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Effusive constrictive pericarditis in systemic sclerosis

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A 69-year old man presented with a 7-day history of worsening heart failure on a background of systemic sclerosis (SSc). He had a history of pericardial effusion, Raynaud's phenomenon, and achalasia (Figure 1). Examination revealed advanced sclerodactyly, mild hypertension, soft heart sounds with pericardial rub, and fine inspiratory crackles bilaterally. Echocardiography demonstrated a left ventricular ejection fraction of 45% and a large pericardial effusion (Figure 2a). Thoracic computed tomography also showed a thickened and densely calcified pericardium (Figure 2b, Figure 2c), leading to a diagnosis of effusive constrictive pericarditis (ECP). Near total pericardiectomy was performed via median sternotomy without using cardiopulmonary bypass (Figure 2c, long arrow). Histopathological examination with hematoxylin and eosin staining reconfirmed calcific pericarditis with extensive hemorrhagic fibrinous exudate (Figure 2d).

Pericardial involvement in SSc may manifest as acute fibrinous pericarditis, pericardial effusion, pericardial adhesion, or chronic constrictive pericarditis. According to the literature, ECP is a rare manifestation of pericardial involvement in SSc; however, it has been reported to occur in patients even before SSc becomes clinically evident (1). ECP is pathologically characterized by a pericardial effusion developing in the presence of a fibrous and inelastic pericardium. The fibrotic layers of pericardium constrict the myocardium but can also exert significant pressure on the effusive fluid, leading to a cardiac tamponade effect (2).

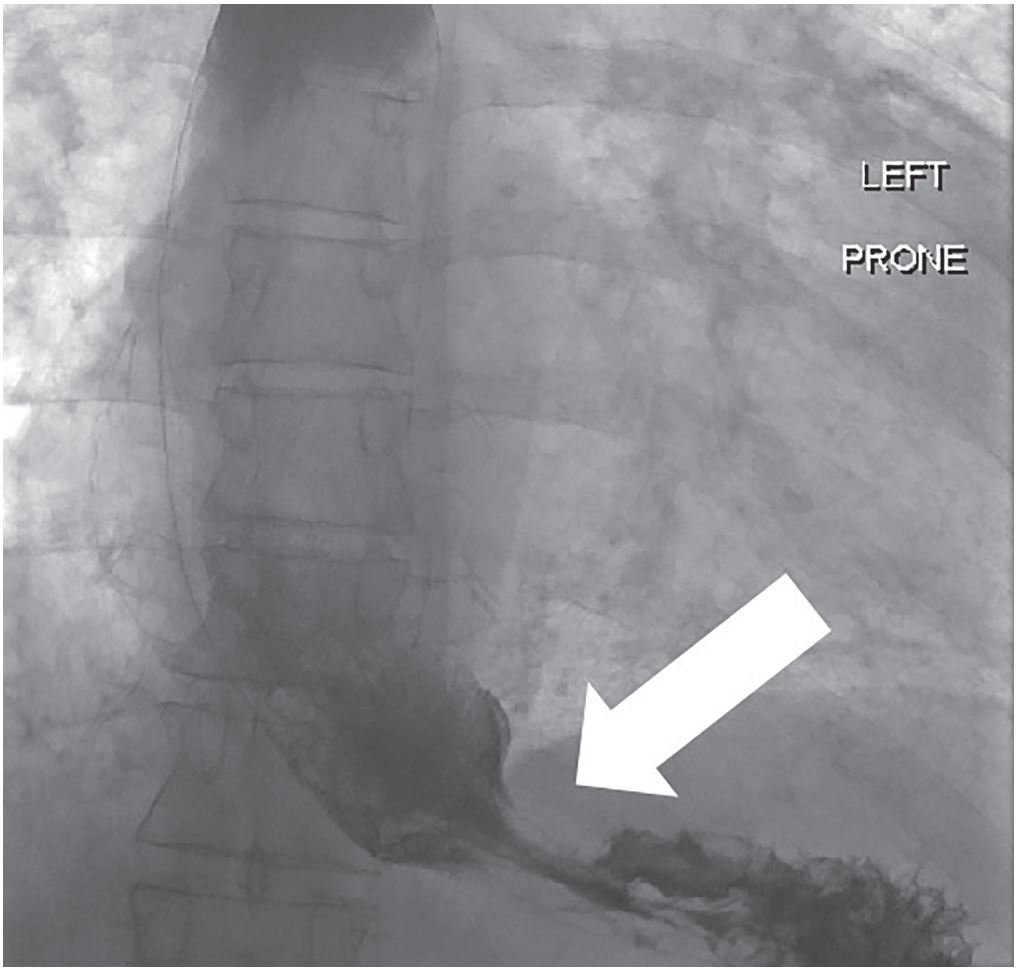

Figure 1. 'Bird Beak' sign of achalasia on barium swallow study.

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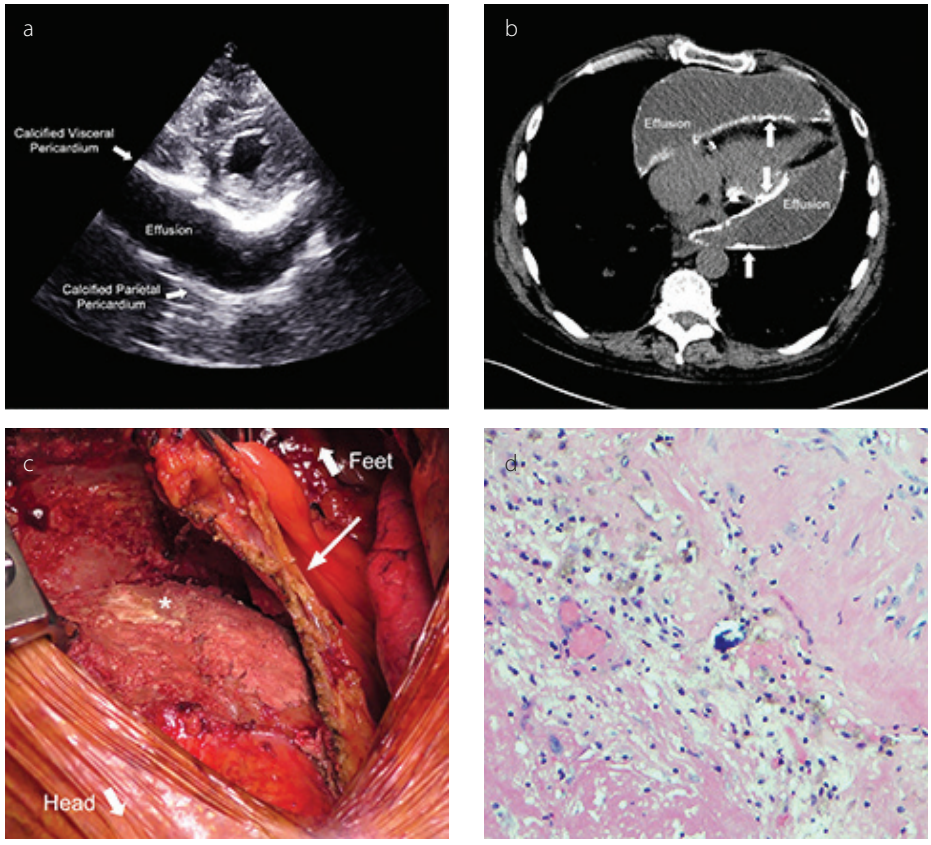

Figure 2. a-d. Calcified pericardial layers (arrows) and effusion on echocardiogram (a), calcified pericardial layers (arrows) and effusion on thoracic computed tomography study (b), pericardiectomy intraoperative image – thickened parietal pericardium (long arrow) and heavily calcified and fibrotic visceral pericardium (asterisks) (c), haemorrhagic fibrinous exudate seen on microscopy with haematoxylin and eosin stain at 40x magnification (d).

Other differentials for the etiology of ECP are commonly idiopathic but also infective, such as tuberculosis, radiation exposure, post-cardiac surgery, and neoplasia. Clinical presentation includes features of pericarditis and right- and left-sided heart failure, varying relative to the rate of onset and the degree of the underlying tamponade effect. Clinical presentation may resemble that of other constrictive pericardial syndromes; however, the distinguishing feature of ECP is persistent elevation of atrial pressure even after drainage of the pericardial effusion (3). Pericardiocentesis has been reported to provide temporary symptomatic relief; however, the underlying fibrous and inelastic pericardium layers will continue to cause diastolic dysfunction until surgically removed. Therefore, pericardiectomy, including the visceral pericardium, is the definitive treatment for ECP (2).

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