A paradoxical rise in the jugular venous pressure on inspiration

Kussmaul's sign in effusive constrictive pericarditis

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Case report

A 67-year old man with mitral valve prolapse and moderate regurgitation was admitted because of dyspnoea, bilateral ankle swelling and hypotension. Close inspection of the jugular veins identified Kussmaul's sign, a typical increase in the central venous pressure during inspiration (fig. 1; arrows). He had no history or clinical evidence of infection, tumours, uraemia, trauma, surgery or radiation. Transthoracic echocardiography revealed moderate diffuse pericardial effusion (PE) (fig. 2, arrows) with paradoxical interventricular septum bounce (see video 1*). Persistence of Kussmaul's sign and symptoms of acute right heart failure after pericardiocentesis (170 ml exudate, no infections and neoplastic cells) prompted the clinical suspicion of idiopathic effusive-constrictive pericarditis. Diagnosis was supported by cardiac magnetic resonance (CMR), showing mild residual PE and diffuse thickening of the pericardium (fig. 3, arrows) with contrast enhancement at the pericardial edges (fig. 4, arrows) and septal bounce (see video 2*). Diagnosis of effusive-constrictive pericarditis was confirmed by typical elevated ventricular filling pressures at cardiac catheterisation (equilibration of ventricular diastolic pressures with dip-plateau waveform) and open surgery (pericardiectomy) showing diffuse parietal (fig. 5A–B) and visceral pericardial thickening (fig. 5C–D). One year follow-up showed complete clinical relief with almost no residual pericardial thickening at CMR.

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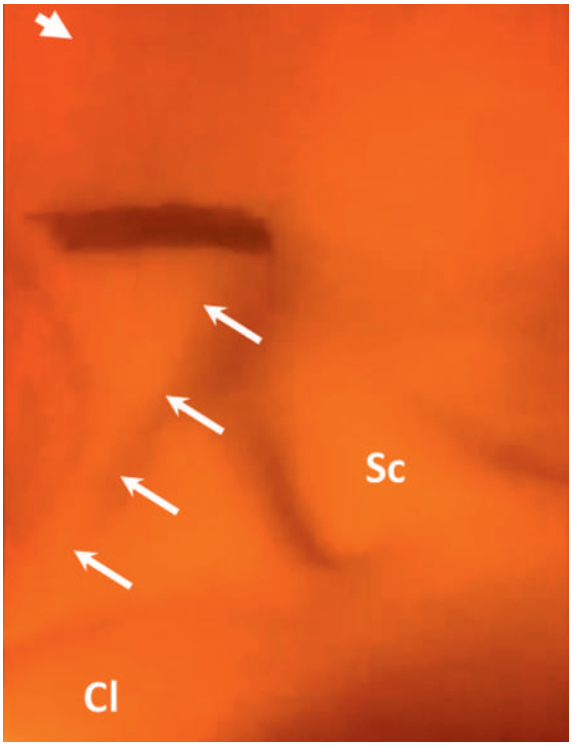


Figure 1: Kussmaul's sign (arrows) is a paradoxical rise in the jugular venous pressure (JVP) (arrows) when the patient breathes in, due to impaired venous flow toward the heart associated with right ventricular constrictive diastolic impairment.
Cl = clavicle; Sc = sternocleidomastoid muscle.

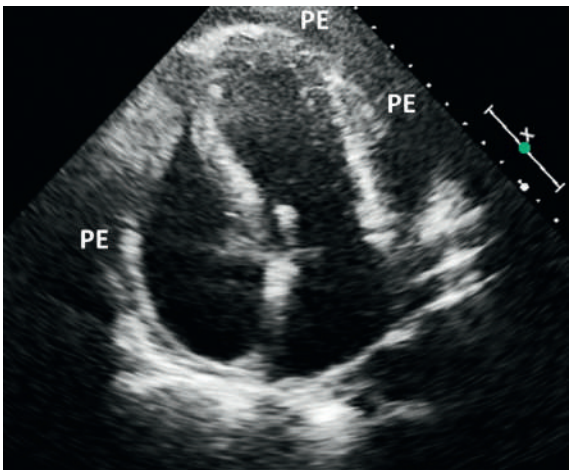


Figure 2: Transthoracic echocardiography; 4 chamber view: it displays a moderate (2 cm) diffuse pericardial effusion (PE), more pronounced on the left side due to partial adhesions. Also ventricular septal bounce due to a paradoxical interventricular septum shift prompted by respiration phases is displayed (see video 1*).

* You can find the videos on http://www.cardiovasc-med.ch/for-readers/multimedia

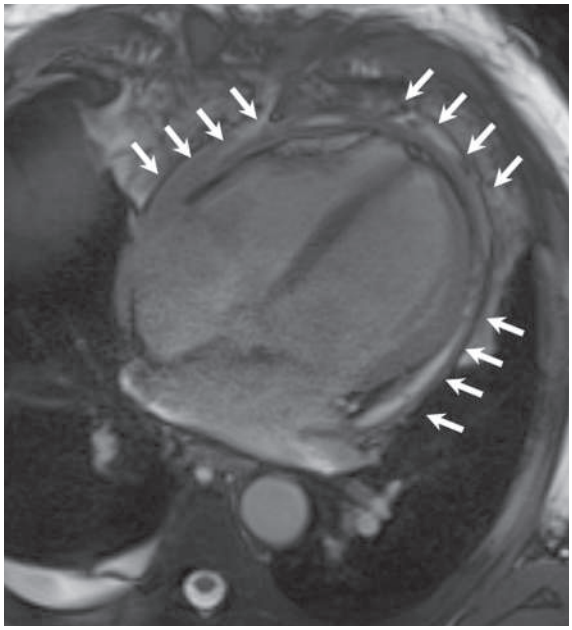


Figure 3: CMR in the 4 chamber orientation showing mild residual pericardial effusion (moderate bright space between pericardial layers), diffuse thickening of pericardial leaflets all around the heart (arrows) and septal bounce (arrows) (see video 2*).

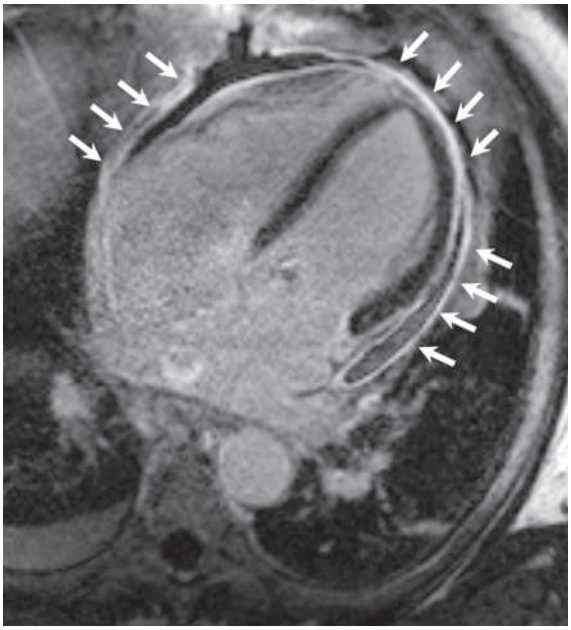
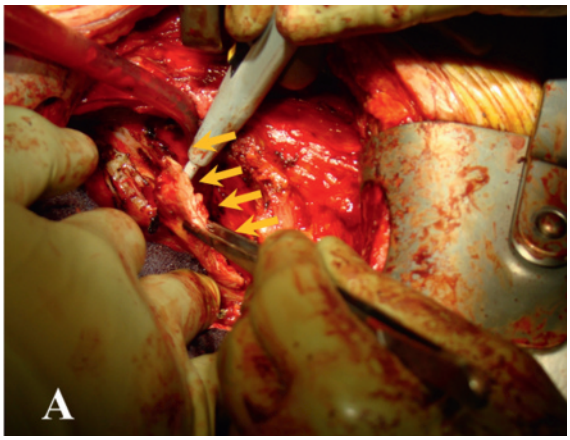
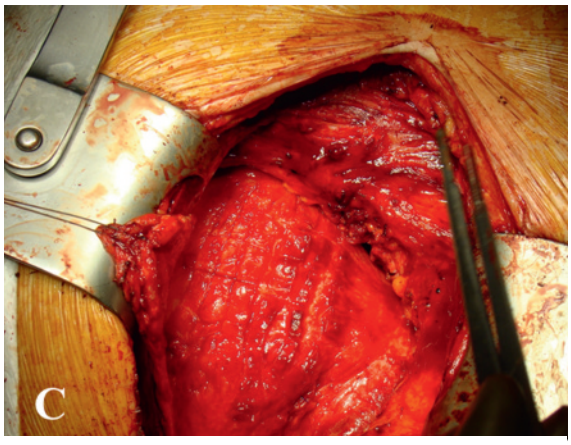
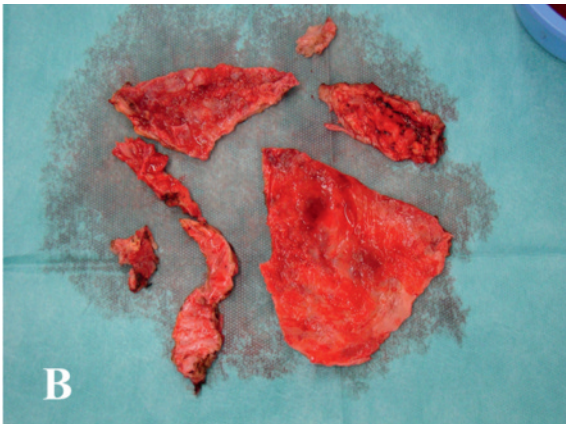


Figure 4: Late-enhancement 4 chamber CMR showing enhancement of the pericardial edges (arrows).







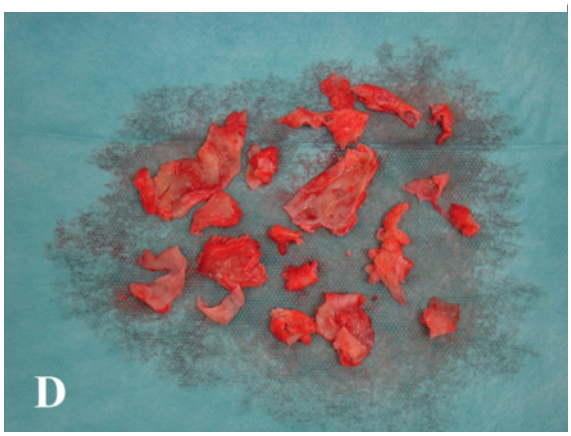


Figure 5: Surgical field and specimens of the thickened pericardium. On the left (A, B) thickened parietal pericardium (arrows) is displayed, while on the right (C, D) visceral thickened pericardium is displayed.

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