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Title: An autopsy case of portopulmonary hypertension associated with idiopathic portal hypertension

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Body: Idiopathic portal hypertension (IPH) is a rare cause of portal hypertension. The present autopsy report documents a case of portopulmonary hypertension (PoPH) associated with IPH, focusing on the portal and pulmonary vasculopathy. In Feb 2011, a 70-year-old woman visited a community hospital, presenting with progressive dyspnea. Right heart catheterization (RHC) indicated pulmonary hypertension, but no further investigations were conducted. In May 2012, she was found to have esophageal varix and referred to our hospital. Second RHC showed marked elevation of mean pulmonary artery pressure (68 mmHg), and comprehensive studies led to the diagnosis of PoPH associated with non-cirrhotic portal hypertension. Medical treatment was started, but to no avail, and she died of right heart failure in Sept 2012. Autopsy showed intact bile ducts and hepatic arteries but obstructed portal veins by fibrosis in the portal tract (A), which was consistent with the pathological features of IPH. In the lungs, pulmonary arterioles showed intimal and medical thickening (B), and plexiform lesions and fibrotic venular obstruction were also noted. Notably, lumens of some arterioles were separated into several compartments (C), suggestive of recanalization. A case of pathologically-proven PoPH associated with IPH was presented. Varied pulmonary vasculopathy appeared to employ in the development of pulmonary hypertension in the present case.