Journal of Cardiology Cases xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

A rare case of spontaneously dissolved calcification of the mitral annulus: Caseous calcification of the mitral annulus

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ARTICLE INFO

Article history:
Received 14 April 2017
Received in revised form 1 November 2017
Accepted 10 November 2017

Keywords:
Mitral annular calcification
Echocardiography
Caseous calcification of the mitral annulus

ABSTRACT

Caseous calcification of the mitral annulus is a rare variant of mitral annular calcification (MAC). MAC is detected using conventional echocardiography and is prevalent in the elderly. However, limited information is currently available on the transformation of MAC. We herein report a case of a sudden liquified change in MAC, which was diagnosed using echocardiography and computed tomography.
<**Learning objective:** The mechanisms underlying the pathogenesis of caseous calcification of the mitral annulus (CCMA) have not been elucidated in detail. This case report revealed the transition from mitral annular calcification to CCMA, indicating the marked transformation of calcification, which leads to spontaneous liquefaction.>

Introduction

Mitral annular calcification (MAC) was first described in 1908 by Bönninger et al. and was reported to be associated with conduction disturbances [1]. It is a degenerative process involving the fibrous annulus of the mitral valve and is more common in the elderly, particularly in osteoporotic women [2]. The occurrence of MAC is rather associated with atherosclerosis than degenerative changes in the mitral valve [3].

Caseous calcification of the mitral annulus (CCMA) is a rare variant of MAC. The incidence of CCMA diagnosed by echocardiography was previously reported to be 0.068% in all the patients undergoing echocardiography and 0.64% in MAC cases [4]. CCMA resembles a large mass, similar to a tumor or abscess [5]. It has the appearance of a soft and echo-dense mass that is composed of a liquefaction lesion with calcium, fatty acid, and cholesterol admixture at the periannular area of the mitral valve [5].

The mechanisms by which CCMA is organized and formed currently remain unclear. One previous study on CCMA proposed a dynamic process based on the transformation of MAC, with CCMA reverting to MAC in some patients [4]. We herein report a case of a 75-year-old woman who presented with a marked transition to CCMA from MAC, as observed using echocardiography and computed tomography.

Case report

A 75-year-old woman visited the attending hospital with hypertension, dyslipidemia, and implantation of a DDD pacemaker due to complete atrioventricular block.

During her visit to London between December 30th, 2014 and January 2nd, 2015, she was hospitalized due to transient cerebral ischemic attack (TIA). After she had returned to Japan, she visited our hospital on January 5th. At the time of her visit to our hospital, her blood pressure was 140/70 mmHg, and her heart rate was 64 beats per minute (bpm). Consciousness was clear, and there were no other remarkable neurological findings. Chest X-ray revealed no pulmonary congestion and no other significant findings. An electrocardiogram showed a heart rate of 65 bpm

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https://doi.org/10.1016/j.jccase.2017.11.001
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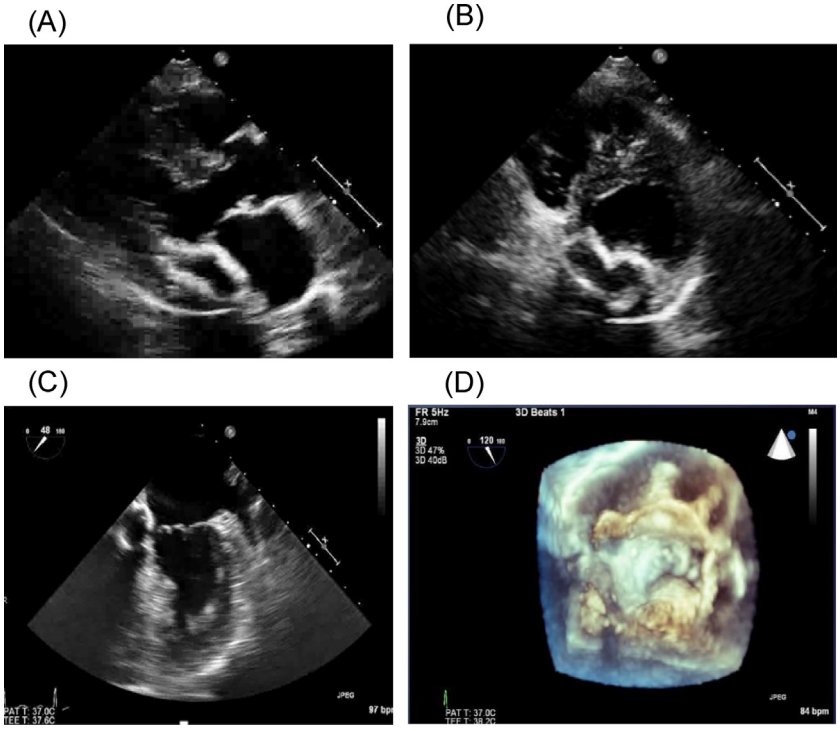

Fig. 1. Transthoracic echocardiography showing a long-axis view (A) and short-axis view (B) at the level of the mitral ring of the echolucent area with encircling calcification. Transesophageal echocardiograph of the mid-esophageal level (C) and a real-time three-dimensional transesophageal echocardiographic image from the left atrium (D). The mass was visualized as a central low echoic area surrounded by calcification at the peri-annular site. The supramitral valve of the left atrium surface was smooth and protruded beyond the mitral annulus level.

and a normal sinus rhythm. Laboratory results were within normal ranges for creatinine, transaminase, and total bilirubin levels as well as blood counts.

Her pacemaker log showed the trace of paroxysmal atrial fibrillation. After returning to Japan, head computed tomography was performed and revealed old cerebellar infarction. Accordingly, she was treated with apixaban (a factor Xa inhibitor).

Echocardiography showed a large mass in the posterior mitral annulus. The mass was soft and moved with the surrounding tissue. Its appearance was echo-lucent in the central area, and a calcified ring encircled the mass (Fig. 1). This echocardiographic finding was different from that of five years previously (Fig. 2). Moreover, computed tomograms revealed a similar change (Fig. 3). The echocardiograms and computed tomograms, which were taken five years before pacemaker implantation, showed markedly different appearance. For these five years, calcification progressed abruptly from MAC to CCMA.

We considered the extirpation of CCMA as a therapeutic strategy. However, CCMA was already widespread around the mitral annulus. If CCMA was removed, reconstructive surgery might have been difficult. After much deliberation, the oral administration of anticoagulants was initiated, and as a result, the cerebrovascular disease never recurred. Careful observation and medication have been continued until now.

Discussion

MAC is characterized by the calcification with chronic fibrosis and degeneration of the ring-like tissue supporting the mitral valve. The prevalence of MAC is 9% and is strongly associated with

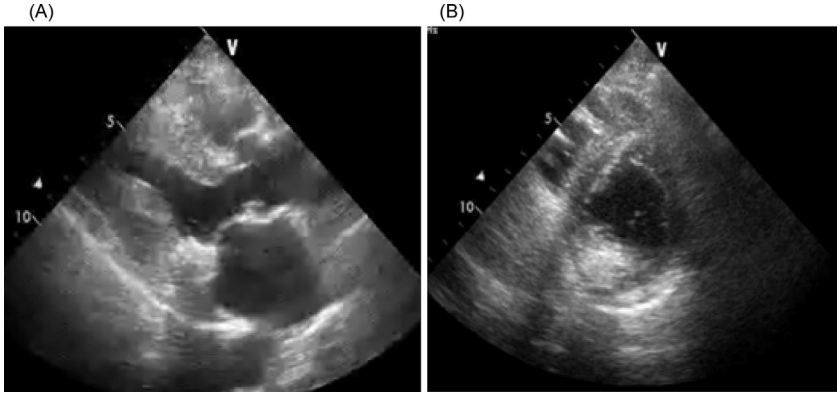

Fig. 2. Transthoracic echocardiography 5 years previously showing a long-axis view (A) and short-axis view (B) at the level of the mitral ring of a bulky calcified homogeneous area with an acoustic shadow under the mass.

Please cite this article in press as: Takeuchi Y, et al. A rare case of spontaneously dissolved calcification of the mitral annulus: Caseous calcification of the mitral annulus. J Cardiol Cases (2017), https://doi.org/10.1016/j.jccase.2017.11.001