An unusual cause of systolic murmur

Une cause inhabituelle d’un souffle systolique

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A 50-year-old male patient was admitted for shortness of breath (New York Heart Association functional class II) and constitutional syndrome of 1 month’s duration. Physical examination was remarkable for a grade 3/6 systolic murmur best heard at the second left intercostal space. Blood pressure was 103/88 mmHg, heart rate was 84 beats/minute and his oxygen saturation at rest was 93%. Electrocardiogram and blood gases at rest were normal. Laboratory tests revealed microcytic hypochromic anaemia, and colonoscopy disclosed a colon adenocarcinoma. Echocardiography was subsequently performed, revealing a mildly dilated right ventricle, mild tricuspid regurgitation, no pulmonary regurgitation and severe elevation of right ventricular systolic pressure (70 mmHg), as well as a 50 × 33 mm mass in the main pulmonary trunk extending into both pulmonary artery branches and producing severe stenosis with minimal residual lumen (Fig. 1). Although computed tomography showed that the mass did not enhance with contrast (Fig. 2), suggesting a giant thrombus as the most likely diagnosis, pathological analysis of the mass confirmed a pulmonary leiomyosarcoma. The patient underwent successful surgical resection of both tumours and remains alive 18 months after the initial diagnosis.
Transthoracic echocardiography (modified parasternal short-axis view at the level of the great vessels) disclosing a mass (asterisk) measuring $50 \times 33$ mm, filling the main pulmonary trunk and extending into both pulmonary branches. LPAB: left pulmonary artery branch; MPT: main pulmonary trunk; PV: pulmonary valve; RPAB: right pulmonary artery branch.

Primary pulmonary artery leiomyosarcomas are rare tumours that may mimic pulmonary thromboembolism. The differential diagnosis between both entities may be taxing but is important to avoid inappropriate therapy. The prognosis is usually ominous, even with surgical resection, with a mean survival time of 10 months. The question of whether the colon adenocarcinoma and pulmonary leiomyosarcoma were somehow related in our patient remains unclear.

**Disclosure of interest**

The authors declare that they have no conflicts of interest concerning this article.