Echocardiographic, Angiocardio graphical, and Surgical Correlations in Right Ventricular Myxoma Simulating Valvar Pulmonic Stenosis

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SUMMARY The echocardiographic, angiographic and surgical correlations in a patient with a right ventricular myxoma simulating valvar pulmonic stenosis are presented. A dense mass of echoes was seen in the body of the right ventricle in diastole; in the right ventricular outflow tract throughout the cardiac cycle; and within the pulmonic valve in systole. These echocardiographic findings suggested the presence of a mass high in the right ventricle, which moved through the pulmonic valve in systole. This impression was confirmed by angiography and surgery.

A 53 mmHg gradient across the pulmonic valve was noted at cardiac catheterization. At operation a myxoma which originated above the crista supraventricularis was found. The tumor moved into the main pulmonary artery in systole. Surgical removal of the tumor resulted in the disappearance of the abnormal echoes.

THE VALUE OF ECHOCARDIOGRAPHY in the diagnosis of left atrial myxoma has been clearly demonstrated.1-3 Right atrial and right ventricular tumors are less frequently seen than left atrial tumors. We describe the echocardiographic features of a patient with a right ventricular myxoma who had been diagnosed previously as having valvar pulmonic stenosis. The tumor prolapsed through the pulmonic valve producing a 53 mmHg gradient across the pulmonic valve.

Case Report

A sixteen-year-old white male presented with a complaint of fatigue and mild exertional dyspnea of six months' duration. He denied a history of dizziness, syncope, or chest pain. A heart murmur had been noted for the first time at the age of 13 years. Two years prior to the present admission he had undergone cardiac catheterization at another hospital, where a diagnosis of valvar pulmonic stenosis had been made. The gradient at the pulmonic valve was 50 mmHg. Surgery was not recommended at that time. His past history was remarkable in that a myxoma (4 x 3.3 x 2.5 cm) had been surgically removed from his left axilla at the age of 12 years.

On examination the only abnormal findings were limited to the cardiovascular system. His pulse was 80/minute and regular and his blood pressure was 110/70. The jugular venous pressure was not elevated; the liver was not palpable and there was no peripheral edema. A prominent heave and a systolic thrill were palpable in the second and third left interspaces. A grade IV/VI long ejection systolic murmur was best heard in the same area. Following the aortic component of the second sound (A2), a low pitched sound which was widely separated from A2, was heard.

The electrocardiogram showed right ventricular hypertrophy and normal sinus rhythm. His chest X-ray revealed right ventricular enlargement and pulmonary artery dilatation with normal pulmonary vascularity. The phonocardiogram showed a long ejection systolic murmur and a low pitched diastolic sound which was 140-160 msec from A2.

Echocardiography was performed with the patient supine and in the left lateral decubitus position. A 2.25 MHz, 10 cm focus, 0.5 inch transducer, an Ekoline 20 Ultrasonoscope and a Honeywell 1856 recorder were used. An M-Mode sweep from the aortic root to the left ventricle was performed. The pulmonic valve was recorded using previously described methods.

Preoperative Echocardiograms

The pulmonic valve echocardiogram is illustrated in figure 1. A dense echo-producing mass is seen to move through the pulmonic valve in systole. There is an echo-free space between the pulmonic valve in the open position and the

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**FIGURE 1.** Echocardiogram of pulmonic valve (PV). Note the dense mass of echoes (T) which moves through the pulmonic valve in systole.
mass. Simultaneous high speed (100 mm/sec) recording of the pulmonic valve, phonocardiogram and carotid pulse tracing is shown in figure 2. A diastolic sound (P) which coincides with maximum anterior motion of the tumor mass and with the delayed closure of the pulmonic valve is seen. Figure 3 shows that tumor echoes are present both in systole and diastole in the right ventricular outflow tract, i.e., anterior to the aortic root. More inferiorly in the right ventricle, tumor echoes are visible only in diastole (fig. 4). The tricuspid valve was normal.

Cardiac Catheterization revealed a mean right atrial pressure of 5 mmHg; a right ventricular pressure of 75/9 mmHg; a pulmonary artery pressure of 22/8 mmHg, and a mean pulmonary wedge pressure of 7 mmHg. There was no evidence of a shunt at the atrial or ventricular level.

Right atrial cineangiography in the PA and lateral projections demonstrated a large lobulated mass high in the right ventricular cavity. The mass was seen to move into the pulmonary artery in systole and flop back into the ventricle in diastole. A systolic frame is shown in figure 5; the tumor is visualized in the pulmonary artery and right ventricular outflow tract.

Operative Findings

The right ventricle was moderately enlarged. The tumor was felt to bump against the anterior surface of the right ventricular outflow tract and move into the main pulmonary artery during systole. The pulmonary artery was enlarged. When the ventriculotomy was performed, a large tumor mass, approximately 8 cm x 3 cm x 3 cm, was identified. A portion of the tumor mass was gray and glistening and the remainder was dark red, apparently from hemorrhage into

FIGURE 2. High speed (100 mm/sec) recording of pulmonic valve (PV), carotid pulse (CA), and phonocardiogram. A diastolic sound (P) is present which coincides with maximum anterior (i.e., towards the right ventricular outflow tract) motion of the tumor mass and with the delayed closure of the pulmonic valve. S, = first heart sound, A, = aortic component of the second heart sound, T = tumor mass.

FIGURE 3. A sweep from the aortic root (AO) to the mitral valve (AMV) is shown. A dense mass of echoes (T) is present anterior to the aortic root both in systole and diastole. LA = left atrium.

FIGURE 4. Ultrasound recording showing an abnormal mass of echoes (T) in the body of the right ventricle (RV) in diastole. LV = left ventricle.

FIGURE 5. A systolic frame from a right atrial angiogram showing the tumor in the main pulmonary artery and the right ventricular outflow tract.
FIGURE 6. Photograph of tumor after it was excised. The white structure on the left border is the stalk of the mass.

The tumor originated from the right ventricular wall just anterior and superior to the crista supraventricularis. The stalk had a firm white fibrous base and when the stalk was removed, the tumor could be delivered through the ventriculotomy without difficulty. The pulmonary valve was grossly normal. There was some dilatation of the pulmonary valve anulus. The tumor (fig. 6) was excised and the ventriculotomy closed.

There was a dramatic change in the cardiac findings following operation. The diastolic sound and the systolic thrill were no longer present. A grade II/VI short ejection systolic murmur was heard.

Postoperative Echocardiograms

The postoperative echocardiogram revealed a normal pulmonic valve (fig. 7). The abnormal mass of echoes in systole was no longer present. The right ventricular outflow tract and the right ventricular cavity were also free of abnormal echoes (fig. 8).

FIGURE 7. Postoperative pulmonary valve (PV) recording. The abnormal systolic echoes seen on the preoperative tracing are no longer present.

FIGURE 8. Sweep from the aortic root (Ao) to the left ventricle (LV) recorded after surgery. The abnormal echoes which were seen anterior to the aortic root and in the right ventricular cavity are no longer present.
Discussion

This patient demonstrated abnormal echoes which were present in the pulmonic valve echocardiogram during systole, in the right ventricular outflow tract in systole and diastole, and more inferiorly in the right ventricular cavity in diastole. The tricuspid valve appeared normal. These findings suggested the presence of a tumor high in the right ventricle, which traversed the pulmonic orifice in systole. This impression was confirmed by angiography and surgery. The diastolic sound noted on the phonocardiogram could have been due to a tumor "plop" or delayed pulmonary valve closure. The auscultatory characteristics of the sound, i.e., a low-pitched thud, were more suggestive of a tumor sound.

Myxomas of the right ventricle are extremely uncommon. They may produce syncope by causing pulmonary outflow tract obstruction. In some instances a right ventricular myxoma may closely simulate pulmonic stenosis. It is of interest that our patient was thought to have valvular pulmonic stenosis on the basis of a pulmonic valve gradient at the time of his first cardiac catheterization. Angiography was not performed at that time. DeMaria and associates described a patient with a right ventricular myxoma, who had a 40 mmHg gradient across the pulmonic valve. They noted a mass of echoes in the right ventricular outflow tract, right ventricle and contiguous to the anterior tricuspid leaflet. They did not describe the appearance of the pulmonic valve in their patients.

A myxoma of the left axilla was excised when our patient was 12 years old. A cardiac murmur was first noted at the age of 13. It is conceivable that tumor cells entered a vein and embolized to the right ventricle at the time of excision of the myxoma of the axilla. An alternative explanation would be the chance occurrence of two myxomata.

This case illustrates the importance of examination of the pulmonic valve in patients who present with a clinical diagnosis of pulmonic stenosis.

References


Echocardiographic Observations on the Association between Mitral Valve Prolapse and Asymmetric Septal Hypertrophy

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SUMMARY One hundred and ninety consecutive patients with mitral valve prolapse (MVP) were studied by echocardiography. Asymmetric septal hypertrophy (ASH) was noted in 16 patients. Three patients had syncope which was associated with supraventricular arrhythmias. Three others had episodes of presyncope which were not related to disturbance of rhythm. One of the patients with MVP and ASH had a family history of idiopathic hypertrophic subaortic stenosis. The septal thickness ranged from 1.6 to 3.1 cm, mean = 2 cm. The posterior wall thickness ranged from 0.7 to 2.1 cm, mean = 1.0 cm. The ratio of the thickness of the interventricular septum to that of the posterior wall ranged from 1.5 to 2.5, mean = 1.9. The percentage of thickening of the septum in systole was reduced in 13 patients. The excursion of the interventricular septum was reduced in three patients. In nine patients the left ventricular end-systolic dimension was below the lower limit of normal. Percentage fractional shortening of the left ventricle was increased in eight patients. Since MVP predisposes to cardiac arrhythmias which are poorly tolerated in the setting of ventricular hypertrophy and reduced left ventricular compliance, the recognition of this combination of MVP and ASH is of clinical importance.

MITRAL VALVE PROLAPSE is a common entity. Although the majority of patients with mitral valve prolapse have a benign clinical course, some subjects experience serious complications such as sudden death, infective endo-

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