Cardiac Hamartoma. Case Report and Literature Review

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INTRODUCTION

Primary cardiac tumors are rare, with a post mortem incidence of 0.001%-0.028%. The symptoms associated with them are very non-specific; electrocardiogram findings and the results of physical examinations and chest x-rays are usually inconclusive. Imaging techniques therefore play an important role in the detection and differential diagnosis of cardiac masses.

CASE REPORT

The patient, who practiced sport habitually, was a 33 year-old man who came for consultation regarding palpitations and exercise-induced dyspnea—symptoms he had noticed for 3 months. A physical examination and chest x-ray were both normal. An electrocardiogram (ECG) showed deep, negative T waves in II, III, aVF, V5, and V6; these had been noticed in an ECG some 8 years earlier. At that time the patient was studied for precordial pain; an arrhythmic cause was ruled out by Holter-ECG. However, echocardiography was not performed. Transthoracic echocardiography (TTE) (Figure 1) now showed a homogeneous intracardiac mass of 45×55 mm on the posterolateral wall of the left ventricle. The echodensity of this mass was similar to that of the myocardium. Although the mass protruded into the cavity, left ventricular systolic function was not compromised. The patient’s biochemical, hemogram, and coagulation results were normal. Tumor markers were negative. Holter-ECG detected isolated ventricular and atrial extrasystoles; no tachyarrhythmia was recorded.

Magnetic resonance (Figure 2) confirmed the presence of an intramural mass with no evidence of myocardial or pericardial infiltration. The mass was hyperintense during the STIR sequence, mildly hyperintense in T1, and isointense or minimally hypointense in T2. Early, heterogeneous highlighting was seen after the injection of gadolinium.


Palabras clave: Miocardiopatía. Imagen. Diagnóstico.
Ventriculography showed a filling defect in the lower part of the ventricle. Coronary angiography showed irrigation of the mass via the obtuse marginal branch and the right coronary artery. Thoracoabdominal computed tomography detected neither adenopathy nor other masses.

A benign heart tumor interfering with left ventricular filling was suspected and surgery scheduled. Following left ventriculotomy, the mass was partially resected and the posteromedial papillary muscle reimplanted.

Anatomopathological examination (Figure 3) showed an irregular distribution of histological elements, including disordered, hypertrophied muscle fibers containing myocytes with large nuclei, surrounded by fibrous and adipose tissue. The final diagnosis was a hamartoma composed of mature cardiomyocytes.

Following surgery, Holter-ECG detected a very weak ventricular extrasystole. Ergometry was negative (patient score 11 METS). The patient followed a cardiac rehabilitation program and has been asymptomatic and has suffered no post-operative problems for three years.

DISCUSSION

The low cost and innocuous nature of echocardiography make it the technique of choice in initial studies of heart masses. The technique can localize such masses, define their shape, size, mobility, and point of anchorage, and determine whether they are solid or cystic in nature.

Magnetic resonance supplies structural and hemodynamic information since it offers both static and dynamic sequences, and images can be acquired from an unlimited number of planes and projections. Tissue characterization via images potentiated in T1 and T2 complete this information. Slightly increased intensity in T1 is seen in tissues with fibrous or muscular contents, but never where there is adipose tissue. Hypointensity in T2 rules out that the mass contains liquid. These tissue characteristics, plus the absence of infiltration, first suggested the present mass might be a fibroma. However, the images obtained after gadolinium injection showed good vascularization. This suggested it not to be a fibroma since these tumors have low metabolic requirements; at this point it was be-
lieved the mass might be a hemangioma or a malignan
tumor. Coronary angiography was performed to
define the vascular supply and to help indicate the sur-
tical technique to follow. Coronary angiography also
supplies data on the presence of obstructive arterial
disease, and on the vascular malformations seen in
rare cases of intramyocardial dissecting hematoma.2,4
It can also be used to ratify a diagnosis of heman-
gioma.3

A hamartoma is a benign overgrowth of the mature,
differentiated cells of the organ in which it is found.
However, these cells are disorganized. The mass re-
results from the anomalous development of embryonic
cells.

Focal hypertrophic cardiomyopathy (FHC) and
rhabdomyoma also involve hypertrophied myocytes,
but with different histological and pathogenic charac-
teristics. A family background and a preference for the
septal region tend to be associated with FHC, whereas
presentation during infancy and an association with
Bourneville’s disease are related to rhabdomyoma.
The definitive diagnosis is anatomopathological.

The first descriptions of hamartomas composed of
mature heart cells appeared in 1998.6,7 The intracardiac
location of such tumors has been reported in small
groups of patients.6-13 An essential characteristic of
cardiac hamartomas is the presence of hypertrophied myocytes lying in a disordered fashion and mixed with
vascular, fibrous and fatty tissue in different propor-
tions. This varied histological presentation has led to a
certain nomenclatural confusion. Hamartomas can ap-
ppear on their own, but on rare occasions can be multi-
ple. They preferably develop on the ventricle wall
but have been reported on valve tissue.9

One of the most common manifestations of these tu-
mors is ventricular tachycardia in children and young
people10-13; a case of a 2 year-old was reported in this
journal.13 Treatment is surgical and good short-term
results are usually achieved. The present patient no-
ticed palpitations; ventricular extrasystoles were de-
tected but no tachycardia was recorded.

The presentation of hamartomas in young adults,
and their slow growth, suggests a possible congenital
origin with hypertrophy and development of the mass
in the first years of life. In the present case, the recor-
ding of electrical anomalies 8 years before the current
events suggests this possibility. Other authors have
described congenital fibrous cardiac tumors (not asso-
ciated with complex hereditary problems) in infancy,
adolescence and early adulthood.2

REFERENCES

med Forces Institute of Pathology; 1996. p. 231.
2. Burke AP, Rosado-de-Christenson M, Templeton PA. Cardiac fi-
broma: clinicopathologic correlates and surgical treatment. J Tho-
3. Piliam MB, Sternlieb JJ. Intramyocardial dissecting hematoma:
an unusual form of subacute rupture. J Cardiovasc Surg. 1993;8:628-
37.
4. Bapat VN, Naik AM, Lokhandwala Y, Tendolkar AG. Intramy-
5. Geiser EA. Hemangioma of the heart. Clin Cardiol. 1998;21:
292-4.
6. Burke AP, Ribe JK, Bajaj AK. Hamartoma of mature cardiac
7. Sturtz CL, Abt AB, Leuenberger VA, Damiano R. Hamartoma of
mature cardiac myocytes: a case report. Mod Pathol. 1998;11:
496-9.
8. Tanimura A, Kato M, Morimatsu M. Cardiac hamartoma: a case
al. Hamartoma of the mitral valve with blood cysts: a rare tumor
10. Greenberg HM, Aretz HT. Case 31-1999: a 33-year-old man with
wide complex tachycardia and left ventricular mass. N Engl J
11. Minh HT, Dinh BA, Galvin JM. Left ventricular hamartoma asso-
JL, et al. Incessant ventricular tachycardia in infants: myocardial
hamartomas and surgical cure. J Am Coll Cardiol. 1987;10:619-
26.
Taquicardia ventricular incesante y hamartomas miocárdicos en
la infancia: remisión a largo plazo tras resección quirúrgica. Rev