Pericardial cystic lymphangioma

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A 43-year-old woman was referred for acute chest pain. She had no relevant past medical history. Her electrocardiogram was normal. However, transthoracic echocardiography revealed an uncommon cystic pericardial effusion. Septations of variable thickness were observed in this multilocular mass (Panel A, see Supplementary data online, Video S1). C-reactive protein raised to 82 mg/L. The patient was treated efficiently with aspirin and colchicine. Serological tests for a bacterial, fungal, or immunological cause were negative.

Thoracic computed tomography scan revealed a paracardiac mass without calcification encompassing the right coronary artery (RCA) (Panel B). At cardiac magnetic resonance imaging (MRI), the mass was well-defined and contiguous to the right ventricle on steady-state free precession (Panel C, see Supplementary data online, Video S2). This lesion had heterogeneous and high signal intensity on T1-weighted with fat saturation perhaps due to the presence of proteinaceous material or blood (Panel D). This mass had high signal intensity on T2-weighted images (Panel E), a finding that reflects their fluid content, and did not enhance after gadolinium administration (Panel F). Thus, a haemorrhagic pericardial cystic lymphangioma was suspected.

Cardiac surgery revealed a pericardial cystic lymphangioma (Panel G), whose resection required partial resection of the RCA which was repaired by venous patch (Panel H). Macroscopic examination did not detect any haemorrhagic changes. Histological analysis (Panel I) showed a cystic tumour with a predominant lymphatic vascular component, lymphoid follicles, and mature adipose tissue. Coronary angiography confirmed the patent lumen of the RCA (see Supplementary data online, Video S3). At follow-up 6 months later, MRI did not detect any residual mass (see Supplementary data online, Video S4).

Cystic lymphangioma in adults is an extremely rare mediastinal benign tumour, which frequently occurs in children and young adults. To our knowledge this observation is the first to describe a pericardial cystic lymphangioma encompassing the RCA.

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

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