Striking pulmonary arteries enlargement in Eisenmenger’s syndrome

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A 54-year-old man was admitted to the Department of Internal Medicine at the University of Brescia (Italy) because of syncope. He was affected by Eisenmenger’s syndrome, due to a congenital ventricular septal defect, diagnosed by an invasive haemodynamic study when he was 18 (at the end of the 17s). The ventricular septal defect had never been surgically repaired. The patient showed a progressively worsening cor pulmonale and chronic hypoxaemic respiratory failure due to the intracardiac right-to-left shunt associated with severe pulmonary hypertension. As expected, even in long-term oxygen therapy, PaO₂ levels were usually <45 mmHg and SaO₂ <75%. The patient showed erythrocytosis, central cyanosis, and strikingly overt finger clubbing (Panel A) and experienced increasing fatigue and disability. Several times in the past, he had refused further clinical assessment to evaluate the feasibility of a total heart–lung transplantation.

The actual chest X-ray revealed an impressive oval opacity of the medial–basal field of the right lung (7 cm maximal diameter) and a right basal parenchymal consolidation feature, neoplastic-like findings (Panel B).

The magnetic resonance imaging confirmed the presence of the ventricular septal defect (pars membranacea) and of a marked dilation of the right ventricle (Panel C—a, right pulmonary artery; b, aortic valve; c, ventricular septal defect; d, descending thoracic aorta).

The chest computed tomography showed a striking dilation of the main pulmonary artery and of its branches: impressively, the right pulmonary artery had a 7 cm diameter and the left pulmonary artery a 5 cm one. The pulmonary vessels were largely obliterated by thrombotic material (Panel D—a, ascending thoracic aorta; b, right pulmonary artery; c, descending thoracic aorta; d, left pulmonary artery; e, thrombus).

Panels E and F (a, thrombus; b, pulmonary artery lumen) show computer tomography images obtained >2 years later (on October 2009), before and after the contrast phase: no enhancement is observed in the ‘mass’, thus confirming its thrombotic nature; further, the finding did not significantly change over a long time, confirming the above-mentioned interpretation.

The patient is still taking oral anticoagulant therapy and bosentan, 62.5 mg twice a day.