Fibrolipoma of the Mitral Valve in A Child

Clinical and Echocardiographic Features

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SUMMARY Echocardiographic examination, performed in a 12-year-old boy who had signs of mitral regurgitation, showed the presence of an abnormal mass of echoes in the left atrium and mitral orifice. These were initially interpreted as representing an atrial myxoma. Surgical exploration showed that the tumor originated from the posterior leaflet of the mitral valve, and, microscopically, the lesion was diagnosed as a fibrolipoma.

The clinical and echocardiographic features of this unique type of cardiac tumor are discussed.

PRIMARY TUMORS OF THE HEART are rare in infancy. The most frequent type is probably the rhabdomyoma which, in most cases, is associated with tuberous sclerosis. Few reports have appeared in the literature regarding primary tumors of heart valves in children.

This report describes the clinical, echocardiographic and anatomical-pathological features of a fibrolipoma of the posterior leaflet of the mitral valve (PML) in an asymptomatic 12-year-old boy.

Case Report

The patient is a 12-year-old male who was referred to the pediatric cardiology department in October 1977 for evaluation of a heart murmur discovered at the time of an emergency room visit for a fracture of the right wrist. The child was asymptomatic with respect to the cardiovascular system and there was no history of rheumatic heart disease.

On physical examination, the patient was well-developed and healthy looking. The blood pressure was 90/60. Peripheral pulses were normal and equal in all four limbs. On palpation of the precordium, the apex was displaced to the sixth left intercostal space at the anterior axillary line. On auscultation, the first heart sound was of normal intensity and the second heart sound was normally split. At the apex there was a grade 3/6 soft pansystolic murmur radiating to the axilla. Also at the apex, there was an S₃, followed by a low frequency grade 2/6 diastolic rumble without presystolic accentuation. There was no opening snap. The lungs were clear to auscultation. The liver and the spleen were not enlarged.

The ECG showed signs of left atrial enlargement. On the chest x-ray, the heart size was normal, with a slight prominence of the left ventricle and dilatation of the left atrium. There was prominence of the pulmonary veins in the upper third of the lungs, suggesting the presence of pulmonary venous congestion. Complete blood count, erythrocyte sedimentation rate and protein electrophoresis were all normal.

Echocardiography (fig. 1)

Standard M-Mode echocardiography was obtained with an Ekoline 20A ultrasonicoscope interfaced with a Honeywell strip chart recorder #1856. At the level of the aortic root, an abnormal mass of echoes was recorded during systole in the left atrium. As the transducer was swept toward the left ventricular outflow tract (LVOT) and the mitral valve, abnormal
Fig. 1. The tumor (T) is recorded during systole in the left atrium. At the level of the left ventricular outflow tract the tumor is recorded both during systole and diastole. During systole it cannot be distinguished from the echoes originating from the closed mitral leaflets. The diastolic motion of the anterior leaflet of the mitral valve (AML) is normal, and its echo is clearly separated from the leading edge of the tumor (slanted arrow). The posterior leaflet of the mitral valve cannot be identified. Ao = aorta; IVS = interventricular septum.

Echoes were registered both during diastole behind the anterior leaflet of the mitral valve (AML) and during systole. During diastole these echoes were clearly separated from the AML but could not be distinguished from the echo originating from the PML. The motion of the AML was normal, with an E-F slope of 125 mm/sec and normal pre-systolic reopening following atrial contraction. The motion of the PML could not be identified. The left ventricular cavity was free of abnormal echo but was slightly dilated, as was the left atrium.

Based on the echocardiographic findings, a diagnosis of left atrial myxoma prolapsing into the mitral orifice during diastole was made, and because of the known risk of systemic emboli from these tumors, the patient was operated on a few days after the diagnosis was made. Cardiac catheterization and angiography were not felt to be necessary in view of echocardiographic findings.

During surgery, after opening of the left atrium a multilobulated mass was seen, filling two-thirds of the left atrial cavity. After inspection of the mass it became obvious that the tumor originated from the PML and occupied its lower two-thirds. Extension of the tumor was also seen along the cordae tendineae. It was not possible to separate the tumor from the PML, so the valve was removed and replaced with a Carpentier-Edwards xenograft.

Pathology

The surgical specimen (fig. 2) consisted of both mitral valve leaflets with attached cordae and portions of papillary muscle. The anterior leaflet was normal, grossly and microscopically. The posterior leaflet was considerably deformed and expanded by a bulky, diffuse and lobulated lesion weighing 4 g, with overall measurements of $6 \times 3.5 \times 2.5$ cm. The surface was whitish, smooth and glistening. The cut surface was pale, whitish yellow and rubbery. The lesional tissue involved the cordae and the apex of the papillary muscle.

Microscopically, the bulk of the lesion consisted of well-differentiated adipose tissue. In many areas poorly cellular nonfasciculated connective tissue contributed to the elaboration of the tumor along with a few fibrous-walled vascular channels (fig. 3). In some areas an interstitial fluid which failed to stain as con-
and composed of acellular hyaline fibrous tissue. Four of these patients presented with signs of chronic endocarditis. According to Prichard, these tumors are closely related to the myxomas. The same author cites Forel who reported a case of lymphangiomma of the posterior leaflet of the mitral valve. Cysts of the mitral valve have been described by Leatherman. Estevez reported a lipoma of the medial leaflet of the tricuspid valve in a 19-year-old boy.

Regarding malignant tumors, Forker reports one case of primary sarcoma of the mitral valve which was considered a malignant transformation of a hamartoma, and Muir describes a patient with a chondrosarcomatous mesenchymoma originating in the vicinity of the mitral valve.

Histologic examination of the tumor in our patient revealed a primary benign tumor, but its precise classification was difficult. After taking into consideration diagnosis such as lipoma, hamartoma and mesenchymoma, a diagnosis of fibrolipoma was chosen in our case because of the histologic similarity to lesions of other sites so designated, in particular, soft pedunculated polyps of the skin otherwise known as molluscum pendulum. In these lesions one finds a mixture of mature adipose and connective tissues, the former usually being more abundant.

Patients with primary tumors of the heart usually come to medical attention because of embolic, obstructive or constitutional complications of the tumor. In some cases, the tumor manifests itself by producing episodes of paroxysmal tachycardia. On physical examination, most patients with tumors originating either in the left atrium or on the mitral valve exhibit signs of mitral stenosis and/or regurgitation. Our patient was totally asymptomatic but had signs of mitral regurgitation on physical examination. Although their absence does not rule out the diagnosis, so-called constitutional signs and symptoms such as fever, anemia and acceleration of the sedimentation rate, as well as embolic complications, are usually associated with the myxomas. Since the tumor in our patient was not of the myxomatous variety and its external surface was not friable, the absence of these two types of complications is not unusual. The mitral regurgitation can be explained by the deformation of the PML by the tumor, preventing complete closure of the valve during systole; but in view of the size of the tumor, the absence of significant obstruction is surprising. However, the dilatation of the left atrium and the redistribution of blood flow in the lungs suggest that the mitral orifice was at least partially obstructed.

Many reports have appeared in the literature relating the usefulness of echocardiography in the diagnosis of intracardiac tumors both atrial and ventricular in origin. To our knowledge, no report exists describing the echocardiographic features of a primary tumor of the mitral valve.

The well-recognized echocardiographic features of left atrial myxoma are the presence of a mass of echoes which appear behind the AML a few milli-
onds after its opening and completely fill the space between the AML and the PML throughout the remainder of the diastole. In most cases, the E-F slope of the AML is diminished, the leaflet being held in an opened position by the tumor, although in some cases, the mid-diastolic closure of the leaflet may be normal. With the transducer directed through the LVOT just below the aortic valve, echoes from the tumor are recorded only during diastole, the area anterior to the closed mitral leaflets in systole remaining free of abnormal echoes. As the transducer is swept toward the aortic valve, the tumor may be seen in the left atrium during systole or diastole.

Since the echocardiogram in our patient showed an abnormal mass of echoes visible in the left atrium during systole and appearing behind the AML during diastole, we initially made the diagnosis of a left atrial myxoma. As suggested by others, we felt that the echocardiogram had clearly established the diagnosis, and the patient was sent to surgery without cardiac catheterization and angiography. During exploration of the left side of the heart during surgery, we found that the tumor was located on and originated from the PML rather than from the left atrial cavity.

Postoperatively, close examination of the echocardiogram of our patient revealed that it differed in many respects from the classical description of left atrial myxoma. At the level of the aorta, the tumor was seen in the left atrium during systole, but as the transducer was directed toward the mitral valve, the abnormal echoes were recorded both during diastole and systole, making it impossible to identify the systolic position of the closed mitral leaflets. As the transducer was directed lower in the left ventricle (position 3 of Feigenbaum), abnormal echoes were present only during diastole behind the AML, and the systolic position of the mitral leaflets could then be identified.

A second distinguishing feature is that the echoes from the proximal portion of the AML were clearly separated from the leading edge of the tumor. The diastolic motion of the AML was entirely normal and an echo-free space was always present between its echo and the leading edge of the tumor during early diastole. The echo from the PML could not be separated from the tumor during systole or diastole.

Although these echocardiographic features are easily explained in our case by the localization of the tumor on the PML, they are not pathognomonic, since a similar echocardiographic picture could be obtained from a myxoma attached to the lower part of the posterior left atrial wall. Their presence should, however, raise the suspicion that this is not the usual form of left atrial tumor, and the surgeon should be alerted of the possible need for mitral valve replacement.

References

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