Recurrent Cardiac Fibroelastoma. Is It Really a Benign Tumor?

Fibroelastoma papilar recurrente. ¿Es realmente un tumor benigno?

To the Editor,

Papillary fibroelastomas are the second most common cardiac tumor. These tumors are benign and typically occur in the valvular endocardium, and most present as solitary masses. Only a small percentage cause symptoms. Recurrence of these tumors is exceptional—until now it has not been described in the literature—and the treatment for such cases is unclear.

We present the case of a 32-year-old man who presented with a stroke. Transesophageal echocardiography showed a mobile mass attached to the mitral valve causing mild regurgitation, with no other abnormalities. Three-dimensional transesophageal echocardiography (3D-TEE) confirmed the presence of a 6 × 5 mm round tumor, with a friable appearance, attached to the atrial aspect of the mitral valve, at the free edge of the posterior leaflet (P1), suspicious of papillary fibroelastoma (Figure 1). In addition, electrocardiography showed negative T-waves in the inferior leads, and consequently magnetic resonance was performed, which showed a small midsegment-basal subendocardial infarct of the posterior septum and inferior wall (Figure 1).

With this suspected embolic pedipillary fibroelastoma, the patient underwent cardiac surgery, in which the mass was resected, with valvuloplasty using a pericardial patch. The surgical result was good, with no residual regurgitation. Pathology showed papillary fronds of connective tissue with few cells, covered with a single layer of endothelial cells, findings characteristic of a papillary fibroelastoma (Figure 1).

At follow-up, the patient remained asymptomatic, and transesophageal echocardiograms performed every 6 months showed no abnormalities.

At 2 years postintervention, the patient had sudden-onset low back pain, and renal embolism was diagnosed. Repeat 3D-ETT was performed, which showed a new 4 × 4 mm tumor, with similar characteristics, this time located on the atrial aspect of the anterior leaflet of the mitral valve (A1), just in front of the location of the previous mass (Figure 2).

Given this rapid tumor recurrence and the aggressive presentation on both occasions (stroke, myocardial infarction, and renal embolism), it was decided to perform mechanical mitral valve replacement (Figure 2). Pathology again confirmed that this was a fibroelastoma.

After myxoma, papillary fibroelastoma is the second most common benign cardiac tumor. These tumors originate in the valvular endocardium, mainly in the aortic and mitral valves, although cases have also been described of attachment to the ventricular walls. They usually present as solitary masses or, rarely,
as multiple lesions. They have a characteristic gross appearance, with multiple papillae, resembling a sea anemone. Histologically they are distinguished by a central core of dense connective tissue surrounded by a layer of hyperplastic endocardial cells.

Most patients are asymptomatic and an incidental diagnosis is made on echocardiography, during surgery, or at autopsy. A small percentage of patients have severe embolic symptoms, such as myocardial infarction, stroke, or peripheral embolism.

The initial diagnosis is made by ultrasound. Currently, 3D echocardiography offers better visualization and understanding of the relationship between cardiac structures than 2D imaging, and allows a more precise diagnosis of this type of tumor.

Surgical intervention is recommended for patients who have had an embolic event, or those with large (> 1 cm) or very mobile tumors. In asymptomatic individuals with small, less-mobile tumors, the therapeutic approach is debated, but tends to consist of conservative treatment.

In the long-term, the prognosis of these tumors is good, and recurrence after surgical resection has not been described in the literature until now.

We present the first case of recurrent papillary fibroelastoma in the mitral valve. Of note, this recurrence involved very rapid tumor growth, as it was not present on a follow-up echocardiogram performed 5 months previously.

Although a very wide resection of the affected area was performed in the first operation, this was insufficient to prevent the development of a new tumor.

Given that the tumor recurred in the contralateral valve leaflet, right in the area of coaptation of the 2 leaflets, it may be attributed to the continual rubbing that both leaflets are subject to during valve closure, or it may be that we are dealing with a more diffuse involvement of the whole valve.

Although this tumor was histologically benign, its malignant biological behavior with a rapid relapse made it a more aggressive tumor. Therefore, although valve repair would have been technically feasible, we opted for mechanical valve replacement.

Belén Díaz-Antón, Francisco Javier Parra Jiménez, Gregorio Cuerpo Caballero, Francisco Pérez Rodríguez, and Jorge Solís Martín

Servicio de Cardiología, Hospital Universitario Madrid Montepríncipe, Boadilla del Monte, Madrid, Spain
Servicio de Cirugía Cardíaca, Hospital Universitario Madrid Montepríncipe, Boadilla del Monte, Madrid, Spain
Servicio de Anatomía Patológica, Hospital Universitario Madrid Montepríncipe, Boadilla del Monte, Madrid, Spain

*Corresponding author: E-mail address: b.diazanton@gmail.com (B. Díaz-Antón).
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Complexity Levels in Patients With Heart Failure Diagnostic Codes in a Population

Niveles de complejidad asistencial en pacientes con códigos diagnósticos de insuficiencia cardíaca en una población

To the Editor,

Heart failure (HF) is a public health issue due to its increasingly high incidence and prevalence, high morbidity and mortality, and the significant associated resource use.1 Although there are several initiatives in Spain that aim to implement HF-specific care models, information is lacking on the distribution and complexity of these patients and, therefore, their classification according to their theoretical care needs. One of the systems used for this purpose is known as the Kaiser pyramid, developed in the United States.2 This allows classification of patients into risk categories and the intensity of recommended health interventions. The base of the pyramid represents healthy individuals and those that are acutely unwell. In the middle segments are those with a chronic disease for whom health care efforts should be directed at disease management. The top level represents complex chronic patients, for whom health care efforts should focus on individual case management.2

The aim of this study was to classify the entire population with a diagnosis of HF in the Valencian Community in Spain according to the Kaiser pyramid categories.

This was a cross-sectional descriptive study of patients with a diagnosis of HF in the Valencian Community in 2015, selected from a population of 4 709 115 patients with a health card and registered with a named physician. HF was defined using the following codes from the International Classification of Diseases 9th Revision, Clinical Modification: 398.91, 402.01, 402.11, 402.91, 404.01, 404.03, 404.11, 404.13, 404.91, 404.93 and 428.XX.3

The study population was classified according to the 9 health statuses, with up to 6 levels of severity per health status, of the clinical risk groups (CRGs), which are a tool for dividing the population according to disease burden.2 These 9 health statuses were grouped into the 4 Kaiser complexity levels: a) healthy or with acute illness; health statuses 1 and 2 of the CRGs; b) low-complexity chronic disease: health statuses 3, 4 and 5 (severity levels 1, 2 and 3) of the CRGs; c) moderately-complex chronic disease: health statuses 5 (severity levels 4 and 5), 6 (severity levels 1 to 4), 7 (severity levels 1 and 2), 8 (severity levels 1 and 2) and 9 (severity levels 1 and 2) of the CRGs, and d) highly-complex chronic disease: health statuses 5 (severity level 6), 6 (severity levels 5 and 6), 7 (severity levels 3 to 6), 8 (severity levels 3 to 5) and 9 (severity levels 3 to 6) of the CRGs. Further details on each of the CRG categories are provided in the supplementary material. The information was obtained from the Alumbra information platform of the Valencian Community Consellería de Sanitat Universal i Salut Pública (Department of Universal Health and Public Health).

The distribution of the total population by Kaiser pyramid category showed that 50.7%, 34.0%, 13.4% and 1.9% were healthy or had acute illness, had low-complexity chronic disease, moderately-complex chronic disease, and highly-complex chronic disease, respectively (Figure 1).

For HF, the prevalence for the 2015 period was 1.46% (68 846 patients). The distribution of patients with a diagnosis of HF by Kaiser pyramid category is shown in Figure 2. Unlike the observations in the general population, the distribution took the form of an inverted pyramid: most patients with HF had at least moderately-complex disease (52.56%), which was severe in 27 597 patients (40.09%).

The findings presented here highlight that most patients with HF have high care needs due to their complexity. According to the Kaiser pyramid postulates, patients with high-complexity illness need personalized health care management with the dual aim of: a) improving clinical prognosis and patient satisfaction, and b) reducing the use of health care resources.

Figure 1. Kaiser pyramid for the general population.