Clinical picture

Giant aortic aneurysm in Marfan syndrome

A 33-year-old man presented to the hospital with progressive dyspnea and palpitations. He had a history of scoliosis 14 years ago, requiring spinal fusion surgery. Physical examination revealed marfanoid habit and irregular heartbeats with a holosystolic murmur of mitral regurgitation and a diastolic murmur of aortic regurgitation. Electrocardiography showed atrial fibrillation and chest radiography revealed a widened mediastinum with an enlarged cardiac silhouette (Figure 1a). Echocardiogram found dilatation of the mitral annulus, aortic valve regurgitation and an ascending aortic aneurysm. Three-dimensional (3-D) reconstruction computed tomographic angiography of the chest showed a 10-cm ascending aortic aneurysm (Figure 1b). The patient underwent mechanical mitral valve replacement and composite graft replacement of the aortic valve, aortic root and ascending aorta, with reimplantation of the coronary arteries into the graft (Bentall procedure) with a full and uneventful recovery. Afterwards, it was found a fibrillin-1 mutation, confirming the diagnosis of Marfan syndrome.1,2

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References