## **CLINICAL IMAGE**

## Multimodal diagnosis of caseous calcification of the mitral annulus in a patient with suspected left ventricular tumor

Katarzyna Kurnicka<sup>1</sup>, Judyta Samul<sup>1</sup>, Dorota Piotrowska-Kownacka<sup>2</sup>, Małgorzata Wiśniewska<sup>2</sup>, Anna Lipińska<sup>1</sup>, Piotr Pruszczyk<sup>1</sup>

1 Department of Internal Medicine and Cardiology, Medical University of Warsaw, Warsaw, Poland

2 1st Department of Radiology, Medical University of Warsaw, Warsaw, Poland

A 77-year-old woman with hypertension, hypercholesterolemia, and an episode of pulmonary embolism 4 weeks before hospitalization, on oral anticoagulation, was referred to our institution with a suspicion of an intracardiac tumor in the left ventricle. The patient presented with mild exertional dyspnea and a low-degree systolic murmur. Two-dimensional transthoracic echocardiography (TTE) revealed an irregular mass (long-axis view,  $26 \times 17$  mm; 4-chamber view,  $28 \times 20$  mm) attached to the mitral valve (MV) annulus, partially involving the MV posterior leaflet. On TTE, the pathological mass showed a calcified rim and central echolucencies (FIGURE 1A). The echocardiographic features of the mass were not typical for a thrombus or myxoma. Despite the turbulence of mitral inflow caused by the mass, no evidence of mitral stenosis was present (mean gradient, 2 mm Hg; MV area >2.5 cm<sup>2</sup>). Additionally, 2 jets of mild mitral regurgitation were detected (FIGURE 1B). To assess the mass more precisely, its characteristics and their relation to the surrounding structures, 3-dimensional transesophageal echocardiography was performed, revealing a round, highly echogenic mass in the middle region of the mitral posterior leaflet (P2 scallop), with a smooth surface (FIGURE 1C and 1D) and without detectable intratumor blood flow on color Doppler ultrasound.

Cardiac magnetic resonance (CMR) showed a low-signal intensity mass (4-chamber view, 26 mm in length) on  $T_1$ - and  $T_2$ -weighted images (FIGURE 1E-1H), and a caseous calcification of the mitral annulus (CCMA) was suggested. Subsequent cardiac computed tomography revealed major calcifications of the lesion, which confirmed the CMR diagnosis of CCMA (FIGURE 1I and 1J). Because the patient had no significant MV dysfunction, she was excluded from surgery. Importantly, follow-up TTE performed at 6 and 12 months showed no progression of the "tumor".

Mitral annular calcification is a well-known degenerative process and a common finding on TTE, whereas CCMA is a very rare variant of mitral annular calcification. Although mitral annular calcification is associated with older age and chronic kidney disease, the pathophysiology of CCMA has not been established. Interestingly, CCMA may mimic intracardiac tumor or abscess.<sup>1</sup> However, its location in the posterior part of the mitral annulus and typical central echolucency surrounded by a hyperechogenic rim, without any flow and no acoustic shadow, can help in the echocardiographic differential diagnosis. The CCMA image on CMR and CT is highly characteristic and supports the final diagnosis without the need for further histological examination.<sup>2,3</sup>

CCMA is benign and often asymptomatic. However, it may cause significant mitral inflow obstruction or embolic stroke and may be an indication for surgical removal.<sup>4</sup> In patients who underwent surgery, microscopic examinations of caseous material evacuated from a calcified envelope revealed amorphous eosinophilic acellular material, surrounded by macrophages and lymphocytes, with no neoplastic cells.<sup>5</sup> In our patient, the diagnosis of CCMA was based only on typical findings of multimodal noninvasive imaging. It seems that echocardiographic follow-up of this rare phenomenon is sufficient in asymptomatic patients. However, surgery should always be considered in cases of uncertain diagnosis, peripheral embolism, or valve dysfunction.

## Correspondence to:

Katarzyna Kurnicka, MD, PhD, Klinika Chorób Wewnetrznych i Kardiologii, Warszawski Uniwersytet Medyczny, ul. Lindleya 4, 02-005 Warszawa, Poland tel.: +48 22 502 11 44, e-mail: kkurnicka@yahoo.pl Received: March 9, 2017. Revision accepted: April 3, 2017. Published online: April 24, 2017 Conflict of interests: none declared. Pol Arch Intern Med. 2017; 127 (4): 281-282 doi:10.20452/pamw.4004 Copyright by Medycyna Praktyczna, Kraków 2017



**FIGURE 1** Noninvasive imaging of the caseous calcification of the mitral annulus (CCMA); **A** – an irregular echodense mass attached to the mitral annulus, with calcified circumference and echolucent areas in the center (arrow); 2-dimensional transthoracic echocardiography (2D-TTE), 4-chamber view; **B** – 2 jets of mitral regurgitation (arrows); 2D-TTE, 4-chamber view, color Doppler; **C**–**D** – highly echogenic structure localized in the posterior part of the mitral annulus including the middle scallop of the posterior leaflet (arrows); 3-dimensional transesophageal echocardiography; **E**–**H** – cardiac magnetic resonance characteristics of the lesion: **E**, **F** – low signal intensity on T<sub>2</sub>-weighted black blood (**E**) and steady-state free precession sequence (**F**) images in 5-chamber orientation; **G** – enhancement deficit of the lesion presented on the first pass perfusion image in the short-axis view; **H** – limited peripheral enhancement of the mass on delayed contrast-enhancement image in the short-axis view (arrows); **I**–**J** – short axis (**I**) and 5-chamber (**J**) reconstructions of electrocardiogram-gated unenhanced 64-slice computed tomography: high-density lesion with double-layer appearance: lower density core and higher density shell (arrows) confirming the diagnosis of CCMA. Abbreviations: LV, left ventricle; LA, left atrium

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