A 20-year-old man, with a recent diagnosis of relapsing polychondritis, was hospitalized for crescendo angina pectoris. Relapsing polychondritis, an auto immune multi-system disease characterized by recurrent inflammation and destruction of different cartilaginous structures, was clinically diagnosed after nasal chondritis (Panel A), left cochllear–vestibular neuritis with deafness, relapsing bilateral anterior uveitis, ankle arthritis and erythema nodosum. Cardiopulmonary examinations as well as left electrocardiogram were normal. Trans thoracic echocardiogram revealed an elongated anterior mitral leaflet with severe prolapse and mild regurgitation, a saccular aneurysm of the left aortic sinus, a small circumferential pericardial effusion and a dilated hypokinetic left ventricle with ejection fraction to 40% (Panel B, Supplementary material online, Videos S1–S4). Cardiac computed tomography demonstrated a compression of the circumflex coronary artery by the aortic aneurysm (Panel C, black arrows). Patient underwent surgical treatment (Panel D) consisting in a Bentall procedure with left ventricular outflow tract reconstruction and mitral mechanic valve replacement. Histological examination of the mitral valve showed a massive fibrinoid necrosis surrounded by histiocytes (Panel E). Post-operative high dose of intra venous corticosteroids associated with cyclophosphamide achieved to reduce the disease activity.

The authors acknowledge Dr Fohlen, radiologist at CHU de Caen, for the assistance in providing computed tomography pictures.

Supplementary material is available at European Heart Journal online.