H60 Massive Portal Vein Thrombosis as a Rare Cause of Unexpected Death in a Non-Cirrhotic Patient

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Learning Overview: The goal of this presentation is to present the case of an unexpected death in a non-cirrhotic patient with acute abdominal pain of recent onset due to a massive Portal Vein Thrombosis (PVT).

Impact on the Forensic Science Community: This presentation will impact the forensic science community by showing that a common presenting symptom, such as abdominal pain, in a young, healthy adult can be a manifestation of a rare diagnosis such as PVT, supporting the relevance of the hospital autopsy as an instrument of audit in the practice of emergency medicine.

PVT as the etiology of abdominal pain in an otherwise healthy young adult is an uncommon occurrence and is capable of leading to serious adverse short- and long-term events in the affected patients. While the occlusion of the extrahepatic portal and splenic vein can provoke complications mainly related to portal hypertension, mesenteric vein obstruction shows a high rate of complications and mortality due to intestinal infarction. The occurrence of this condition is probably underestimated given the fact that the clinical appearance can vary from asymptomatic patients incidentally diagnosed to patients with severe complications. The underlying cause of PVT includes malignancy, chronic liver diseases, local inflammatory processes, systemic disorders, including myeloproliferative disorders, and thrombophilia.

In a high percentage of patients, two or more risk factors are present. In population studies, the cohort of patients with occlusion in the portal tributary system can be divided into three similarly large subcohorts of major etiologies: malignant thrombosis mainly due to gastrointestinal, hepatobiliary, or pancreatic cancer; chronic liver diseases, especially liver cirrhosis with portal hypertension; and non-malignant, non-cirrhotic PVT. While the underlying disease determines the natural history and outcome of patients in the case of malignant thrombosis, the two other groups of patients are characterized by an outcome influenced by PVT. Furthermore non-malignant, non-cirrhotic PVT has often been described as an entity of its own, leading to non-cirrhotic portal hypertension, which represents more of a consequence than a complication of another disease. It has been estimated that in non-cirrhotic PVT patients, thrombophilic states account for approximately 40% to 60% of PVT cases, and local factors are thought to be the causative factor in 10% to 50%. In about one-third of patients, the etiology is unknown. The site, extent, chronicity, and course of thrombosis determine clinical presentation as well as complications in affected patients. While partial PVT is usually discovered incidentally by routine diagnostics and remains clinically silent, the complete occlusion of the vein (90%–100% of the lumen) is associated with abdominal and/or lumbar pain characterized by sudden onset or progressive development over the course of a few days. Acute and complete thrombosis is usually associated with intestinal congestion and occasionally with non-sanguineous diarrhea. The most feared complication is intestinal infarction with a mortality of 20%–60%, leading to extended resections with a high risk of postoperative complications. In contrast to intestinal congestion, infarction often presents with persistent pain, hematochezia, guarding, contractures, ascites, or multiorgan failure with metabolic acidosis. This complication is usually found when the mesenteric veins are involved.

Case Report: A 61-year-old man reached the emergency department of the local hospital complaining of onset of acute abdominal pain for two days. Hypotension (80/60mmHg) and bradycardia (55bpm) were detected when he suddenly collapsed. Resuscitation maneuvers were attempted, unsuccessfully. Hospital autopsy was performed the day after death to investigate cause of death. Diffuse mesenteric ischemia was observed due to massive PVT extended to splenic and mesenteric veins. Liver was increased in volume (32cm x 20cm x 10cm), non-cirrhotic, with mild steatosis. Histological assessment of thrombus samples with immunohistochemistry was also performed to establish chronological stage. Audit and mortality and morbidity review with clinicians was performed after postmortem investigation.

Portal Vein Thrombosis, Hospital Autopsy, Mesenteric Ischemia

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