G58 Spontaneous Pulmonary Arterial Dissection: A Case Report

Margherita Neri, PhD, Stefania Bello, MD, Raffaella Bisceglia, MD, and Carmela Fiore, MD, Department of Forensic Pathology, University of Foggia, Viale degli Aviatori 1, Foggia, 71100, ITALY*

The goal of this presentation is to present a fatal case of spontaneous pulmonary arterial dissection with a wide immunohistochemical study about alteration of pulmonary wall.

This presentation will impact the forensic science community for the rarity of the deaths due a spontaneous dissection of the pulmonary trunk.

Unlike dissection in systemic arteries, pulmonary artery or main pulmonary branche dissection, is usually lethal. So the diagnosis of this condition is very rarely made during life and most commonly diagnosed at autopsy in cases of sudden and unexpected death. Hemorrhagic pericardial effusion and cardiac tamponade usually follow the outward rupture of the proximal main pulmonary artery.

With regard to pathogenesis, pulmonary artery dissection is strongly associated with primary and much more frequently, secondary pulmonary hypertension. Secondary pulmonary hypertension most often results from congenital cardiac lesions, above all with various forms of left-to-right shunting, most commonly patent ductus arteriosus, or congenital ventricular septal defect. These cardiac conditions predispose individuals to the development of pulmonary artery aneurysm by generating sustained high pulmonary flow rates and pulmonary artery pressure. However, other possible causes are Marfan syndrome and other connective tissue diseases, infectious processes and inflammatory conditions, such as Behcet disease. Anyway pulmonary artery dissection is exceedingly rare in the absence of pulmonary hypertension or other pathologic conditions.

The clinical presentation of pulmonary artery dissection is highly variable and the symptoms are nonspecific, most frequently chest pain, dyspnea, cyanosis, and hemodynamic compromise. Diagnostic instruments for this condition are noninvasive imaging techniques, including echocardiography, CT, and magnetic resonance imaging (MRI).

The vascular histopathologic changes associated with the majority of pulmonary artery dissections involve medial degeneration, with fragmentation of elastic fibers. These changes may represent an intrinsic weakness in the vessel wall which is compounded by the increased hemodynamic shear stresses of pulmonary hypertension, thereby predisposing an intimal tear. The pathogenetic mechanism of dissection in absence of histopathologic alterations remains substantially unclear.

The case presented concerns sudden death due to spontaneous pulmonary artery dissection.

A 72-year-old woman was admitted to the Emergency Department for chest pain, spread to mandible, dyspnea, and jugular tightness, and she referred these symptoms after bleach inhalation during housecleaning.

Physical examination, ECG and CT were unremarkable. Cardiac ultrasonography showed concentric ventricular hypertrophy and ascending thoracic aorta ectasia (50 mm). Laboratory blood values demonstrated neutrophilia, lymphopenia, monocytosis and increased erythrosedimentation rate. Two days later she died.

A postmortem examination was performed and revealed a large hemorrhagic area in left posterior mediastinum and pericardial sac containing approximately 150 ml of blood and 250 g of clotted blood. The source of hemorrhage was readily identified as a 2 cm tear in the wall of the pulmonary trunk and so dissection and rupture of the artery.

Microscopic sections of the pulmonary artery revealed regular morphology of the wall layers. The medial layer showed fragmentation of elastic fibers, marked fibrosis and copious erythrocytes. In a section the intimal tear was identified as initial site of dissection.

The immunohistochemical investigation of the pulmonary artery samples in whole artery wall and in laminar dissection was performed with antibodies anti TGF-beta-1, TGFBR1 (ALK-5) e TGFBR2, ALK-1, fibrillin and endoglin. Fibrillin showed a massive and diffuse positive reaction of the whole pulmonary artery near the dissection, but it showed negative reaction in laminar dissection; Endoglin showed a weak positive reaction in the whole artery and a negative reaction in the laminar dissection; TGF-beta1 revealed a weak positive reaction in the whole pulmonary artery and a strong reaction in the laminar dissection; TGFBR1 and ALK-1 showed a moderate positive reaction in the whole pulmonary artery wall and a massive positive reaction in laminar dissection; TGFBR2 revealed a massive positive reaction of the whole pulmonary artery, but it showed moderate reaction in laminar dissection.

A fatal hemopericardium caused by spontaneous pulmonary artery dissection was recorded as the cause of death. The histological investigation of the pulmonary artery samples revealed the absence of hypertensive arterial changes and the immunohistochemical showed the absence of any connective tissue disease of the pulmonary trunk. So the presented case illustrates a very rare cause of sudden death in a

 ** Presenting Author*

spontaneous dissection of a normal pulmonary trunk.

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 ** Presenting Author*