H50 A Case of Pulmonary Artery Dissection in a Woman With Chronic Pulmonary Hypertension

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Learning Overview: After attending this presentation, attendees will better understand pulmonary artery dissection as a fatal complication of chronic pulmonary hypertension and its autopsy findings.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by raising awareness of sudden death in adults with long-standing pulmonary hypertension.

A 57-year-old woman with a history of pulmonary hypertension and a remote history of polysubstance abuse (1998) was found unresponsive in her locked residence by a neighbor after reportedly feeling unwell for a couple days.

At autopsy, external examination noted digital clubbing. Opening the chest cavity revealed a tense, discolored pericardial sac containing 650mL of liquid and clotted blood. Hemorrhage was identified at the root of the pulmonary artery, and careful examination revealed a 2mm full thickness tear, located 5mm distal to the pulmonic valve. The dissection extended 6.0cm distally to the left main pulmonary artery, terminating 5mm proximal to the take-off of the apicoposterior and anterior segment branches. The pulmonary trunk was dilated, measuring 3.8cm in diameter at the pulmonary root, 3.8cm at the left main pulmonary artery, and 3.2cm at the right main pulmonary artery. There was diffuse atherosclerosis extending into the distal branches of the pulmonary artery. The heart weighed 530 grams and displayed right ventricular hypertrophy with a wall thickness of 8mm, as well as dilated tricuspid, pulmonic, and mitral valves (12.5cm, 9.3cm, and 11.0 cm, respectively).[1] Multiple cavernous hemangiomas of the liver ranging in size from 5mm to 35mm were incidentally identified.

Histologically, the pulmonary artery showed extensive medial degeneration with a large defect extending from the tunica media into the adventitia with associated hemorrhage. The small and large pulmonary vessels showed medial hypertrophy, highlighted with elastin stain. Scattered plexiform lesions were identified. Elastin stain further highlighted fragmentation and loss of elastic fibers with increased intralamellar and translamellar extracellular mucoid material noted on Alcian blue stain. The aorta showed intact elastic fibers with no increase in extracellular mucoid material.

While right ventricular failure is the main cause of death in patients with pulmonary hypertension, autopsy is rarely performed in these cases of sudden death in otherwise stable patients. It has been reported that 28% of pulmonary artery hypertension patients die unexpectedly within three years of diagnosis. Dilatation of the pulmonary artery has been independently associated with unexpected death in these patients with two possible mechanisms: pulmonary artery rupture/dissection and potentially arrhythmogenic pulmonary artery compression of the left main coronary artery. In one study, 2 of 26 unexpectedly deceased pulmonary hypertension patients underwent autopsies, and both showed cardiac tamponade due to pulmonary artery dilatation and dissection.[2]

Pulmonary artery dissection is a rare, but fatal, complication of chronic pulmonary hypertension. Some of the underlying etiologies include: congenital cardiac diseases, pulmonary artery obstruction due to emboli (thrombotic, tumor or foreign material), fibrosing pulmonary diseases, emphysema and left-sided cardiac conditions. If there is no underlying etiology discovered, a diagnosis of primary or idiopathic pulmonary hypertension is made. Histologic findings are often most pronounced in small pulmonary arteries and include medial thickening due to smooth muscle hyperplasia and hypertrophy, intimal thickening due to intimal smooth muscle cells or fibrosis, plexiform lesions, and dilation lesions.[1]

Overall, pulmonary artery dissection as a result of chronic pulmonary hypertension is a rare but fatal entity that may be seen in the forensic pathology community.

Reference(s):

1. Buju, L.M., Butany, J., ed. *Cardiovascular Pathology*. London: Elsevier Press, 2016.
2. Zylkowska J., Kurzyna M., Florczyk M., Burakowska B., Grzegorczyk F., Burakowski J., Wieteska M., et al. Pulmonary Artery Dilatation Correlates with the Risk of Unexpected Death in Chronic Arterial or Thromboembolic Pulmonary Hypertension. *Chest* (December 2012): 142(6): 1406-1416.

Pulmonary Hypertension, Pulmonary Artery Dissection, Hemopericardium

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