Multi-imaging assessment of successful surgical treatment of pulmonary artery dilatation and dissection in Marfan syndrome

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A 42-year-old male patient with a 1-month history of dyspnoea of great effort, daily oppressive chest pain with intensity 5/10 irradiated to the shoulder and left scapular region, palpitations and haemoptysis. He was diagnosed according the new Ghent criteria of Marfan syndrome. He had aortic dilatation and systemic score 13/20. Transthoracic echocardiography showed dilatation of the trunk of the pulmonary artery (70 mm) and right and left branches and dissection of the pulmonary trunk, above the valvular plane with a wide communication site, extending to the left pulmonary branch (A and B; see Supplementary data online, Movie S1), and dilatation of the ascending aorta (43 mm). Computed tomography corroborated these findings. (C, D and E). The histopathological study of the pulmonary artery demonstrated extensive replacement of the elastic mid layer by repairing connective tissue fibroblasts, mild cystic necrosis in the middle elastic layer and extensive recent haemorrhage in the adventitia suggesting acute dissection (F). The patient went for surgical treatment with a surgical time of 5 h and 32 min.

The incidence of aneurysmatic dilatation and dissection of pulmonary artery has been described with fatal outcome. In this case, the patient went immediately to surgery and the postoperative evolution was satisfactory (G). He is in New York Heart Association functional class I. We consider that the diagnosis and timely treatment change the fatality of the prognosis in Marfan syndrome patients with involvement of pulmonary artery.

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