Echocardiography of Right Ventricular Tumors

THOMAS A. PORTS, M.D., NELSON B. SCHILLER, M.D., AND BRIAN L. STRUNK, M.D.

SUMMARY The clinical and echocardiographic findings of four patients with right ventricular tumors (three myxomas and one metastatic melanoma) are presented. All four patients previously had been evaluated by competent cardiologists, and the diagnosis of a right ventricular tumor was missed. The initial echocardiograms were interpreted as not showing any right ventricular tumor; but on review we found that the correct diagnosis could have been made. The presence of the right ventricular myxoma was confirmed at angiography and surgery. However, the attachment of the stalk of the myxoma could not be determined confidently by angiography alone, and in two of the three cases two-dimensional echocardiography was required to identify correctly the location of the base of the stalk.

This paper emphasizes the usefulness and technical difficulties of M-mode and two-dimensional echocardiography in the diagnosis of a right ventricular tumor.

INTRACAVITARY CARDIAC TUMORS are uncommon; when encountered, they are usually atrial myxomas.1 Primary and secondary intraventricular cardiac tumors are exceedingly rare and their clinical presentation highly variable.2-4 As a result, the antemortem diagnosis of these potentially curable lesions is frequently missed. Echocardiography is useful in the diagnosis of atrial tumors.6-9 By contrast, echocardiographic experience with intraventricular tumors in both children and adults is limited.10-13 In this paper, we review four cases of right ventricular tumor, with emphasis on the spectrum of M-mode and two-dimensional real-time echocardiographic findings. Echocardiographic patterns are correlated with the angiographic, surgical or postmortem anatomic findings. Echocardiographic techniques useful in the diagnosis of right ventricular mass lesions and the potential sources of interpretive error are also discussed.

Materials and Methods

Patients ranged in age from 26 to 65. Only one patient had a prior history of cardiac disease. In none of the patients was right ventricular tumor suspected as the initial clinical diagnosis.

Echocardiographic Studies

M-mode echocardiographic studies were performed using either a Picker EV 10 echocardiograph, Unirad series 100 echocardiograph interfaced with a Honeywell 1856 strip chart recorder, or Smith Kline Echoline-20A Ultrasonoscope interfaced with an Irex 101 strip chart recorder. A variety of transducers of differing focal length and frequency were used for each patient in order to maximize the anteriorly reflected signals. Recordings were done with the patient in the supine or 30° left lateral decubitus position with the transducer positioned in the third, fourth or fifth intercostal space at the left sternal border. The ultrasonic beam was directed to obtain the left ventricular, mitral valve and aortic root echoes in the usual manner and then angled to the right to obtain echoes in the plane of the tricuspid valve. The transducer was then angled toward the left shoulder obtaining a sweep of the right ventricular outflow tract. In each case, the ultrasonic beam was directed to show simultaneously the anterior right ventricular wall, right ven-

cular cavity and interventricular septum. Coarse gain attenuation was varied during the outflow tract sweep, and the near gain and depth compensation were set in order to receive optimally echoes from the right ventricular cavity and were varied systematically to verify that the encountered intracavitary echoes were indeed produced by tissue and were not artifact.

Two patients were studied by two-dimensional echocardiography. The instrument employed was a prototype of a Varian Associates 32-element phased array, wide angle (80°) sector scanner. Two-dimensional images were obtained in the long axis or sagittal plane by directing the echo sweep between the apex and the base of the heart. Short axis or transverse views were obtained by directing the plane of sweep along a line drawn between the right hip and the left shoulder perpendicular to the long axis of the left ventricle and following the course of the right ventricular outflow tract.14

The images were permanently recorded on video-tape. The illustrations presented here were obtained from Polaroid photographs of stop-action single frame scan images from the video-tape recordings. This process results in a reduction of image quality and loss of the visual appreciation of motion normally present in phased-array real-time recordings.

Patients

Case 1

The patient was a 65-year-old black female who had sustained previous large myocardial infarctions in 1960 and 1969. She had been maintained on digitalis and diuretics for severe biventricular failure since 1964. A permanent demand transvenous right ventricular pacemaker was implanted in 1974 for symptomatic bradycardia. When the patient was hospitalized in 1975 for worsening congestive heart failure, murmurs of tricuspid regurgitation and mitral regurgitation were first noted. An echocardiogram at that time revealed enlarged left and right ventricular dimensions. Paradoxical septal motion was noted, as were linear echoes in the right ventricular cavity which were thought to represent the right ventricular pacemaker. Right heart catheterization, done at the bedside, showed the right atrial pressure to be 12, the right ventricular pressure 32/12, the pulmonary artery pressure 35/14 and the pulmonary capillary wedge pressure 18. The arterial pressure was 94/80 and the cardiac output 3.8 L/min. Nitroglycerin ointment was added to the patient's therapy with hemodynamic improvement. In August 1975 an epicardial pacemaker was inserted, replac-

From the Department of Medicine, Cardiovascular Division, University of California, School of Medicine, San Francisco, California.

Address for reprints: Thomas A. Ports, M.D., Room 1186-M, University of California, San Francisco, California 94143.

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ing the failing transvenous pacemaker. The patient was hospitalized again in October 1976 for worsening congestive heart failure and pacemaker battery replacement. After the battery pack had been changed, the patient became febrile, developing new pulmonary infiltrates and worsening right sided congestive heart failure. An echocardiogram was done to aid in the evaluation of possible bacterial endocarditis.

Echocardiographic Findings

The right and left ventricular dimensions were again enlarged, and paradoxical septal motion was noted. Again, unusual dense linear echoes were noted in the right ventricle about the tricuspid valve. In the right ventricular outflow tract sweep these echoes were seen in both systole and diastole and were initially thought to be valvular vegetations (fig. 1). On expanded view they were noted to be separate from the tricuspid valve (fig. 2). Two-dimensional echocardiography demonstrated the presence of a mobile right-ventricular mass attached to the interventricular septum just below the tricuspid valve (fig. 3).

Angiographic and Surgical Findings

Following a right atrial injection, a right ventricular forward angiogram was done which confirmed the presence of an approximately 2 × 2 cm mobile right ventricular mass. The patient subsequently underwent surgery and a 1 × 2 cm right ventricular mass was resected. The tumor was attached to the interventricular septum just below the level of the septal leaflet of the tricuspid valve. On histologic examination the tumor was found to be a myxoma.

Case 2

The patient was a 36-year-old black female referred to the echocardiographic laboratory for evaluation of a previously undetected cardiac murmur. She was asymptomatic, except for the presence of sharp anterior chest pains for approximately one year which were not related to exertion. On physical examination the vital signs were normal. The lungs were clear. The jugular venous and carotid pulses were normal, as were the first and second heart sounds. There was a loud harsh systolic ejection murmur at the lower left sternal border with several prominent intermittent systolic clicks. The murmur increased in intensity with inspiration. The chest X-ray and electrocardiogram were normal.

Echocardiographic Findings

Echocardiographic examination demonstrated normal mitral and aortic valves and left ventricular dimensions. The right ventricle was mildly enlarged. Dense echoes were noted in the right ventricular cavity in diastole and in the right ventricular outflow tract in systole (fig. 4).

Angiographic and Surgical Findings

Right heart catheterization demonstrated normal right atrial and ventricular pressures. Following a right atrial injection, a right ventricular forward angiogram was done, and the presence of a 2 × 2 cm freely mobile intraventricular mass was noted. The patient subsequently underwent successful resection of a right ventricular myxoma attached by a long stalk to the interventricular septum at the level of the tricuspid valve. The postoperative echocardiogram (fig. 5) was normal.

Case 3

The patient was a 26-year-old white male, previously reported in the medical literature for a hormonally-active,
benign, interstitial cell tumor of the testes. During numerous physical examinations as a child and adolescent no cardiac murmurs were detected. There was no history of rheumatic fever. The patient was quite active and able to do heavy construction work without cardiac symptoms. During a recent hospitalization for presumed viral gastroenteritis, a harsh systolic ejection murmur was noted for the first time. Blood cultures were negative. The electrocardiogram demonstrated right ventricular hypertrophy. The chest X-ray was normal. An echocardiogram done by the referring cardiologist demonstrated enlarged right ventricular dimensions, normal left ventricular function and normal mitral, tricuspid, pulmonic and aortic valves. The patient was referred for catheterization to evaluate his murmur. On physical examination the vital signs were normal. The lungs were clear. The jugular venous and carotid pulses were normal. There was a left parasternal lift. The first heart sound was normal, and the second heart sound was felt to be widely split with a loud pulmonic component. There was an ejection click noted in the pulmonic area. A grade III/VI harsh systolic ejection murmur, which decreased with inspiration, was present over the right ventricular outflow tract. Cardiac catheterization revealed a 60 mm gradient across the right ventricular outflow tract. A right ventricular angiogram demonstrated the presence of a large, mobile, lobulated mass moving out the right ventricular outflow tract and appearing to strike the pulmonic valve in systole. The origin of the tumor was unclear from the angiographic studies.

**Echocardiographic Findings**

Echocardiographic studies subsequent to the angiogram demonstrated a normal left ventricle and enlarged right ventricular dimensions. The mitral and aortic valves were normal, as was the tricuspid valve. On the right ventricular
sweep multiple dense linear echoes were observed in the right ventricular outflow tract extending to and striking the pulmonic valve in systole (figs. 6 and 7). A simultaneous phonocardiogram documented the presence of a systolic ejection murmur and a systolic click thought to be produced by the tumor striking the pulmonic valve. Two-dimensional echocardiography confirmed the presence of a large mobile mass in the right ventricular outflow tract and demonstrated its point of attachment to the interventricular septum above the tricuspid valve (fig. 8). It appeared to strike the pulmonary valve in systole.

**Surgical Findings**

The patient underwent successful removal of a large 4 × 6 cm right ventricular mass, which was attached to the interventricular septum just above the tricuspid valve. Histologically, the tumor was a myxoma. Postoperative M-mode and two-dimensional echocardiography were normal, and the previously-noted dense intracavitary echoes were no longer present (figs. 6, 7 and 9).

**Case 4**

The patient was a 42-year-old white male with no previous history of cardiopulmonary disease. In 1974 he had a malignant melanoma excised from the midscapular area. He did well for approximately one year until the diagnosis of metastatic disease to the lungs and liver was made. He entered the hospital in 1976 with acute shortness of breath. Electrocardiogram showed a new right bundle branch block, and blood gases showed arterial hypoxemia. A lung scan was abnormal. A presumptive diagnosis of pulmonary embolism was made and the patient was given anticoagulants. Except for tachypnea, the admission physical examination was unremarkable, and no abnormalities were detected in the cardiorespiratory system. After some initial improvement, the shortness of breath became progressively worse,

**Figure 5.** Postoperative M-mode echocardiogram from case 2. Previously detected echoes in the right ventricle are gone.

**Figure 6.** Left panel shows right ventricular outflow tract sweep in case 3. Dense echoes (myxoma) are seen beneath the pulmonic valve. Right panel shows absence of these echoes after removal of the tumor. AAW = anterior aortic wall; PAW = posterior aortic wall; PV = pulmonary valve.
and the cardiac silhouette was noticed to be enlarging. An echocardiogram was done to exclude the possibility of a pericardial effusion. The echocardiogram was felt to be a technically suboptimal study and interpreted as showing no significant pericardial effusion, mitral valve prolapse, and paradoxical septal motion. Dense intracavitary echoes were present in the right ventricle, but were thought to be artifactual. Five days later the patient experienced acute hemodynamic decompensation with marked neck vein elevation and Kussmaul's sign. It was felt the patient was having pericardial tamponade, possibly secondary to intrapericardial hemorrhage, exacerbated by his anticoagulation. An emergency echocardiogram was done (figs. 10 and 11) and was interpreted as showing no pericardial effusion. Dense intracavitary echoes in the region of the right ventricle were again noted and felt to be consistent with a space-occupying mass. Hemodynamic monitoring was attempted in the Coronary Care Unit, but the catheter could not be passed beyond the right atrium, in which a pressure of 28 mm Hg was recorded. Pericardiocentesis was attempted but was unsuccessful. The patient died shortly thereafter with intratable heart failure.

**Autopsy Findings**

Postmortem examination revealed that the cavity of the right ventricle of the heart was 90% occluded by a large polypoid mass protruding from the myocardium into the chamber. The tumor adhered to the tricuspid valve leaflets, making them immobile. Smaller tumor nodules were also
FIGURE 9. Systolic stop-frame image from case 3 after removal of right ventricular tumor. Previously detected mass in right ventricular outflow tract is gone.

Discussion

Primary or secondary space-occupying tumors of a cardiac chamber may be the actual pathology in patients initially suspected of having pericardial, myocardial or valvular heart disease. The relative rarity of cardiac tumors combined with the wide spectrum of clinical presentation makes their diagnosis most challenging. Early recognition of the presence of such tumors offers the patient the possibility of improvement or cure with surgical excision. Establishing the diagnosis prior to cardiac catheterization can be important because the diagnostic accuracy of the angiographic study is frequently increased and the potential risk of tumor embolization is diminished.16 Although echocardiography has been established as a safe and reliable technique for the diagnosis of atrial tumors,6** the echocardiographic experience with nonatrial tumors is extremely limited, and the two-dimensional echocardiographic findings have not been

FIGURE 10. M-mode sweep from base to apex with anterior focus transducer in case 4. Dense columns of echoes (tumor) are seen occupying almost the entire right ventricle. Pattern of pansystolic mitral valve prolapse is seen.
reported previously. Allen et al.\textsuperscript{10} reported the echocardiographic features of a right ventricular tumor in a neonate, and Farooki et al.\textsuperscript{11} discussed the echocardiographic features in two infants with ventricular septal rhabdomyoma. DeMaria et al.\textsuperscript{12} reported a case of a right ventricular myxoma diagnosed by echocardiography four years after the resection of a left ventricular myxoma.

Our four cases represent a spectrum of echocardiographic experience with right ventricular tumors. Cases 1 and 2 represent small pedunculated, mobile right ventricle myxomas occurring in the outflow tract. The tumors are represented echocardiographically by a cluster of dense echoes appearing in the right ventricular outflow tract intermittently in various phases of the cardiac cycle. In case 2 these abnormal echoes disappeared after successful operative removal of the tumor (fig. 5). In case 3 a large mobile right ventricular myxoma is represented echocardiographically by dense echoes seen in both systole and diastole in the right ventricular outflow tract (figs. 6 and 7). A postoperative echocardiogram (figs. 6 and 7) illustrates the absence of these echoes following successful removal of the tumor. Case 4 represents near-total replacement of the right ventricular cavity by a metastatic tumor. This case is perhaps the most difficult to diagnose echocardiographically because no right ventricular valves can be identified and familiar landmarks are difficult to appreciate. The tumor is represented by columns of dense echoes filling the right ventricular cavity which are not obliterated by maximum gain attenuation (figs. 10 and 11). These echoes appear to move with the heart cycle, but in an abnormal manner. The variable density of the echoes and the clear space observed at the right ventricular side of the septum may represent the inhomogeneity of this large polypoid tumor. Exaggerated paradoxical septal motion is noted, perhaps a result of right ventricular distention by the tumor. The pattern of mitral valve prolapse is also observed. It is either artifactual because of the peculiar systolic motion imparted to the heart by the infiltrating intraventricular tumor or true prolapse secondary to the papillary muscle involvement by metastatic tumor which was found at autopsy.

Because the right ventricle is the most anterior of the four cardiac chambers and lies directly beneath the sternum, it is often the most difficult to evaluate echocardiographically. As a result, the right ventricle is frequently ignored or suboptimally examined in routine echocardiography. With the relative technical difficulty in echoing the right ventricle and the interpretive problems arising from echoes that are not directly related to a known cardiac structure, it is not surprising that in each of the four cases presented here at least one experienced echocardiographer missed the diagnosis of the right ventricular tumor. In case 1 the abnormal echoes were initially recorded without difficulty but interpreted first as being a pacemaker wire and one year later as vegetations on the tricuspid valve. In case 2 the echoes likewise were recorded, but were felt possibly to be originating from a tangential view of an abnormal tricuspid valve. The prominent abnormal echoes in case 3 were missed by over-damping the anterior signals, and the study was interpreted as normal. The echoes in case 4 were thought to be artifactual.

These cases illustrate some of the problems and potential errors encountered in the interpretation of M-mode echocardiography of right ventricular masses. It is obvious that the diagnosis will be missed if the tumor does not traverse the path of the echo beam. Because right ventricular myxomas are often mobile and in the outflow tract,\textsuperscript{17} the diagnostic accuracy of the echocardiographic study is greatly enhanced if a right ventricular outflow tract sweep is carefully done. Figure 12, a preoperative echo from case 3, when compared with figures 6 and 7 illustrates that even a large mobile tumor can be missed if a right ventricular outflow tract sweep is not done and the echocardiographer in-

\begin{figure}[h]
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\caption{Expanded M-mode echocardiogram of right ventricle in case 4 showing dense echoes filling the right ventricle which are not obliterated by damping (right panel).}
\end{figure}
stead concentrates on views of the tricuspid and pulmonary valves. Likewise, if the anterior gain setting is too low, the entire right ventricle or any mass contained within it obviously will be missed. For this reason, it is important that the gain setting and depth compensation be adjusted to receive signals adequately from the anterior right ventricle and interventricular septum. In this regard, we have found that the use of a 3.5 MHz transducer often greatly improves the resolution of the right ventricle. Just as overattenuation and improper depth compensation can result in the obliteration of an anterior right ventricular mass, a nonexistent mass can be created by too much anterior gain. The echocardiographer must determine if anterior signals received are artificial or attributable to real tissue. It is important to attenuate the gain systematically and observe if the intraventricular echoes are still perceived at the time that the anterior right ventricle and septal signals are received. Real tissue (tumor) is usually represented by dense echoes, often with a motion consistent with the heart cycle, whereas artifactual anterior echoes caused by too much anterior gain appear as diffuse, fuzzy and nonmobile echoes. When confusing signals are received from the right ventricle, we have found that the use of an expanded view of the area in question is helpful in determining whether the echoes originate from a valvular or adjacent structure (fig. 2).

Real-time, two-dimensional echocardiography has proved to be extremely useful in further defining intracavitary masses. With two-dimensional echocardiography accurate visualization of the right ventricular body and outflow tract can be accomplished consistently. The acoustic nature and anterior location of right ventricular myxomas make them appear as bright, mobile masses. The mobile nature of the tumor can easily be appreciated, and its point of attachment, or stalk, can be visualized accurately. This information may be extremely important in planning catheterization and surgery.

As these cases illustrate, the echocardiographic pattern of right ventricular tumors can be quite dramatic. But as obvious as they may seem, their diagnosis is not simple. Clinical suspicion combined with careful and systematic echocardiographic examination is required for accurate diagnosis. With the use of two-dimensional echocardiography to complement M-mode techniques, the noninvasive diagnosis of right ventricular tumors should be more reliable.

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**References**

Two-dimensional Echocardiographic Findings in Atrial Septal Defect

WILLIAM LIEPPE, M.D., RALPH SCALLION, M.D., VICTOR S. BEHAR, M.D., AND JOSEPH A. KISSLO, M.D.

SUMMARY Two-dimensional echocardiographic findings in a group of 24 patients with atrial septal defects were correlated with findings obtained by cardiac catheterization and M-mode echocardiography. The prevalence of mitral prolapse was 95% by two-dimensional echocardiography and 59% by angiography in patients with secundum and sinus venosus atrial septal defects. The majority of the group with echocardiographic prolapse manifested a distinctive pattern of prolapse, with predominant involvement of the anterior mitral leaflet. Thirty-eight percent of the patients in this series manifested paradoxical septal motion by M-mode and/or two-dimensional echocardiography. In the patients with abnormal septal motion, the net systolic anterior movement of the septum was caused by an exaggerated systolic anterior movement of the main body of the left ventricle. The atrial septal defect could be visualized with confidence by two-dimensional echocardiography only in the two patients with ostium primum atrial septal defects. Cleft anterior mitral leaflets were also clearly demonstrated in these two patients.

PATIENTS WITH ATRIAL SEPTAL DEFECTS and other forms of right ventricular volume overload frequently manifest right ventricular enlargement and paradoxical septal motion on echocardiography. However, several recent studies have offered conflicting explanations for the paradoxical septal motion. Other reports have documented the frequent association of secundum atrial septal defect with angiographically manifest prolapse of the posterior mitral leaflet.

Two-dimensional echocardiography is a relatively new technique which has been shown to provide spatial information concerning cardiac structures not readily available by conventional M-mode echocardiography. Furthermore, it was suggested recently by Gilbert et al. that cross-sectional echocardiography may also be superior to angiography for the detection of certain types of mitral prolapse.

This study correlates the cross-sectional echocardiographic, M-mode echocardiographic and angiographic findings in 24 patients with catheterization documented atrial septal defects in an effort to determine the relative role of two-dimensional echocardiography in the evaluation of patients with this entity.

Methods

Patients

The initial patient group included a total of 29 consecutive patients with atrial septal defects who were referred to the Duke Cardiovascular Laboratory for catheterization between September 1974 and October 1976. Each patient underwent a complete clinical evaluation, which included physical examination, electrocardiogram, and chest X-ray. Two-dimensional echocardiograms were performed on all patients. For these studies to be considered technically acceptable, both mitral leaflets and the endocardium of the posterior free wall and septum had to be visualized throughout the cardiac cycle. In five of the initial group of 29 patients (17%) the two-dimensional echocardiogram was technically inadequate, and these patients were excluded from the study. M-mode echocardiograms were available on 17 of the remaining 24 patients (71%). Both the two-dimensional and M-mode echocardiograms were done within three days of cardiac catheterization.

Table I includes a summary of pertinent clinical, hemodynamic, angiographic, and echocardiographic data from the 24 patients included in the study.
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