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Caseous calcification of the mitral annulus: an uncommon cause of acute mitral regurgitation

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A 63-year-old female patient was being followed-up for a caseous calcification of the mitral annulus (CCMA) associated to a mild mitral stenosis (MS) and mild mitral regurgitation (MR). A multi-modality approach was used to diagnose the CCMA: transoesophageal echocardiography

(TOE)—Panel A, cardiac computed tomography—Panel B, and cardiac mag-



netic resonance—*Panels C* and *D*. She presented to the emergency department with acute cardiogenic shock. A TOE was performed to further elucidate the cause of the cardiogenic shock. TOE showed significant changes in the size and shape of the CCMA as well as an acute severe MR (*Panels E–H*, Supplementary data online, *Videos S1–S3*). The spontaneous rupture and discharge of the caseous mass inside the left ventricle probably led to severe MR which explained the cardiogenic shock. CCMA is a rare form of mitral annular calcification, which can be found in 0.06–0.07% of the population. In CCMA, there is a caseous transformation of the core of a calcified mass involving the mitral annulus, which resembles at autopsy to a toothpaste like material—a mixture of cholesterol, fatty acids, and calcium. The exact mechanism leading to this phenomenon remains unknown. CCMA occurs more often in elderly patients (mean age 69 years), is more frequent in women and is often associated with hypertension. It is usually an incidental finding at the time of TOE and is considered to be benign. However, it may be complicated by systemic embolization, abnormal diastolic flow across the mitral valve, usually mild MS and MR, conduction abnormalities when located close to the atrioventricular conduction system, and unnecessary surgeries when mistaken for tumours or abscesses. CCMA leading to acute severe MR and cardiogenic shock is rare.

Supplementary data are available at European Heart Journal - Cardiovascular Imaging online.

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