

Hamartoma of the Mitral Valve with Blood Cysts: A Rare Tumor Detected by Echocardiography

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A 16-year-old boy was submitted to a cardiac examination after an episode of faintness. A transthoracic echocardiogram was performed, which revealed a very mobile multicystic tumor attached to the mitral valve. A small solid mass adherent to the cysts was better defined by transesophageal echocardiography. The patient was submitted to cardiac surgery consisting of tumor resection and a mitral prosthesis implant. The surgery was successful. The tumor consisted of three bright red tense cysts with hematic content, each 1 to 2

cm in diameter. The cysts were coalescent and adherent to a solid mass attached to the posterior papillary muscle head. Histopathologic examination revealed a hamartoma with a cystic part composed of the proliferation of myofibroblast cells in a stroma with vessels, collagen, and elastin fibers. Valvular hamartoma with blood cysts is a very rare cardiac tumor both for its histopathology and its localization. (*J Am Soc Echocardiogr* 1998;11:832-6.)

Cardiac tumors are not frequent. Primary cardiac tumors are only seen in 1 of 10,000 routine autopsies and are most often histologically benign.¹ Cardiac involvement by secondary growths or direct extension is found 10 to 20 times more often.

Although cardiac tumors are mostly asymptomatic, we must bear in mind the importance of emergent diagnosis because sudden death may be the first manifestation of this disease.²

Echocardiography, presently a routine procedure, is part of most check-up studies. This technique made the diagnosis of cardiac tumors possible, which would otherwise remain unknown.

CASE REPORT

A 16-year-old boy who was almost asymptomatic, except for an episode of faintness (typically vasovagal), was submitted to cardiac examination.

Physical examination showed the patient was of slender build and afebrile with a regular pulse of 60 beats/min and blood pressure at 100/60 mm Hg. Cardiac examination showed no palpable thrill, normal sounds, and a systolic ejection murmur that was audible along the

left sternal border, becoming more intense in the third left intercostal space (grade II/IV). The character of the murmur was not changed by Valsalva maneuver position or handgrip.

Sinus bradycardia and first-degree atrioventricular block were present on the electrocardiogram. A chest radiograph showed a thoracic ratio of 50% with no changes in the pulmonary fields. Ambulatory 24-hour monitoring revealed nothing, with the exception of the electrocardiographic findings. Laboratory values were in the normal range; the sedimentation rate in particular was normal, and there were no eosinophilia. Abdominal echography was normal.

Echocardiographic Findings

The transthoracic echocardiogram (Figures 1 and 2) showed a thin leaflet mitral valve with a very mobile multiple cystic image (43 mm × 18 mm × 23 mm) attached to the anterior leaflet, chordae tendineae cordis, and papillary muscle. A small solid mass stood between the posterior papillary muscle and the cystic image with involvement of the muscle head. Despite its dimension and mobility, this tumor apparently did not interfere with mitral valve motion, causing no significant obstruction or regurgitation. In addition, it did not cause obstruction of left ventricular outflow. The other valvular structures were normal. Cardiac cavity dimensions were not increased, and left ventricular function was normal.

A transesophageal echocardiogram was also performed (Figure 3), defining more precisely the small solid mass adherent to the cystic structures and to the head of the posterior papillary muscle.

After echocardiographic diagnosis of the cystic tumor was made, serology for hydatid cyst was performed and

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Figure 1 Transthoracic echocardiogram: Parasternal long-axis view. Cystic tumor is seen attached to anterior leaflet of mitral valve. In this incidence, length of tumor (longest dimension) is 43 mm. Cyst height is 18 mm. Left atria and left ventricle are not dilated.

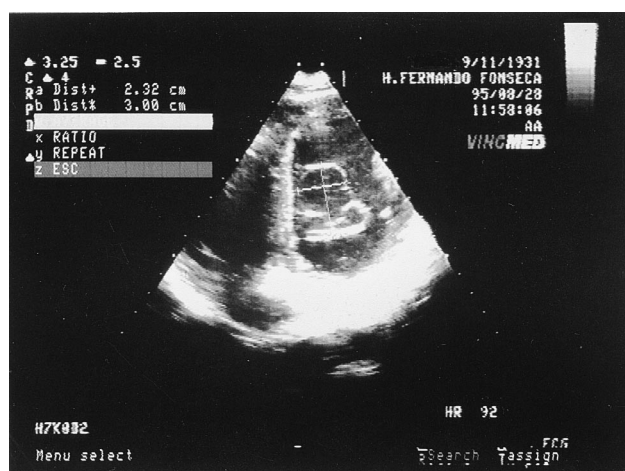


Figure 2 Transthoracic echocardiogram: Apical four-chamber view (angulated). Tumor consists of three cysts, the largest measuring 23 mm in width. This view does not show longest dimension of tumor.

proved negative. When questioned, the patient revealed close contact with dogs during childhood.

Magnetic resonance imaging did not provide any additional information for the diagnosis.

Operative Findings

On operation it was not possible to dissect the cyst, therefore the valve was resected and a number 27 St. Jude mitral prosthesis implanted. The excised tumor (Figure 4) consisted of three bright red tense cysts, each ± 1 to 2 cm in diameter. The cysts, of hematic content, were coalescent and linked to the head of the posterior papillary muscle by the solid mass, which occupied the whole extension of the posterior commissure of the valve.

A histopathologic examination of the valve showed a partial cystic hamartomatous lesion composed of myofibro-

blastic cell proliferation in a stroma with vessels, collagen, and elastin fibers. There was no adipose tissue.

The patient had an uneventful postoperative recovery and remained asymptomatic in a 12-month follow-up.

DISCUSSION

Primary cardiac tumors are rare, especially in youths. In particular, heart valve tumors are very rare conditions, representing less than 10% of all cardiac neoplasms. Each of the four valves is involved with approximately the same frequency.² Most valvular tumors are incidental findings at autopsy; however, there has been a recent increase in clinical diagnosis^{3,4}

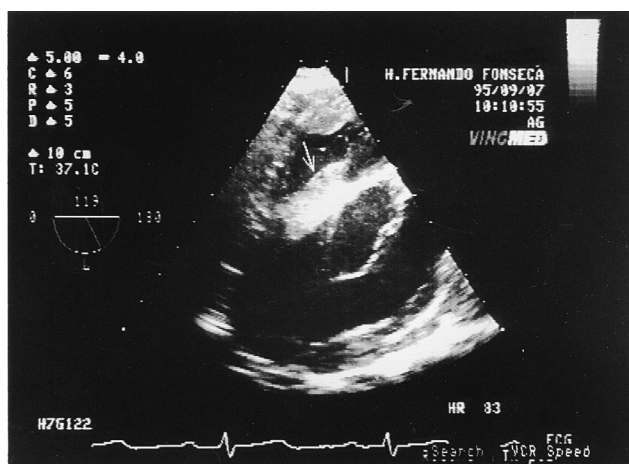


Figure 3 Multiplane transesophageal echocardiogram at 120 degrees. Very echogenic solid mass is seen adherent to cystic structure and attached to posterior papillary muscle head.

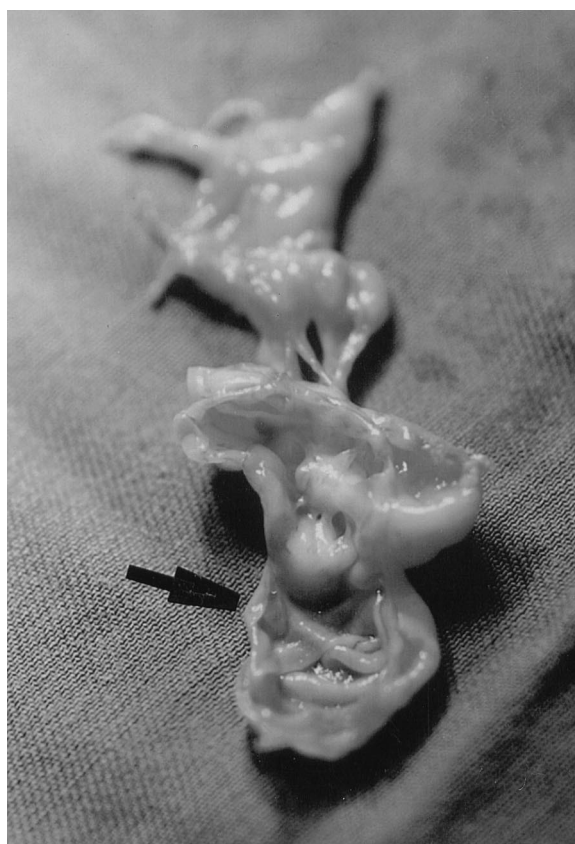


Figure 4 Pathoanatomic piece. Macroscopic aspect of tumor with ruptured cysts (*arrow*) after resection.

because of a greater use of diagnostic techniques, especially echocardiography. Compared with extra-valvular tumors, they are more likely to be benign and asymptomatic. Tumors of the mitral valve are mostly asymptomatic; however, they may produce symp-

toms such as congestive heart failure, neurological manifestations, and sudden death, in decreasing order of frequency.²

Primary cardiac valve tumors are less likely to produce serious complications compared with nonvalvu-

lar tumors,² nevertheless, because of their potential to interfere with valve motion and to release tumor fragments or fibrin clots formed on the tumor surface, they have been associated with sudden death, embolic events, and heart failure.⁴⁻⁶ This potential for life-threatening complications indicates that cardiac valve tumors, even those that are histologically benign and do not cause metastatic or erosive disease, should be managed surgically. Therefore, once the diagnosis of valvular tumor is made, prompt surgical resection is advisable. This is particularly true given the ability to repair rather than replace many affected valves.

In the large series of Edwards et al.,² 92.8% of the tumors were benign, with 41 papillary fibroelastomas, 5 myxomas, 4 fibromas, 1 hemangioma, and 1 hamartoma. The hamartoma in these series was located in the mitral valve and manifested by congestive heart failure.

Besides these neoplastic tumors, we must bear in mind inflammatory or infectious and congenital tumors.

In our case, the cystic echocardiographic characteristic of the mass restricts our diagnostic range because most of the cardiac tumors are solid.

Faced with the cystic nature of the tumor, we considered the hypothesis of a hydatid cyst, hemangioma, or hematogenic cyst of the mitral valve.

Cardiac involvement of hydatidosis is uncommon (0.5% to 2%)⁷ if compared with hepatic (65%) or pulmonary localization (25%).⁸ Cardiac presentation is usually associated with the involvement of other organs. We excluded hepatic and pulmonary involvement in our patient.

Chest pain is the most common manifestation of cardiac hydatidosis; however, other symptoms and manifestations such as arrhythmias and sudden cardiac arrest have also been observed.⁸ In our case, the lipotimia could have been provoked by the interference of the mass with mitral valve closure, although the character of the murmur did not change with maneuvers and we could not demonstrate any interference by echocardiography nor detect any arrhythmia or conduction defect.

We seriously considered the hypothesis of a hydatid cyst, which was favored by patient contact with dogs and the fact that hydatid cysts are endemic in some areas in the south of Portugal, where the patient lived. Although the serology for hydatid cyst was negative, this does not exclude hydatidosis because hydatid serology sensitivity and specificity is not 100%.^{8,9} Serologic tests (latex agglutination and immunoelectrophoresis) are positive in only 60% of pulmonary and 90% of hepatic lesions. Eosinophilia

were not present, although this is not a constant in hydatidosis.

In addition, hydatid cysts are more frequently intramyocardial, located in the interventricular septum, followed by the posterior wall of the left ventricle and the right ventricular free wall in decreasing order of frequency.¹⁰ One case was described in the papillary muscle, and our case had the solid mass related to the posterior papillary muscle head, which could be an inflammatory reaction to the cyst. Valvular locations have not been described because the parasitic agent arrives from the blood stream, through the coronary circulation. In our case it could have developed from the papillary muscle circulation.

In most of the patients with cardiac echinococcosis, the bidimensional echo showed images of spheroidal cystic masses. These are usually single, intramyocardium-located cysts affecting mostly the intraventricular septum. The average size is 3.5 cm in diameter, and they usually protrude into one or both ventricular chambers without producing any symptoms of mechanical interference. Nevertheless, the echocardiographic patterns may differ from this typical pattern in many patients. Cysts may be multiple instead of single, as in our case.¹⁰

The tumor in this case could otherwise correspond to hemangiomatous malformations. Although infrequent¹¹ and most being incidental autopsy findings,¹² hemangiomatous malformations of the heart correspond to blood-filled cysts of the heart valves, which were first reported in 1844 by Boyd and El-sasser.¹³ Small blood-filled cysts of cardiac valves are normally found in infants at necropsy but are rare after the first year of life. They are usually seen as small, rounded multiple nodules on the atrial surfaces of the atrioventricular valves and occur less often on the ventricular surfaces of the semilunar valves. They are believed to be cystic dilations at the end of canals lined with endothelium in the substance of the valves. Single, large-sized valvular blood cysts occurring in older children and adults have seldom been reported; there are, however, a few reports in adults.

Independent of the nature of the tumor, which was uncertain, we decided to refer the patient to surgery. We agree with Pasaoglu et al.¹⁴ in that all the cystic tumors of the heart must be excised, specially in a valvular location because the precise diagnosis can be made only by intraoperative examination.² In this case the surgical indication was easy to establish because it was a valvular cystic tumor of unknown origin, the mass was large, and the patient had previously had lipotimia, which had not been completely clarified and could be related to this mass.

Finally, histopathologic examination of the solid

mass taken from our patient revealed a hamartoma with a hematogenic cyst of the mitral valve, an extremely rare diagnosis with a very unusual location. On reviewing the literature, we found only one case of hamartoma of the mitral valve,² without any associated cyst. Hamartomas are benign tumors¹⁵; however, because of all the possible complications already described, of which some are life-threatening, surgical resection of the tumor was obligatory.

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REFERENCES

1. Nadas HS, Ellison RC. Cardiac tumors in infancy. *Am J Cardiol* 1968;21:363-66.
2. Edwards FH, Hale D, Cohen A, Thompson L, Pezella T, Vimani R. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127-31.
3. Shub C, Tajik AJ, Seward JB, et al. Cardiac papillary fibroelastomas: two-dimensional echocardiographic recognition. *Mayo Clin Proc* 1981;56:629-33.
4. Topol EJ, Biem RO, Reitz BH. Cardiac papillary fibroelastoma and stroke: echocardiographic diagnosis and guide to excision. *Am J Med* 1986;80:129-32.
5. Kasarkis EJ, O'Connor W, Earle G. Embolic stroke from cardiac papillary fibroelastomas. *Stroke* 1988;19:1171-3.
6. Chitwood WR. Cardiac neoplasms: current diagnosis, pathology and therapy. *J Cardiac Surg* 1988;3:119-54.
7. Dighiero J, Canabal EJ, Aguirre CV, Hazan J, Horjales JO. Echinococcus disease of the heart. *Circulation* 1958;17:127-32.
8. Miralles A, Bracamonte L, Pavie A, Bires V, Rabago G, Grandjbakhch I, Cabral C. Cardiac echinococcosis: surgical treatment and results. *J Thorac Cardiovasc Surg* 1994;107:184-90.
9. Munoz C, Portus M. Estudio comparativo de diversas tecnicas serologicas para el diagnostico de laboratorio de la hidatidosis. *Rev Iber Parasitol* 1982;(suppl):345-58.
10. Oliver JM, Sotillo JF, Dominguez FJ, et al. Two-dimensional echocardiographic features of echinococcosis of the heart and great blood vessels. *Circulation* 1988;78:327-37.
11. McAllister HA, Fenoglio JJ. Tumors of the cardiovascular system, *Atlas of Tumor Pathology*, vol 15. Washington, DC: Armed Forces Institute of Pathology, 1978.
12. Reece IJ, Cooley DA, Frazier OH, Hallman GL, Pomens PL, Montero CG. Cardiac tumors: clinical spectrum and prognosis of lesions other than classical benign myxoma in 20 patients. *J Thorac Cardiovasc Surg* 1984;88:439-46.
13. Boyd TAB, Elsasser GS. Blood cysts on the heart valves of newborn infants. *Arch Pathol* 1949;25:757-9.
14. Pasaoglu I, Dogan R, Nazli N, Gungen Y, Bozer AY. Blood cyst originating from tricuspid septal leaflet. *J Cardiovasc Surg* 1991;32:589-91.
15. Becker AE. Tumors of the heart and pericardium. In: Fletcher CDM, editor. *Diagnostic histopathology of tumors*. Edinburgh: Churchill Livingstone, 1995:7-41.

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