Cardiac calciphylaxis presenting as endocarditis

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A 45-year-old man presented with dyspnoea and fever. His medical history included chronic renal failure requiring long-term haemodialysis. On examination, he was pyrexial, had multiple nail fold splinter haemorrhages, and a mid-systolic murmur. Blood tests revealed white blood count 11.9 × 10^9/L and C-reactive protein 184 mg/L.

Transthoracic and transoesophageal echocardiography was performed (http://www.internationaljournalofcardiology.com/article/S0167-5273(07)01389-7/fulltext-fig1, Panels A–D; LA, left atrium; LV, left ventricle; CV, calcified vegetation, see Supplementary material online, Movies 1–5). This demonstrated a calcified mobile mass in the aortic outflow tract, and another at the apex of the left ventricle. The myocardium appeared unusually bright and speckled with prominent aortic valve and mitral annular calcification. A diagnosis of endocarditis was made.

Blood cultures and serology were negative. Inflammatory markers remained elevated despite multiple courses of antibiotic therapy. The patient developed extensive necrosis of his hands, toes, penis, and buttocks. Plain X-rays revealed vascular calcification in the hands and feet. Two weeks later, the patient died. Post-mortem examination confirmed systemic calciphylaxis with large areas of calcification present within the media of coronary vessels (Panel E, black arrows) and within the myocardium (Panel F, black arrows).

The mobile masses seen on echocardiography were likely to represent healed vegetations that had calcified as a result of calciphylaxis. This condition, better termed ‘calcific uremic arteriopathy’, is characterized by intimal proliferation and endovascular fibrosis with calcification. Subsequent thrombosis leads to tissue infarction and necrosis.

To our knowledge, there have been no previous reports of calciphylaxis involving the heart demonstrated by echocardiography and histology.

Supplementary material is available at European Heart Journal online.