Pulmonary artery intramural haematoma: an unusual cause of sudden death in patients with chronic pulmonary hypertension

A 70-year-old woman with history of chronic idiopathic pulmonary hypertension was admitted to the cardiology department because of sudden onset of chest pain and dyspnoea.

A transthoracic echocardiogram showed moderate pericardial effusion without signs of cardiac tamponade and extremely severe pulmonary hypertension (figure 1A).

CT of the chest ruled out a pulmonary embolism, but documented a huge dilatation of the pulmonary trunk (48 mm) extending to the bifurcation of right and left pulmonary arteries (figure 1B).

The day after admission the patient suddenly collapsed and died despite advanced resuscitation measures, including pericardiocentesis.

Postmortem examination disclosed haemopericardium due to a single stab wound of the heart wall caused by emergent pericardiocentesis, and a large haematoma affecting the main pulmonary trunk and both pulmonary arteries (figure 1C,D). No intimal tear and no clear-cut defect were seen in the pulmonary adventitia (figure 1E,F).

Pulmonary artery intramural haematoma is extremely rare.[1] Pulmonary artery dissection, although rare, has been found more frequently.[2]

The haemorrhage into the pulmonary artery media and the absence of an entry tear define the non-communicating pulmonary artery dissection. Extreme arterial dilatation, severe

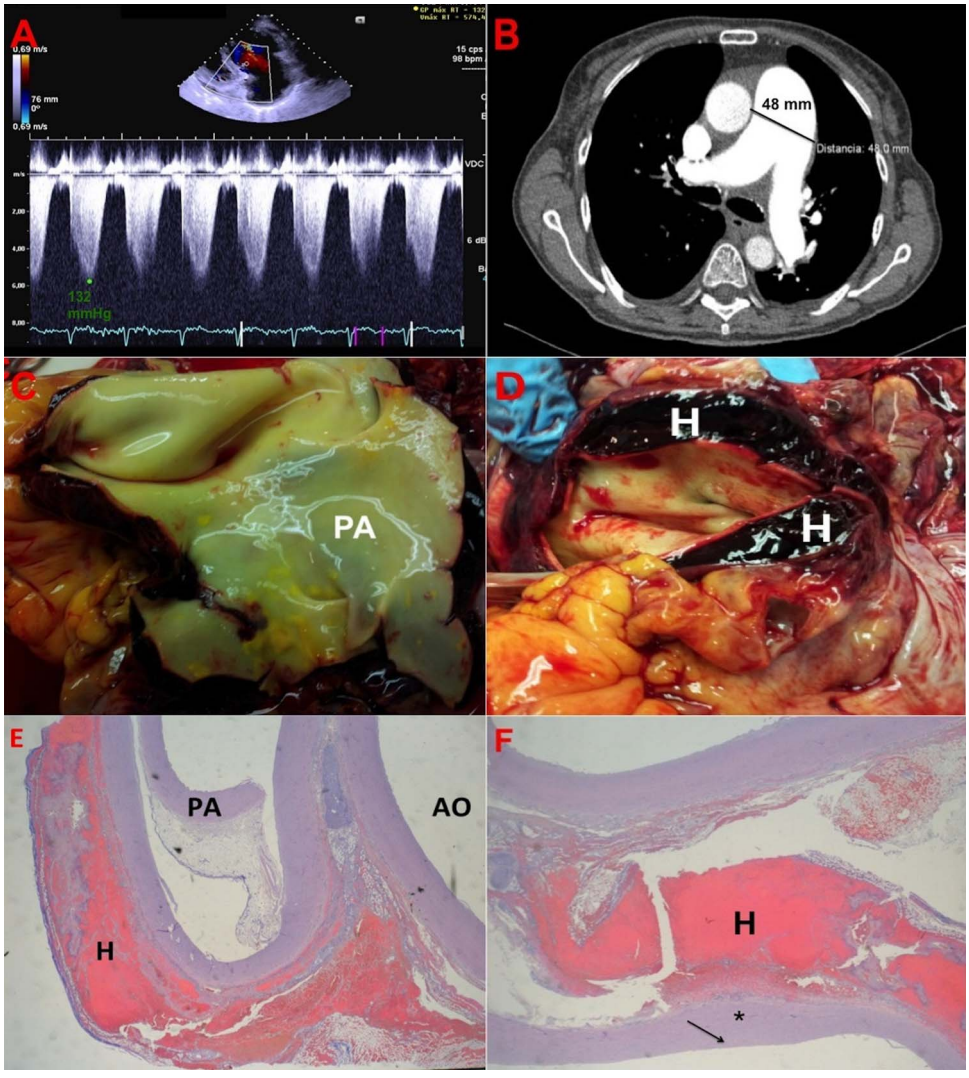

Figure 1 (A) Severe pulmonary hypertension estimated by echo Doppler. (B) Huge dilatation of the pulmonary trunk (48 mm) and both pulmonary arteries in the contrast-enhanced CT. (C) Severe dilatation of the pulmonary artery (PA) at postmortem examination. (D) Postmortem macroscopic examination of the large haematoma (H) involving the main pulmonary trunk, and extending to both pulmonary arteries and the aortic root. (E) Postmortem histological study of the pulmonary trunk, haematoxylin–eosin stain. The proximity of the ascending aorta and the PA is clearly seen, as well as the PA intramural haematoma (H). (F): Histological study of the right PA. Undamaged intima (arrow) and media (*) layers. Intramural haematoma (H).

pulmonary hypertension and medial degeneration with fragmentation of elastic fibres, may have a significant pathogenic role.

Pulmonary artery dissection and haematoma are often lethal and must be taken into consideration when treating patients with severe pulmonary hypertension.[1 2] Sudden onset of dyspnoea or chest pain are the main alarm symptoms, and non-invasive imaging techniques are the preferred diagnostic method.

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