was abnormal in 80% of cardiac tumors while definitive detection was made by angiocardiography in 94% of the cases. Cardiac scintigraphy has had limited use in the diagnosis of cardiac tumors, but has been diagnostic in 100% of the cases in a small series of myxomas.

Key words: Heart, neoplasms – Neoplasms, metastases – Heart, ultrasound studies – Heart, radionuclide studies – Angiocardiography – Cardiac catheterization.

Cardiac tumors, although rare, carry serious consequences that make their identification for operative removal vital. However, the majority of such tumors escaped medical diagnosis and surgical treatment until the past decade when the widespread availability of sophisticated diagnostic procedures, such as angiocardiography, echocardiography, and cardiac isotope scanning, as well as more refined surgical techniques, greatly enhanced the clinical management of this condition. While in the past, primary cardiac tumors were most often diagnosed at necropsy, most such tumors are now detected during life, permitting surgical treatment, which is generally curative. While the clinical, pathologic, and radiologic aspects of cardiac tumors have been the subjects of earlier reviews [1–14], no current body of data is available on the prevalence of various radiologic, hemodynamic, and echocardiographic abnormalities in patients with primary or secondary tumors of the heart. Accordingly, the purpose of this article is to present new information regarding the accuracy of diagnostic procedures in the detection of primary and secondary cardiac tumors and to delineate the frequency and prognosis of these tumors. Since echocardiography has proved to be a highly useful, noninvasive approach for identifying cardiac tumors, the ultrasound techniques are considered in greater detail than the other diagnostic methodologies. In addition to our own experience, the present study is based on all cardiac tumors reported in the English literature from 1972 through 1977, a period that saw the widespread application of newly developed diagnostic procedures. For a review of the literature prior to 1972, the reader is referred to the Review Articles [1–14].

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Cardiac Tumors

Incidence

Primary cardiac tumors are rare, as is indicated by their infrequent mention in the literature. Thus, no cardiac tumors were observed in one series of 30,000 autopsies, and only one case was reported in another series of 60,000 (0.002%) [14]; 44 such tumors were

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Table 1. Distribution of 219 primary cardiac tumors reported 1971–1977

Tumor	No.	
	Adult	Pediatric
Cardiac malignant	44	2
Pericardial malignant	5	1
Malignant	49	3
Cardiac benign myxoma	101	7
Cardiac benign non-myxoma	19	33
Pericardial benign	3	4
Benign	123	44
Total	172	47

Table 2. Adult malignant cardiac tumors reported 1971–1977

Tumor	No.
Right atrial angiosarcoma	14
Rhabdomyosarcoma evenly distributed in the four cardiac chambers	12
Left atrial malignant mesenchymoma	5
Right ventricular leiomyosarcoma	2
Fibrosarcoma	2
Fibromyosarcoma	1
Plasmacytoma	1
Reticular cell sarcoma	1
Malignant teratoma	1
Osteosarcoma	1
Myxosarcoma	1
Liposarcoma	1
Chondromyxosarcoma	1
Undifferentiated sarcoma	1

found in a general population of 160,000 (0.028%) [5]. A slightly higher frequency of nine in 11,000 (0.08%) was described in children at postmortem examination [11].

As noted before, most cardiac tumors had previously been diagnosed at autopsy [10–12], while currently they are generally detected during life. Thus, an analysis of the prevalence of these tumors, as encountered today by the cardiovascular radiologist and cardiologist, is more clinically meaningful. We have observed primary cardiac tumors in four of 7,000 procedures performed in the catheterization laboratory at the University of California Davis Medical Center during the past six years. The English literature contains reports of 219 primary cardiac tumors in the six-year period of 1972–1977, most being single case reports (see references); of these, more than 75% had been diagnosed during life, with 60% survival after treatment.

The prevalence of specific types of cardiac tumors, benign or malignant, and endocardial, myocardial, or pericardial in adults and children is also difficult to determine because of the sparsity of reported data. In the 219 cases from the literature, the distribution of cardiac tumors in adults and children according to location in the myocardium or pericardium and their differentiation as benign or malignant tumors are shown in Table 1. Of the 219 primary cardiac tumors, 47 occurred in children, which suggests that cardiac tumors are equally common in children and adults, considering the age distribution of the population studied. The ratio of malignant to benign primary tumors is greater in pericardial tumors (6:7) than in myocardial tumors (1:3.5), an observation in agreement with earlier autopsy findings [1]. Further, both benign and malignant pericardial tumors are considerably less frequent than myocardial tumors in adults as well as in children.

Malignant Cardiac Tumors in Adults

Of the 44 reported cases [15–50], there were 14 angiosarcomas, all of which arose in the right atrium; 12 rhabdomyosarcomas, which were essentially evenly distributed in the four chambers of the heart; and five malignant mesenchymomas, all found in the left atrium (Table 2). The most common malignant cardiac tumors in adults were right atrial angiosarcomas and rhabdomyosarcomas arising in any cardiac chamber, followed by left atrial malignant mesenchymomas. There have been occasional reports of angiosarcomas originating in the pericardium or left atrium [6]. The remaining 13 malignant tumors are listed in Table 2.

Benign Cardiac Tumors in Adults

As documented in Table 1, the predominant benign cardiac tumor was the myxoma, representing 84% of all primary tumors. Left atrial myxomas were by far the most common (68%), followed by those in the right atrium (15%), right ventricle (8%), and left ventricle (3%) (Table 3). Six percent of myxomas were multiple.

Benign Cardiac Tumors in Adults Excluding Myxomas

Nineteen such tumors were reported [98–115] (Table 4), eight of which were endomesotheliomas of

Table 3. Cardiac chamber distribution of 101 reported myxomas [58–97]

Chamber	No.
Left atrium	69
Right atrium	15
Right ventricle	8
Left ventricle	3
Right atrium and left atrium	3
Left atrium and left ventricle	1
Right atrium and pulmonary artery	1
Right atrium, right ventricle, and left atrium	1

Table 4. Adult benign cardiac tumors, excluding myxomas

Tumor	No.
Endomesothelioma of atrioventricular node	8
Fibroma	3
Lipoma	3
Hemangioma	2
Pheochromocytoma	1
Papilloma	1
Neurilemmoma	1

Table 5. Benign Cardiac Tumors in Children

Tumor	No.
Rhabdomyoma	23
Fibroma	7
Myxoma	7
Teratoma	4
Hamartoma	1
Schwannoma	1
Neurofibroma	1

the atrioventricular (AV) node [101]. It appears that the AV node mesothelioma is the second most common benign cardiac tumor in adults. This particular tumor is quite small, only a few millimeters in diameter, and causes death by intractable arrhythmias; because of its small size it has never been diagnosed in life. Other tumors found were three left ventricular (LV) fibromas, two of which were calcified; three lipomas, two in the AV groove and one in the right atrium (RA); two hemangiomas in the RA and LV; a left atrial (LA) pheochromocytoma; a papilloma of the aortic valve; and a right atrial neurilemmoma (Table 4).

Benign Pericardial Tumors in Adults

Only three cases in this group were reported from 1972 to 1977: a lipoma, a teratoma, and a diffuse angiomatosis [113–115]. Other tumors, such as hamartoma, leiomyoma, and dermoid, were described in the earlier literature. Thyroid, thymic, parathyroid rest tumors, bronchogenic cysts, and inclusion cysts occur in the pericardium but are not true pericardial tumors, nor are pericardial cysts and diverticula.

Malignant Cardiac Tumors in Children

Although cardiac tumors are approximately as common in children as in adults, malignant tumors are quite rare in children. In the literature reviewed, only three such tumors were found, two cardiac (one left ventricular rhabdomyosarcoma [133], and one right atrial Kaposi sarcoma [129]) and one pericardial (a pericardial fibrosarcoma [119]).

Benign Tumors in Children

The most common cardiac tumor in children is the rhabdomyoma, followed by fibroma and myxoma (Table 5). There were four benign pericardial tumors, all of which were teratomas, one hamartoma, one schwannoma, and one neurofibroma of Vagus. The rhabdomyomas were most commonly multiple (10 cases). In cases in which there was a single lesion, six were in the left ventricle, three in the right ventricle, two in the left ventricle extending into the left atrium, one in the left atrium, and one in the right atrium. Myxomas, which are by far the most common adult cardiac tumors, are substantially less common in children.

Tumor Extension

Tumor extension into the heart from an extracardiac tumor was reported in 11 cases [151–158], seven of which occurred in the right atrium and four in pulmonic veins and the left atrium. Symptoms of tumor extension into the heart may be the first manifestation of a primary extracardiac tumor. Of the seven RA tumors extensions there were three cases of Wilms' tumor, two of renal cell carcinoma, one hepatoma, and one retroperitoneal osteosarcoma. The four tumors extending into pulmonic veins and left atrium comprised two cases of squamous cell lung carcinoma, one oat cell carcinoma, and one pleural mesothelioma.

Metastatic Tumors

Metastatic tumors of the heart and pericardium are approximately 20 to 40 times more common than primary cardiac tumors [12]. Metastatic cardiac tumors have been reported in 1–5% of all autopsies

[1, 8] and in 10–25% of patients dying from malignant disease [2]. Pericardial metastases are approximately twice as common as myocardial [2]. The most common primary tumors metastasizing to the heart or pericardium are lung carcinoma, carcinoma of the breast, lymphoma-leukemia, and malignant melanoma [2, 8, 12, 171, 174], but any malignant tumor may metastasize to the heart or pericardium. Up to 60% of all melanomas and 25–50% of all bronchial carcinomas [2, 14] have been reported to metastasize to the heart or pericardium. Metastases are unusual in children, reflecting the rarity of pediatric malignancies. Pratt [179] reported that eight of 23 children with peripheral rhabdomyosarcoma had cardiac metastases, a frequency similar to that in adults.

Cardiac metastases are so common that most are no longer reported when encountered by clinicians or pathologists. The literature review of the recent six-year period is, therefore, under-representative of the true frequency of cardiac metastases. Twenty-five references have been found reporting 33 cases of malignant tumor with cardiac metastases, as well as eight review articles discussing numbers of cases [159–189]. The most commonly reported malignant tumors in these recent references are again lymphosarcoma-leukemia, lung carcinoma, melanoma, and breast carcinoma, with single case reports of metastases from tumors in various other locations, such as the cervix, trachea, and kidney.

Methods of Diagnosis and Prognosis of the Cardiac Tumors

When a cardiac tumor is suspected on the basis of clinical history and physical examination, the following methods are generally used to confirm or exclude the diagnosis: (1) x-ray examination of the chest, sometimes including fluoroscopy and tomography; (2) echocardiography; (3) cardiac catheterization; (4) angiocardiography; and, sometimes, (5) cardiac scintigraphy. The use of these methods and their results in the 263 cardiac tumors, metastases, and tumor extensions reported in the English literature from 1972 to 1977, are given in Table 6. After the diagnosis was confirmed, exploratory thoracotomy and tumor resection were generally performed. At the present time, the diagnosis is only occasionally made at exploratory thoracotomy, without preoperative detection of benign cardiac tumors; such is also the case in at least 50% of malignant cardiac tumors, both primary and secondary (Table 6).

The survival rate was 51% for all cardiac tumors including secondary tumors, 60% for primary benign and malignant tumors, and 74% for benign tumors. The prognosis for survival with malignant primary cardiac tumors was poor (15%) and was even worse for secondary tumors (7%). The follow-up period in the series studied was generally several years (range, 6 months to 10 years).

A further breakdown of the material into adult malignant versus benign tumors, myxomas, tumors in children, secondary tumors, and tumor extension, and other categories is depicted in Tables 7–10. Myxomas have the best prognosis (87% survival), better than other benign tumors in adults (47% survival), and considerably better than malignant cardiac tumors in adults (14% survival). Cardiac tumors in children, which carry a 55% prognosis for survival, are almost all benign. The discrepancy between the prognosis in benign tumors in children (57% survival) compared to in adults (80% survival), is likely due to the relative rarity of myxomas in children. The survival rate in adult benign cardiac tumors, except myxomas, is similar to that in children. Malignant tumors, either primary or secondary, have the worst prognosis, but occasionally patients with cardiac metastases can survive for several months after diagnosis and treatment, as illustrated by a case of cervical carcinoma with pericardial metastases reported by Charles et al. [162], which had a follow-up period of 1.5 years.

The prognosis for survival is greatly enhanced when the cardiac tumor is detected preoperatively. In the 134 surviving patients, the diagnosis was made preoperatively in 128, and only six were diagnosed exclusively at explorative thoracotomy. Of the 129 non-survivors, 78 died without preoperative or preautopsy diagnosis, and 51 died with such diagnosis. Of the 83 patients without a preoperative or preautopsy diagnosis, 77 died and only six survived. Of the 43 patients with benign cardiac tumors, 24 of the non-survivors were without preoperative diagnosis while 19 died with a known diagnosis. A few instances of mortality in benign cardiac tumors were attributed to preoperative demise [126]; two children died at cardiac catheterization [117, 138], and two children died of complicating congenital heart disease [124, 146]. Although reported within the six-year period surveyed, an occasional patient had died several years before the availability of open heart surgery. Of the 13 non-surviving cases of myxoma, for example, four were from the pre-open heart surgery era [62, 70, 73, 76], three had no or an incorrect preoperative diagnosis [66, 68, 73], and six died during or soon after surgery [62, 67, 88].

Diagnostic Methods Employed

X-ray Examination of the Chest

This demonstrated pathology in 83% of all cases of cardiac tumor in which the examination had been

Table 6. Accuracy of diagnostic methods in cardiac tumors

	No.	Chest x-ray			Echocardiography			Cardiac catheterization			Angiocardiography			Isotope study			Exploratory thoracotomy			Dead	Alive	Percent survival
		+	−	Not done or stated	+	−	Not done	+	−	Not done or stated	+	−	Not done or stated	+	−	Not done	+	−	Not done			
All tumors, metastases, and tumor extensions	263	175 83%	35 17%	53	69 94%	4 6%	190	112 80%	28 20%	123	158 94%	10 6%	95	14 88%	2 12%	247	179 98%	3 2%	81	129	134	51
All true cardiac tumors	219	151 87%	22 13%	46	63 97%	2 3%	154	93 77%	28 23%	98	144 94%	10 6%	65	13 87%	2 13%	204	168 98%	3 2%	48	88	131	60
All benign cardiac tumors	167	111 87%	17 13%	39	60 100%	0	107	80 76%	25 24%	62	124 95%	7 5%	36	11 100%	0	156	142 100%	0	25	44	123	74
All malignant primary cardiac tumors	52	40 89%	5 11%	7	3 60%	2 40%	47	13 81%	3 19%	36	20 87%	3 13%	29	2 50%	2 50%	48	26 90%	3 10%	23	44	8	15
Metastases and tumor extensions	4	24 65%	13 35%	7	7 88%	1 12%	36	19 100%	0	25	14 100%	0	30	1 100%	0	43	11 100%	0	33	41	3	7

Table 7. Accuracy of diagnostic methods in benign adult cardiac tumors

	No.	Chest x-ray			Echocardiography			Cardiac catheterization			Angiocardiography			Isotope study			Exploratory thoracotomy			Dead	Alive	Percent survival
		+	−	Not done or stated	+	−	Not done	+	−	Not done or stated	+	−	Not done or stated	+	−	Not done	+	−	Not done			
Benign cardiac tumors, except myxomas	19	10 91%	1 9%	8	1 100%	0	18	0	7 100%	12	7 78%	2 22%	10	1 100%	0	18	9 100%	0	10	10	9	47
Benign peri- or epicardial tumors	3	3 100%	0	0	1 100%	0	2	1 50%	1 50%	1	1 50%	1 50%	1	0	0	3	2 100%	0	1	1	2	67
Myxoma	101	60 83%	12 17%	29	46 100%	0	55	58 91%	6 9%	37	82 96%	3 4%	16	10 100%	0	91	99 100%	0	2	14	87	86
Total	123	73 85%	13 15%	37	48 100%	0	75	59 81%	14 19%	50	90 94%	6 6%	27	11 100%	0	112	110 100%	0	13	25	98	80

Table 8. Accuracy of diagnostic methods in adult malignant cardiac tumors

	No.	Chest x-ray			Echocardiography			Cardiac catheterization			Angiocardiography			Isotope study			Exploratory thoracotomy			Dead	Alive	Percent survival
		+	−	Not done or stated	+	−	Not done	+	−	Not done or stated	+	−	Not done or stated	+	−	Not done	+	−	Not done			
Malignant cardiac tumors	44	33 89%	4 11%	7	2 40%	3 60%	39	9 75%	3 25%	32	15 83%	3 17%	26	1 33%	2 67%	41	21 91%	2 9%	21	38	6	14
Malignant pericardial tumors	5	5 100%	0	0	0	0	5	3 100%	0	2	4 100%	0	1	0	0	5	3 75%	1 25%	1	4	1	20
Total	49	38 90%	4 10%	7	2 40%	3 60%	44	12 80%	3 20%	34	19 86%	3 14%	27	1 33%	2 67%	46	24 89%	3 11%	22	42	7	14

Table 9. Accuracy of diagnostic methods in pediatric cardiac tumors

	No.	Chest x-ray			Echocardiography			Cardiac catheterization			Angiocardiography			Isotope study			Exploratory thoracotomy			Dead	Alive	Percent survival
		+	−	Not done or stated	+	−	Not done	+	−	Not done or stated	+	−	Not done or stated	+	−	Not done	+	−	Not done			
Benign cardiac tumors	44	38 90%	4 10%	2	12 100%	0	32	21 66%	11 34%	12	34 97%	1 3%	9	0	0	44	32 100%	0	12	19	25	57
Malignant cardiac tumors	3	2 67%	1 33%	0	1 100%	0	2	1 100%	0	2	1 100%	0	2	1 100%	0	2	2 100%	0	1	2	1	33
Total	47	40 89%	5 11%	2	13 100%	0	34	22 67%	11 33%	14	35 97%	1 3%	11	1 100%	0	46	34 100%	0	13	21	26	55

Table 10. Accuracy of diagnostic methods in cardiac metastases and tumor extensions

	No.	Chest x-ray			Echocardiography			Cardiac catheterization			Angiocardiography			Isotope study			Exploratory thoracotomy			Dead	Alive	Percent survival
		+	−	Not done or stated	+	−	Not done	+	−	Not done or stated	+	−	Not done or stated	+	−	Not done	+	−	Not done			
Cardiac metastases	33	22 73%	8 27%	3	5 100%	0	28	16 100%	0	17	7 100%	0	26	0	0	33	7 100%	0	26	32	1	3
Tumor extension into the heart	11	2 29%	5 71%	4	2 67%	1 33%	8	3 100%	0	8	7 100%	0	4	1 100%	0	10	4 100%	0	7	9	2	18
Total	44	24 65%	13 35%	7	7 88%	1 12%	36	19 100%	0	25	14 100%	0	30	1 100%	0	43	11 100%	0	33	41	3	7

performed or reported (175 of 210 examined cases, Table 6). In the majority of patients, the pathologic findings were inconclusive (e.g., cardiomegaly), but occasionally the specific lesion, such as a calcified tumor, could be observed by plain x-ray examination of the chest. In metastases to the heart or tumor extension into the heart, x-ray examination of the chest was somewhat less frequently pathologic (65%) than in primary cardiac tumors (87%).

Echocardiography

Echocardiography has evolved as a highly accurate, noninvasive diagnostic tool in the detection of cardiac tumors. It was used in only 73 of the 263 reported cases [190–232], but its use has increased in all cardiac diagnostic centers and, at present, it is employed in all cases of suspected cardiac tumor. Echocardiography showed positive findings, in most cases diagnostic of a cardiac tumor, in 94% of the cases in which it was used. It was negative in only four cases (Tables 6–10): three malignant tumors and one tumor extension into the heart. In two of these negative cases, echocardiography showed pericardial effusion but did not demonstrate tumor [194, 299]; in the third case technical echocardiographic difficulties were encountered [208].

Cardiac Catheterization

This examination was positive in 80% and negative in 20% of the cases in which it was performed (Table 6), but it was done or reported in only approximately 50% of cases. Hemodynamic findings were not diagnostic, but suggested cardiac obstruction, which was an indication for further evaluation with angiocardiography. With the exception of myxomas, cardiac catheterization was negative in adult benign cardiac tumors (Table 7). These seven negative cases were intramural tumors, usually fibromas [99, 108, 110] with an occasional angioma [107], neurilemmoma [102], pheochromocytoma [102], and endothelioma [104]. Cardiac catheterization also frequently had negative findings in benign cardiac tumors in children. Almost all these tumors were also intramural (usually fibromas or rhabdomyomas) and did not cause obstruction to blood flow [120, 127, 137, 143, 144, 146, 147] (Table 9). Occasionally myxomas in the RA or LA had negative catheterization findings when they did not prolapse into the AV valves [62, 76, 81, 118]. Some malignant intramural tumors, such as leiomyosarcomas, on occasion also had normal catheterization findings [21]. In summary, cardiac catheterization is most often positive and initiates further studies with angiocardiography, but may be negative in 20% of cases, almost all intramural tumors.

Radionuclides

Nuclear medicine procedures have occasionally been used in the diagnosis of cardiac tumors (Table 6). Of the 16 reported cases in which radionuclides were used [233–239], 10 were myxomas, all of which were diagnosed by gated cardiac blood pool scintigraphy, which showed a mobile defect in the left or right atrium [233, 237–239]. Harford et al. [235] diagnosed a metastasis to the left ventricular myocardium from a carcinoma of the lung by finding concentrations of technetium-99m stannous pyrophosphate in the metastasis. Radionuclide studies were applied in four malignant tumors and were diagnostic in a pericardial fibrosarcoma in a child [234] and in an adult pulmonic valve sarcoma [36], but did not detect a diffuse rhabdomyosarcoma [19] or a right atrial angiosarcoma [29] in two adults. A benign right atrial hemangioma [111] was also detected by radioisotopes. In summary, radionuclide studies have proved quite useful in the diagnosis of myxomas, but in other cardiac tumors, present experience is limited.

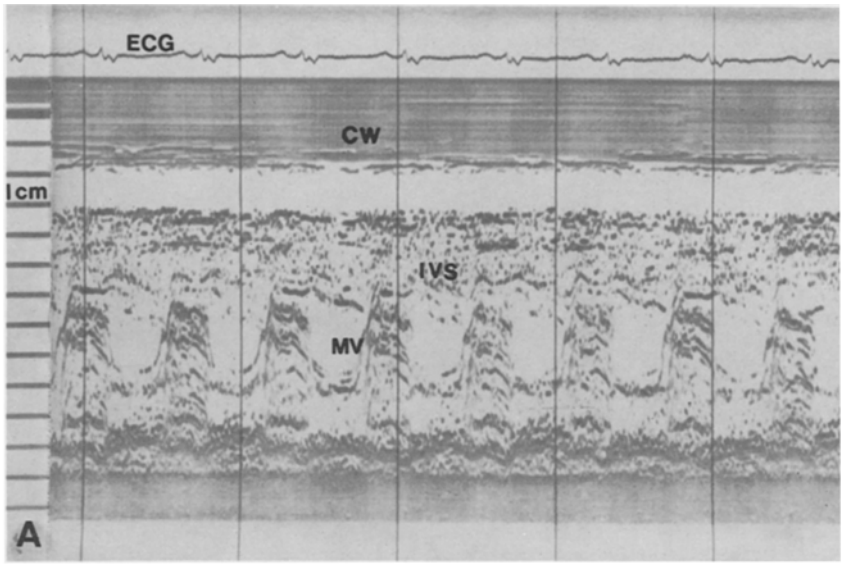
Angiocardiography

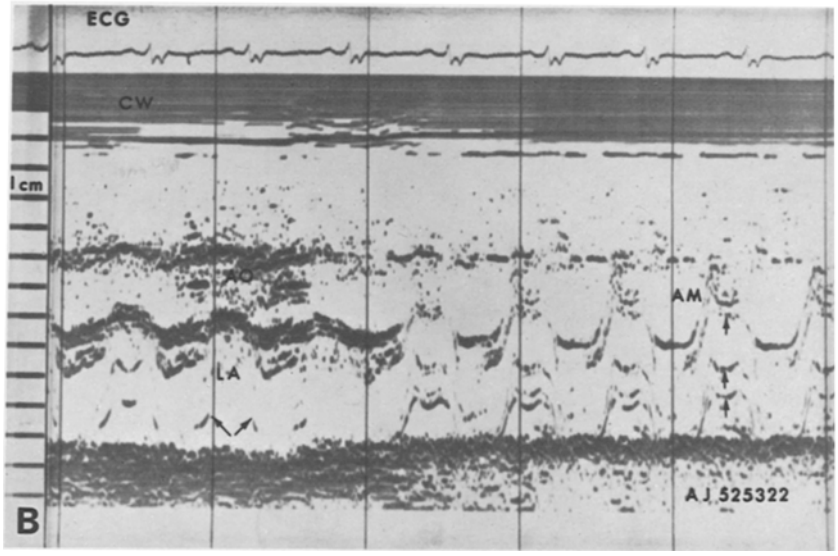
Angiocardiographic findings were positive in 94% and negative in 6% of all cases in which the procedure was performed or reported (168 of 263 cases) as shown in Table 6. In two cases, a benign right atrial tumor not seen at angiography was found at surgery performed for congenital heart disease [102, 146]. In four patients in whom cardiac tumors were subsequently found at surgery or autopsy, the angiographic diagnosis was pericardial effusion [21, 28, 45, 114]. The four remaining negative cases were one small AV node endothelioma [104] and three cases of myxoma. Two left atrial myxomas were not seen by angiography due to poor angiographic quality [76]. Finally, biatrial myxomas were not detected at angiocardiography in one case [84]. In summary, angiocardiography, if properly performed, is almost always diagnostic in cardiac tumors, with the exception of small intramural tumors. In cases with other cardiac lesions, such as congenital heart disease or pericardial effusion, a coexisting cardiac tumor may not be observed.

Considerations in the Diagnosis of Cardiac Tumors

Plain Film Diagnosis

The plain film diagnosis of cardiac tumors has been described in detail in earlier review articles [1, 3, 13].





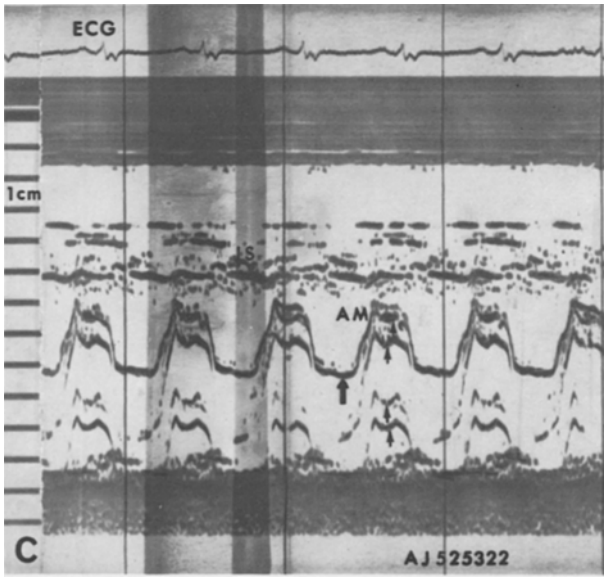


Fig. 1A–C. Echocardiography in left atrial myxoma. **A** Typical echocardiogram of left atrial myxoma showing a dense conglomeration of echoes behind the mitral valve in diastole, indicating a left atrial myxoma. This tracing could only be obtained with a single critical transducer position and angle. **B and C** Echocardiography in the same patient with routine transducer position and angle. **B** Echocardiography through the left atrium (LA) transversing the plane or aorta (AO) (left) and mitral valve. Discrete linear echos are seen in the LA posterior to the aorta and behind the anterior mitral valve leaflet (AM) (*arrows*). **C** An echocardiogram focused on the mitral valve shows multiple linear echoes (*small arrows*), and mid-systolic posterior movement of the anterior mitral leaflet is seen (*broad arrow*). *ECG*, electrocardiogram, *CW*, chest wall, *IS*, interventricular septum.

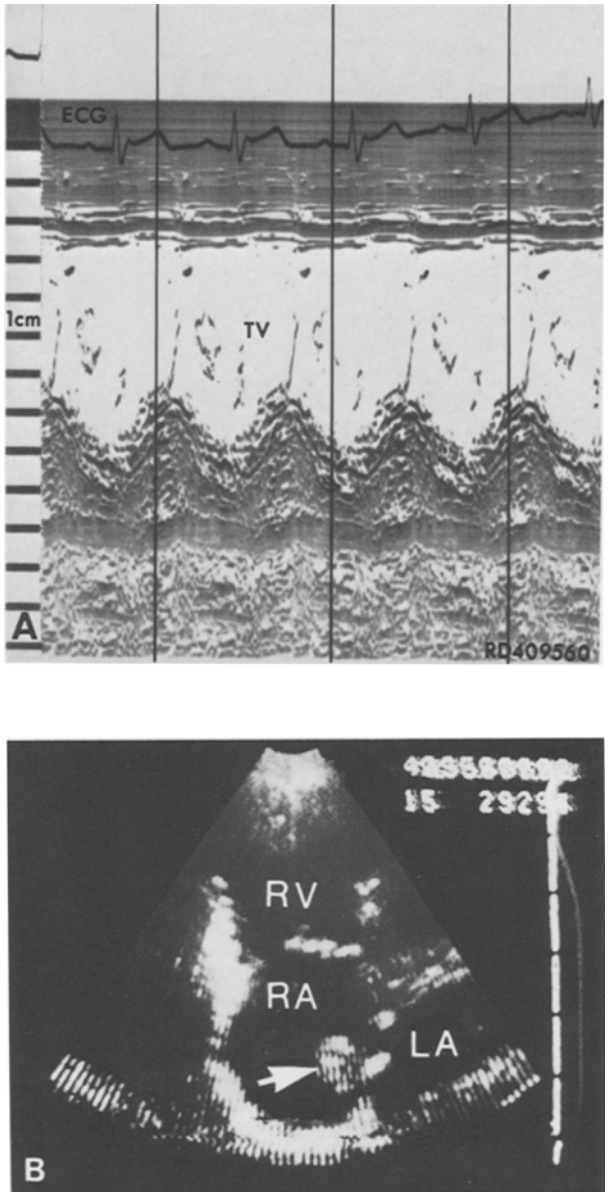


Fig. 2A and B. Echocardiography in right atrial myxoma. **A** A dense cloud of echos behind the tricuspid valve is seen on this M-mode echocardiogram. **B** Two-dimensional echocardiogram in the truncated apical view of the right atrial myxoma (*arrow*).

An analysis of the frequency of tumor calcification was made in the present study. Of the 263 cases reported, only six were described as having calcified tumors. All were primary tumors; calcification has never been described in metastatic tumors [25]. Four of the calcified tumors were fibromas, two in children [248, 255] and two in adults [253, 254]; one was a right atrial myxoma [249]; and the sixth was a left atrial malignant mesenchymoma earlier described by one of the present authors [252]. We have recently encountered a case of right atrial myxoma that was heavily calcified, and left atrial myxomas are also known to calcify occasionally [3]. The frequency found in this study (2%) is low compared to earlier descriptions in the literature in which calcifications in cardiac fibromas, hamartomas, rhabdomyomas, and angiomas have been described as occurring in as many as 20% of the cases [1]; it is likely that this higher rate is due to autopsy evaluation.

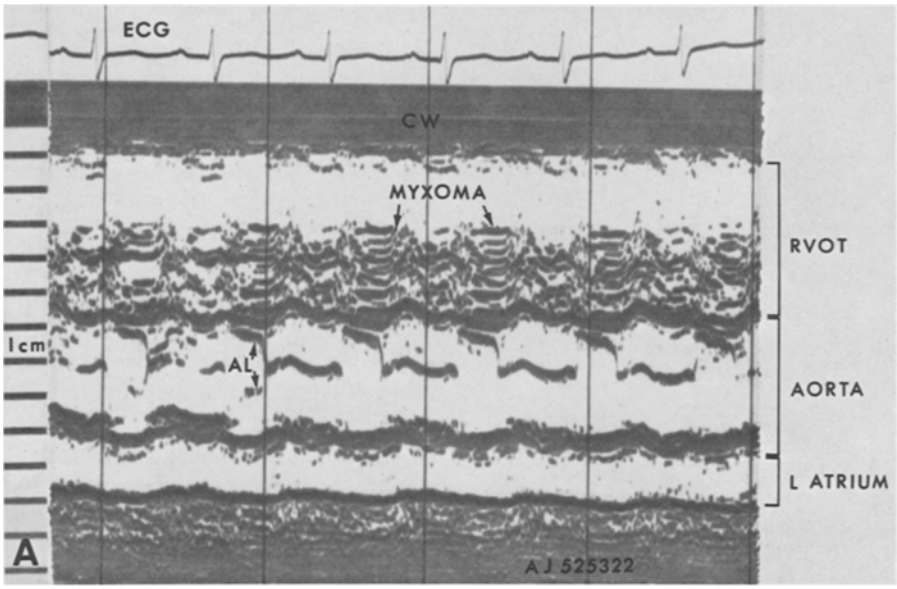
Echocardiography

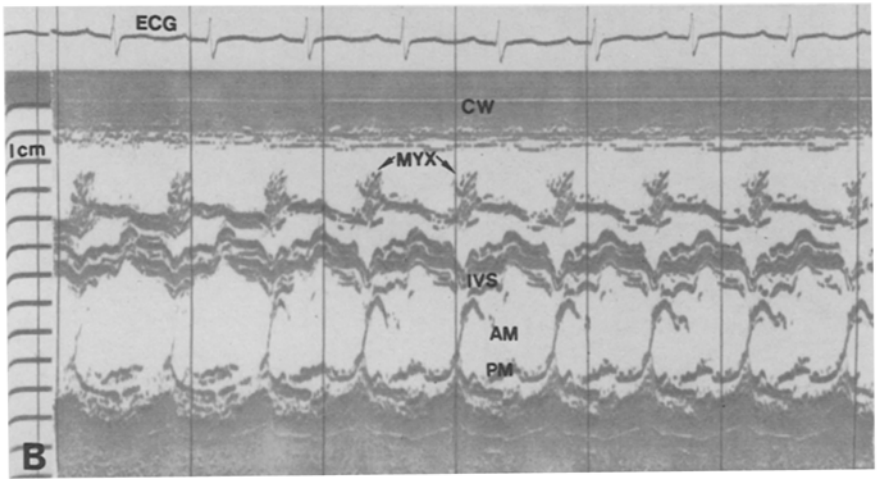
Due to the rapid advances in M-mode and two-dimensional echocardiography, a more detailed description of these ultrasound techniques in detecting tumors is of considerable importance. M-mode echocardiography provides an excellent nontraumatic technique for the detection of intracavitary as well as intramural cardiac tumors by indirectly delineating intracardiac anatomy [190–232]. The recent development of cross-sectional or two-dimensional echocardiographic techniques enables direct visualization of nearly all areas of cardiac anatomy and permits simultaneous visualization of multiple chambers [228].

The most prevalent cardiac tumor is the myxoma, and the predominant location for this benign tumor is the left atrium. Typically, left atrial myxomas are pedunculated and move freely within the left atrium throughout the cardiac cycle. They prolapse into the mitral valve orifice during diastole and are propelled back into the left atrium during ventricular systole. A left atrial myxoma is, therefore, potentially recognizable, not only as an intracavitary mass, but also by virtue of the motion exhibited.

The recognition of left atrial myxomas represented one of the earliest diagnostic applications documented for M-mode echocardiography [200]. The left atrial myxoma is shown as a cloud or dense conglomeration of echoes which characteristically appears within the mitral orifice, just behind the echo of the anterior mitral leaflet during diastole, and is subsequently propelled into the left atrial chamber during systole (Fig. 1A). An additional finding of diagnostic value in patients with left atrial myxomas is flattening of the mitral E-F slope in a manner similar to that found with mitral stenosis. There is often also a delay between the opening of the mitral leaflets and the appearance of the left atrial myxoma within the valve orifice in early diastole due to the time required for the tumor to prolapse into the orifice.

Although a cloud of echoes behind the anterior mitral leaflet in diastole is the echocardiographic hallmark of left atrial myxoma, certain tumors give rise to other, more unusual, echoes. Some left atrial myxomas, for example, are sessile, and therefore not capable of prolapsing into the mitral orifice. In patients with such tumors it is necessary to scan a large area





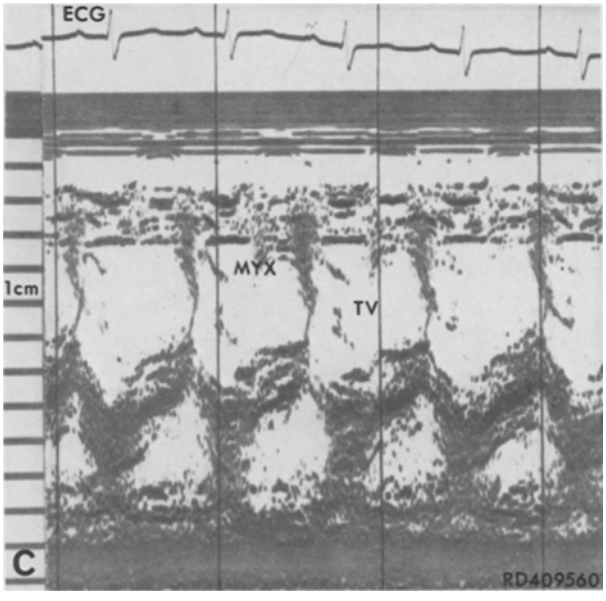
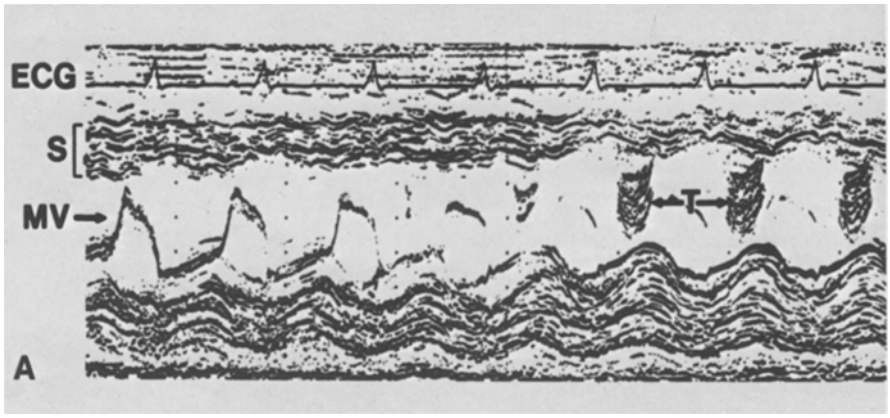


Fig. 3A–C. Right ventricular myxoma in a patient who earlier had a left atrial myxoma and would later present with a right atrial myxoma. **A** Echocardiogram showing multiple echoes in the right ventricular outflow tract (RVOT) indicating a tumor. **B** Echocardiogram at the level of the mitral valve demonstrating myxoma (MYX) in the right ventricle. **C** Echocardiogram of the anterior tricuspid leaflet (TV). *AL*, aortic leaflets, *PM*, posterior mitral leaflet, *RV*, right ventricle, *RA*, right atrium, *LA*, left atrium.



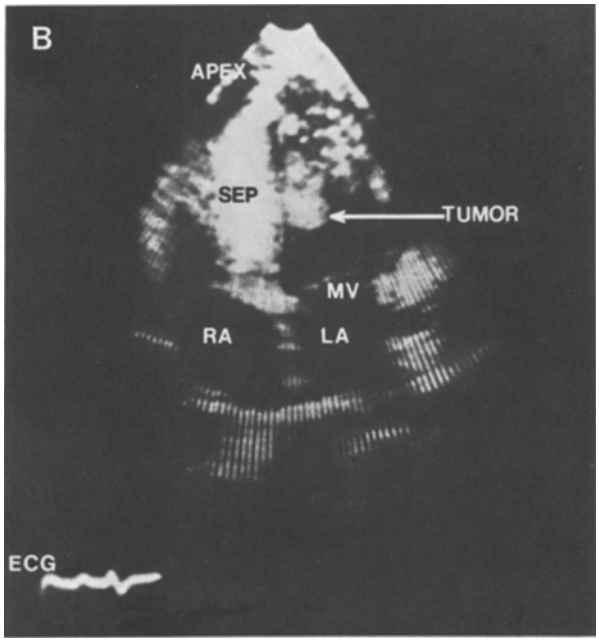


Fig. 4 A and B. Echocardiography in a case of melanoma metastatic to the left ventricle. **A** Dense mobile echos within the left ventricle in systole represent metastatic tumor. **B** Systolic stop frame from a two-dimensional echocardiogram, apex four-chamber view. The metastatic tumor mass is seen within the left ventricle. The apex of the heart is at the top of the figure, the base at the bottom. *LA*, left atrium, *MV*, mitral valve, *RA*, right atrium, *S* and *SEP*, septum, *T*, tumor. (Reproduced with permission from [222], courtesy of author and publisher).

of the left atrium to identify the immobile lesions. In addition, some myxomas are manifested only by a series of parallel linear echoes in the mitral valve orifice rather than by a dense cloud or conglomeration of ultrasonic signals (Fig. 1 B and C). This linear appearance of left atrial myxomas is attributable to an inhomogeneity of tissue structure within the tumor itself.

Myxomas are also found in cardiac chambers other than the left atrium. Right atrial myxomas appear as a dense cloud of echoes behind the tricuspid valve on M-mode echocardiograms (Fig. 2); they may also prolapse through the tricuspid orifice and present as masses within the right ventricular outflow tract. Indeed, patients with such tumors present clinically with symptoms of pulmonic stenosis, rather than of tricuspid stenosis. Primary myxomas have also been recognized by echocardiography in the left and right ventricles (Fig. 3A–C). The echocardiographic presentation of other intracavitary tumors is similar to that of myxomas, except that other tumors are rarely as mobile. Two examples of left ventricular tumors are seen in Figures 4 and 5.

The recent availability of two-dimensional echocardiography has contributed significantly to the echocardiographic assessment of cardiac tumors. Two-dimensional echocardiography is capable of directly imaging a cardiac mass rather than giving indirect evidence as does M-mode echocardiography; in addition, the ease with which the left atrium is visualized in multiple projections by two-dimensional echocardiography facilitates the detection of sessile myxo-

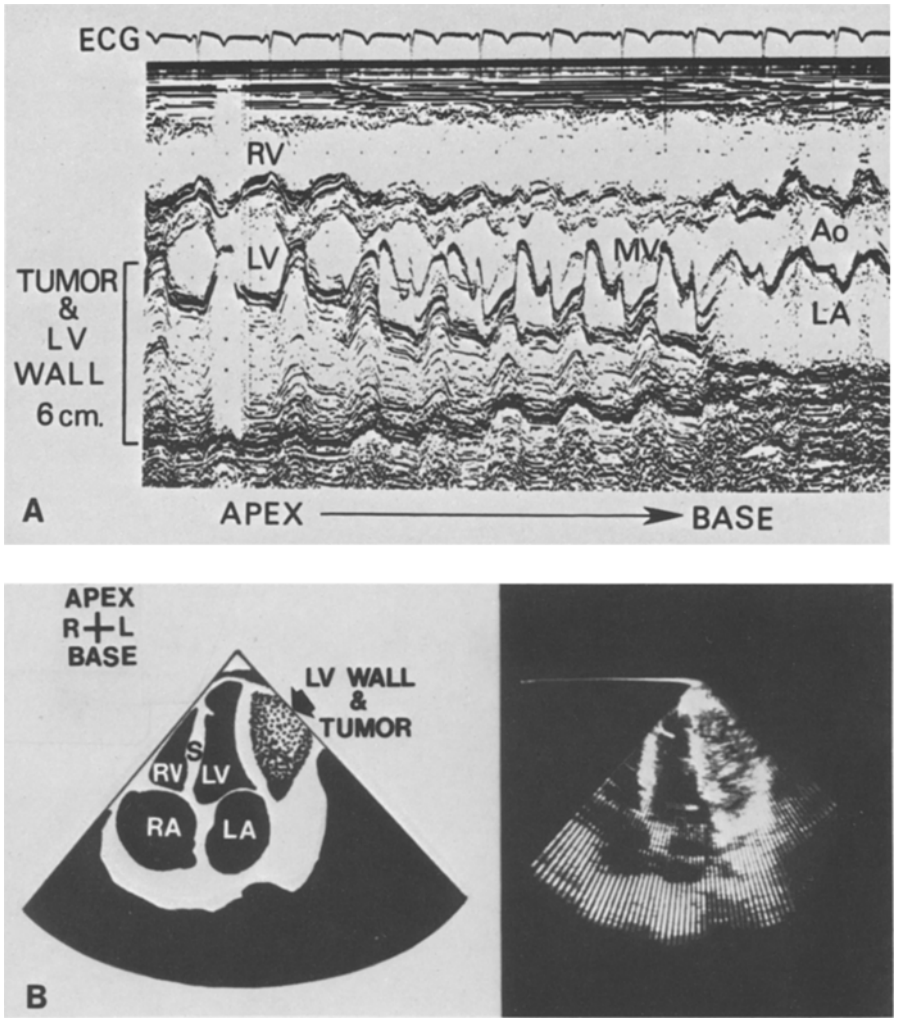


Fig. 5A and B. Echocardiography in left ventricular hemangioma. **A** M-mode sweep of the left ventricle from apex to base. Note the markedly thickened posterior left ventricular wall caused by the intramural tumor. **B** Two-dimensional echocardiogram, long axis view. Note the infiltration of tumor into the posterior left ventricular wall. *AO*, aorta, *LA*, left atrium, *LV*, left ventricle, *MV*, mitral valve, *RV*, right ventricle, *S*, septum. (Reproduced with permission from [222], courtesy of author and publisher.)

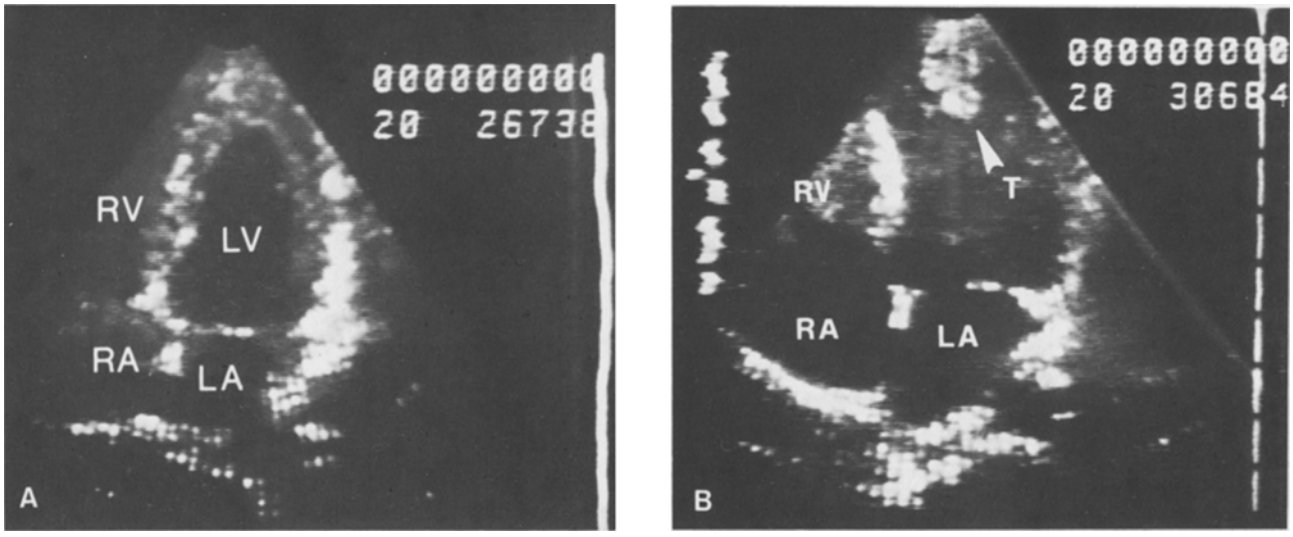


Fig. 6A and B. Two-dimensional echocardiogram in the four-chamber view of a left ventricular thrombus. **A** normal, **B** thrombus.

mas. Two-dimensional echocardiography enables simultaneous visualization of the right and left atria and thereby facilitates detection of biatrial cardiac myxomas (Fig. 2B). By cross-sectional echocardiography the point of origin of a tumor may be identified, making it possible to distinguish myxomas, which are characteristically attached to the atrial septum, from atrial thrombi. The four-chamber view clearly visualizes a left ventricular metastatic tumor mass in Fig. 4B. Another left ventricular tumor, a hemangioma, is seen in the long-axis view in Fig. 5B. Finally, the images produced by two-dimensional echocardiography often contain enough information about a cardiac tumor, such as a myxoma, that surgery can be undertaken on this basis alone.

It is important to know that a cloud or conglomeration of echoes at echocardiography is not specific for intracardiac tumors, but may be produced by a number of other cardiac lesions, the most common of which is an intracardiac thrombus (Fig. 6). In addition to thrombi, calcific deposits in cardiac valves and cardiac vegetations secondary to infectious endocarditis also appear as dense conglomerations of echoes on both M-mode and two-dimensional echocardiography. As with calcification of cardiac valves, the recognition of a cardiac vegetation is usually made by virtue of its association with a valvular structure on two-dimensional echocardiography. However, in certain instances vegetative and inflammatory masses involving chordae tendinae and the endocardium may be mistaken for cavitary tumors. It is therefore important to know that the echocardiographic appearance of a cardiac mass is similar, whether the mass consists of tumor, calcific deposits, thrombus, or vegetation. The location and motion pattern of the mass are extremely important in determining whether or not the lesion is neoplastic.

Echocardiography is also valuable in the detection of intramural tumors of the heart [222] by virtue of its ability to identify the two endocardial surfaces of the interventricular septum as well as the endocardial and epicardial surfaces of the free walls of the right and left atria and ventricles. The echocardiographic abnormalities characteristic of intramural tumors include a marked increase of the mural width in the area of the heart involved by the tumor. As with intracavitary tumors, the increase in wall thickness induced by cardiac tumors does not have a specific ultrasonic appearance, and one must distinguish thickening due to neoplasm from that induced by cardiac hypertrophy or myocardial infiltration with substances such as amyloid-glycogen and from endocardial thickening caused by mural left ventricular thrombus. An important factor in the distinction of these various causes of myocardial thickening involves the localized nature of the abnormality which is typically found with tumors. Thus, the disproportionate thickening of one area of the heart strongly suggests a cardiac tumor.

Finally, echocardiography has proved of value in the recognition of pericardial effusion secondary to malignant cardiac involvement. Malignant pericardial effusions are readily detected by both M-mode and two-dimensional echocardiography. The most common cause of such effusions is metastatic disease from carcinoma of the lung and breast, and typically these effusions are large. Unfortunately, ultrasonography cannot determine whether pericardial effusions are hemorrhagic or serous, nor can the actual metastatic growth in the pericardium usually be visualized [211].

In summary, we believe that echocardiography is a very accurate method for the detection of cardiac tumors. However, its accuracy is highly dependent on a careful and thorough examination producing high quality M-mode or two-dimensional echocardiograms and, perhaps most importantly, on an experienced interpreter.

Coronary Angiography

Although arteriography has been extensively employed in the diagnosis of tumors elsewhere in the body, its use in cardiac tumors has been limited. Myxomas are known to have a high degree of neovascularity and small arteriovenous fistulae formations [240, 244, 245]. However, the angiographic findings are not diagnostic, as left atrial and ventricular thrombi exhibit similar neovascularity and fistulae formation [243]. Neovascularity in a left ventricular tumor, presumably a cardiac hemangioma or hamartoma, has been described in a child [246]. A massive intrapericardial lipoma with neovascularity and vessel displacement [242] and an intrapericardial pheochromocytoma with neovascularity [241] have also been reported.

Endomyocardial Biopsy

There is no report of any cardiac tumor diagnosed by myocardial biopsy, although the technique could probably be used to diagnose cardiac tumors [260, 261].

Differential Diagnosis

The most common mimickers of cardiac tumors at angiocardiography are thrombi, most of which are

seen in the left atrium or the left ventricle [256, 259]. Rare cases of left atrial aneurysms have been reported to mimic left atrial tumors [258], and we have seen a greatly enlarged left atrium in a case of cor triatriatum that resembled a tumor. Pericardial cysts may be difficult to distinguish from tumors. Rare hydatid cysts have been described as mimicking tumors [257].

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