Clinics in Respiratory Medicine

Clinical Image
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Massive Dilation of Pulmonary Artery

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Clinical Image

Sixty-year-old African American woman presented to the hospital with progressively worsening shortness of breath. She underwent an extensive workup that included chest X-ray, CT angiogram of the chest and an echocardiogram. In the transthoracic echocardiogram, she was seen to have elevated right and left sided cardiac pressures with preserved left ventricular function with impaired relaxation. In addition, she was also found to have massively dilated pulmonary artery trunk extending bilaterally. Accompanying images show massively dilated pulmonary artery trunk with borderline dilated right and left atrium as seen on the CT angiogram of the chest (Figure 1). Diameter of the pulmonary artery trunk was measured on the CT chest to be 11.5 cm. Our patient was labeled as having pulmonary artery hypertension secondary to heart failure with preserved ejection fraction. No valvular, other congenital heart problems were found after the extensive workup. She was initiated on a regimen consisting of calcium channel blockers, diuretics and cardiac rehab with which her symptoms improved. Patient declined to undergo any surgical intervention. She gets an annual chest imaging to keep a close eye on the size of the pulmonary artery (Figure 2).

The diameter of a normal pulmonary artery as measured through CT scan of the chest is considered to be 2.72 cm (SD=0.3). In patients who have pulmonary artery hypertension this measurement increases to 3.47 cm (SD=0.33). It is suggested that, using unenhanced axial 10 mm CT sections, the upper limit of normal main pulmonary artery diameter is 3.32 cm [1]. The Framingham investigators reviewed the non-contrasted chest CT scans from 706 individuals who were deemed "healthy" (defined by the lack of obesity, hypertension, or history of chronic obstructive pulmonary disease [COPD], pulmonary embolism, or cardiovascular disease) and found a mean (SD) main PA diameter of 25.1 (SD: 2.8) mm, with an upper limit of normal of 28.9 mm in men and 26.9 mm in women [2]. Massive dilation of the pulmonary artery and/or the pulmonary trunk is mostly

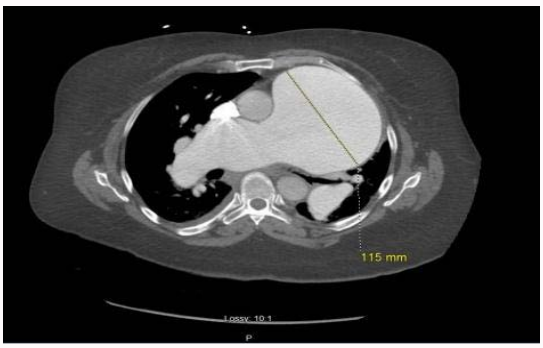

Figure 1: CT angiogram axial view showing massive dilation of pulmonary artery trunk with secondary compression of left main stem bronchus and passive atelectasis of the left lung.

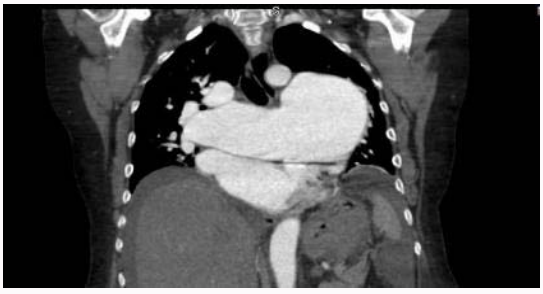

Figure 2: CT angiogram coronal view showing dilated pulmonary artery branches.

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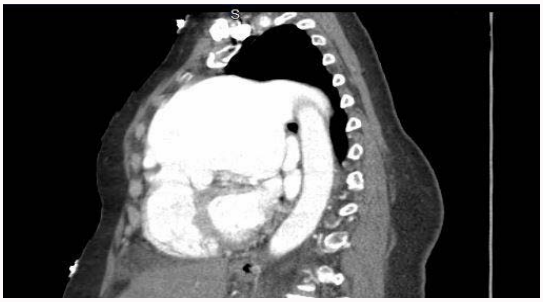

Figure 3: CT angiogram sagittal view depicting the impingement of surrounding structures by dilated pulmonary artery.

seen associated with congenital abnormalities (Figure 3). Congenital pulmonary valve stenosis has been associated with the development of massive pulmonary arterial dilatation [3]. Idiopathic dilation of pulmonary artery (IDPA) is a rare disease with little known about its etiology and pathogenesis. Kaplan et al. [4]. Postulated it as mal-development of the entire pulmonary tree and congenital weakness of the arterial wall whereas assman [5] thought of it as an unequal division of truncus arteriosus communis. Over time, this dilatation may distort surrounding structures and lead to compression of the left main coronary artery (LMCA) or the left main stem bronchus.

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