

Caseous Necrosis of the Mitral Valve: A Rare Case Report

David Makarious*, Suzanne Eshoo and Peter Fahmy

Department of Cardiology, Blacktown Hospital, Sydney, NSW, Australia

*Corresponding Author: David Makarious, Department of Cardiology, Blacktown Hospital, Sydney, NSW, Australia.

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Abstract

Caseous necrosis of the mitral valve (CNM) is a rare variant of mitral annular calcification (MAC). Diagnosis is made radiologically. Notable risks include fistulisation and arterial embolization. Management options include surgical correction or echocardiographic surveillance, yet no consensus has been reached in the literature.

Keywords: Caseous Necrosis; Mitral Valve; Takutsubo Cardiomyopathy; Mitral Annular Calcification; Stroke

Abbreviations

CNM: Caseous Necrosis of the Mitral Valve; MAC: Mitral Annular Calcification; PML: Posterior Mitral Leaflet

Introduction

Caseous necrosis of the mitral valve (CNM) is a rarely described condition, thought to be a variant of mitral annular calcification (MAC). Whilst the pathogenesis of CNM is unknown, it has been postulated to be associated high serum calcium loads. Comorbidities commonly identified with CNM include hypertension (61.5%), dyslipidemia (23.8%), diabetes mellitus (16.9%) and end-stage renal failure on chronic hemodialysis (11.5%) [1].

Case Report

70-year-old female presented to hospital with chest pain and congestive cardiac failure following a grief event on a background of hyperlipidaemia and hypertension. High sensitivity troponin-T-assay was elevated at 124 ng/L. Electrocardiogram revealed anterior T-wave inversion in V2-V4. A coronary angiogram revealed mild disease. Transthoracic echocardiogram showed hypokinesis of the apical segment and a diagnosis of Takutsubo cardiomyopathy was made. An incidental finding of an immobile echogenic mass situated on the posterior mitral annulus measuring 25 x 28 mm was found (Figure 1). Trans-oesophageal echocardiogram demonstrated a 20 x 15 mm hyper-echoic mass attached to the posterior mitral leaflet (PML), suggestive CNM. Computed tomography findings were in keeping with CNM, revealing a bi-lobe contrast-filling cavity in relation to the PML measuring 29.4 mm in diameter (Figure 2). Given that no functional compromise of the mitral valve was present, a decision was made for on-going surveillance.

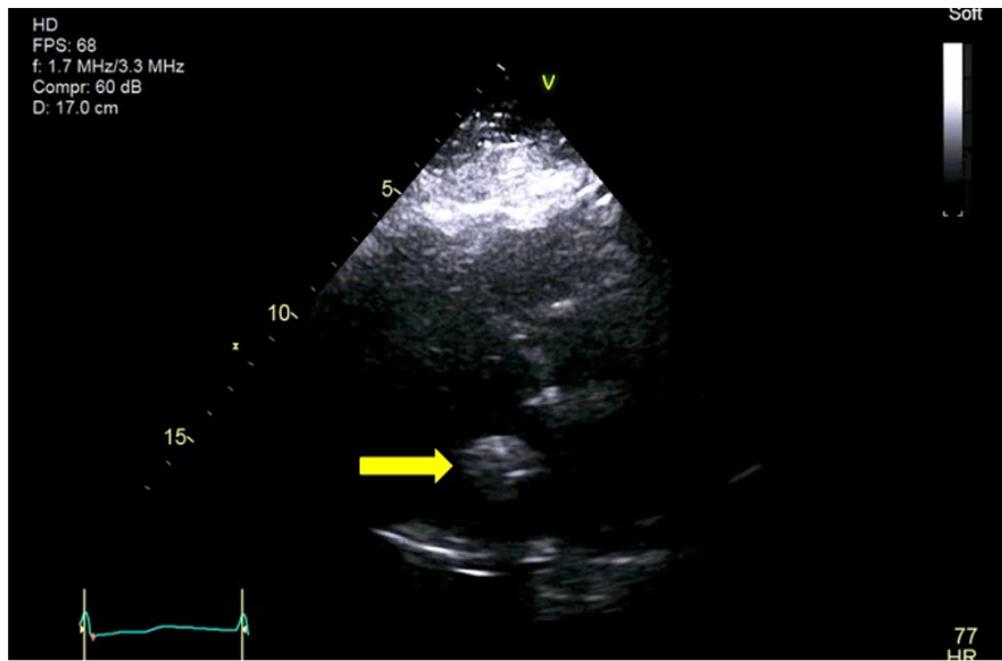


Figure 1: Transthoracic Echocardiogram demonstrating a 25x28mm mass on the posterior leaflet of the mitral valve.



Figure 2: Bi-lobe contrast-filling cavity measuring 29.4 mm on the posterior leaflet of the mitral valve.

Discussion

CNM accounts for 0.63% of diagnosed cases of MAC and carries a general population prevalence of 0.067% [2]. Necroscopy studies revealed a prevalence of CNM of 2.7% in patients with MAC, highlighting a discrepancy in the sensitivity of echocardiography and necroscopy [3]. Under diagnosis via echocardiography when compared to necropsy could be due to the differences in sensitivity or the lack of clinician awareness given the rarity of the condition. As a consequence of the rarity of caseous necrosis, no clear management guidelines/consensus exist.

Current literature suggests that patients with embolic phenomenon or mitral insufficiency, would be candidates for surgical management via valvular replacement or repair. Yet no consensus on the management of asymptomatic patients exist. Currently, most clinicians opt for regular echocardiographic surveillance. Interestingly, a review of the literature by Dietl, *et al.* revealed that 20% of patients with CNM suffered an embolic stroke, with CNM thought to be the precipitant [1]. All reported cases of CNM associated strokes demonstrated an absence of any thrombus formation or other intra-cardiac source of embolization. They postulated that spontaneous fistulisation and embolization of caseous necrotic debris acted as the catalyst for an embolic stroke, negating any benefit of prophylactic anti-coagulation/platelet therapy. Consequently, they suggest elective surgical repair in asymptomatic patients whom are good surgical candidates, in which the risk of surgery is outweighed by the risk of stroke.

Conclusion

In summary, caseous necrosis remains a rare and difficult condition to diagnose and manage due to the lack of high-level evidence to support clinical decisions.

Conflicts of Interest

There are no conflicts of interest.

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