A 36-year-old woman with a 6-month history of exertional fatigue was referred for echocardiography. In physical examination, a low-pitched systolic murmur (Grade III/VI) was heard best in the left upper sternal border. Transthoracic echocardiography revealed dilated right cardiac chambers with severe tricuspid regurgitation. Continuous-wave Doppler echocardiography showed an 80 mmHg right ventricular outflow tract gradient with normal pulmonary valve function. Careful examination of great vessels showed a large mass in the main pulmonary artery. Left cardiac chambers and valves were normal. Transoesophageal echocardiography revealed right ventricular and atrial enlargement (Figure 1A) with no evidence of left-to-right shunt (Figure 1B). A large mass (3.5 × 2.6 cm) originating from the main pulmonary artery was detected (Figure 1C). Continuous-wave Doppler showed 84 mmHg gradients in the main pulmonary artery (Figure 1D). The patient underwent cardiac surgery and the mass was excised successfully. Pathological examination of the mass revealed diffuse infiltration of many lymphocytes, plasma cells, and macrophages (Figure 1E). Immunohistochemistry examination of the tissue showed positive CD68 (Figure 1F). These findings were in favour of xanthogranuloma.

Xanthogranuloma, a rare benign disease, is a macrophage response to non-specific tissue injury with subsequent granulomatous reaction. There have been rare reports of myocardial xanthogranulomatosis. In the present report, we described a case of isolated pulmonary artery xanthogranuloma that causes right-sided heart failure. To the best of our knowledge, there is no published report of pulmonary artery xanthogranuloma.

Figure 1 Transoesophageal echocardiography of a 36-year-old woman with a history of dyspnoea on exertion and syncope. (A) Transoesophageal echocardiography revealed right ventricular and atrial enlargement. (B) No evidence of left-to-right shunt was found by saline contrast injection. (C) A large mass originating from the main pulmonary artery was detected. (D) Continuous-wave Doppler showed 84 mmHg gradients in the main pulmonary artery. (E) Sections from a pulmonary artery mass. Diffuse infiltration of many lymphocytes, plasma cells, and macrophages is seen (H&E × 400). (F) Immunohistochemistry examination of the tissue shows positive CD68. These findings are in favour of xanthogranuloma.