Pulmonary hypertension

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A 29-year-old man presented with exertional dyspnea and syncope. He had been diagnosed with idiopathic pulmonary hypertension (PH) 7 years prior. A chest CT showed marked dilatation of the pulmonary artery trunk (Figure 1).

PH is a progressive disease of the pulmonary arteries that is characterized by marked remodeling of the pulmonary vasculature and a progressive increase in pulmonary vascular pressure, leading to right ventricular hypertrophy and remodeling. PH is defined as a mean pulmonary artery pressure greater than 20 mmHg at rest, leading to increased pulmonary vascular resistance. It may be idiopathic or arise in the setting of other clinical conditions. Diagnosis and treatment planning are made on the basis of clinical and hemodynamic criteria, pulmonary function test results, and radiological and histological findings, usually at specialized referral centers. The clinical picture is nonspecific and may include dyspnea, limitations in daily activities, retrosternal and chest pain, dizziness, cyanosis, and hemoptysis, among other findings. Death usually results from right ventricular failure. It is of note that pulmonary arterial hypertension is a subgroup of PH. This subgroup includes the idiopathic form.[1,2]

The classic imaging findings of PH can be divided into three categories: vascular, cardiac, and parenchymal. The chief vascular finding is dilatation of the pulmonary artery trunk, which should be measured in the axial plane at the level of its bifurcation and orthogonal to its long axis. A diameter equal to or greater than 29 mm should be considered abnormal. We should also consider the ratio of the diameters of the pulmonary artery and aorta, which should be measured in the same plane as that in which the aforementioned finding is measured. The pulmonary artery diameter should not be greater than the ascending aorta diameter. It should, however, be considered that main pulmonary artery dilatation may develop in pulmonary fibrosis patients in the absence of PH. Other vascular findings include a ratio of the diameters of arteries and bronchi > 1, observed in three or four lung lobes, and bronchial artery dilatation (hypertrophy). Cardiac findings include hypertrophy or dilatation of the right cavities, leading to inversion of the interventricular septum. Parenchymal findings include the presence of a mosaic attenuation pattern and the presence of centrilobular ground-glass nodules.[1,2]

Extensive clinical, laboratory, and hemodynamic studies did not determine the specific cause of this patient's PH, which was therefore classified as an idiopathic form.

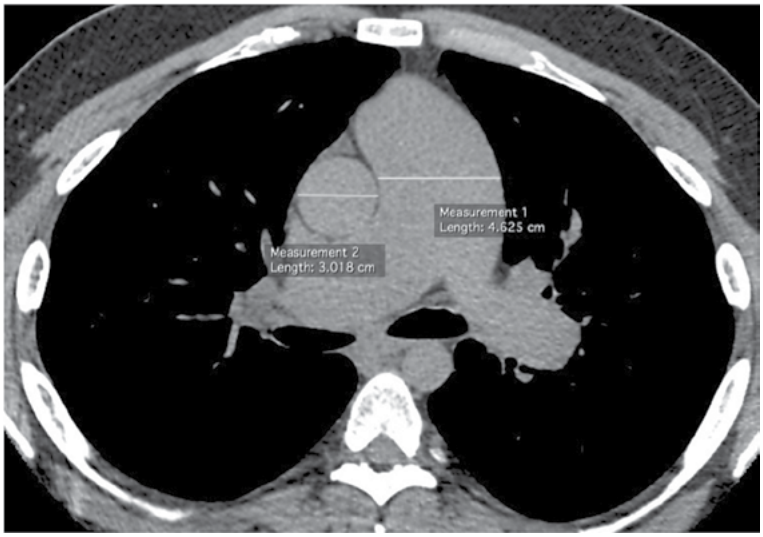

Figure 1. Axial chest CT scan with mediastinal window settings shows an increased pulmonary artery diameter (46 mm — normal up to 29 mm). Also note that this diameter is greater than the aortic diameter measured in the same plane (30 mm).

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