LETTER to the EDITOR

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Ruptured Collateral Venous Aneurysm Associated with IVC Agenesis Resulting in a Massive Retroperitoneal Hematoma

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Dear Editor,

The embryonic development of inferior vena cava (IVC) is a complex process with regressions and anastomoses of embryonic veins. Congenital abnormalities of the IVC and its tributaries were first described by Abernethy (1) in 1793 with the demonstration of a congenital mesocaval shunt and continuation of IVC with azygos vein in a 10-month-old child. IVC agenesis (IVCA) is one of the rare anomalous developments of IVC having a controversial pathophysiological mechanism in which defective development of the embryonic veins and intrauterine/perinatal IVC thrombosis are both suggested (2). Many patients remain clinically silent but, it may also be diagnosed following its serious complications, most commonly unprovoked multiple deep vein thromboses (DVT) in young individuals, particularly in the lower extremities (3). However, unusual presentations may also occur. We present an extremely rare presentation of IVCA complicated by massive retroperitoneal hematoma (RPH) in a young patient with acute abdominal pain. A 35-year-old previously healthy young male person was admitted to the emergency department with acute onset left abdominal pain. He was hypotensive, and the laboratory findings showed decreased hemoglobin levels in addition to metabolic acidosis. He had no bleeding disorders and was not taking any anticoagulant treatment. IV contrast-enhanced abdominal computed tomography revealed a massive RPH with the largest diameter of 20 cm with a mass effect on adjacent structures. The arterial phase images excluded the arterial etiology of the retroperitoneal bleeding (Figure 1). The intrahepatic IVC was absent with hypoplasic hepatic IVC. Superiorly, the hepatic veins were draining into the suprahepatic IVC. Inferiorly, the common iliac veins were draining into the dilated ascending lumbar veins, which in turn drain into the azygous/hemiazygous system via paravertebral varicoid collaterals, of which most were thrombosed (Figure 2). The renal veins were also draining into the dilated azygous system. A filling defect was observed in the joining part of the

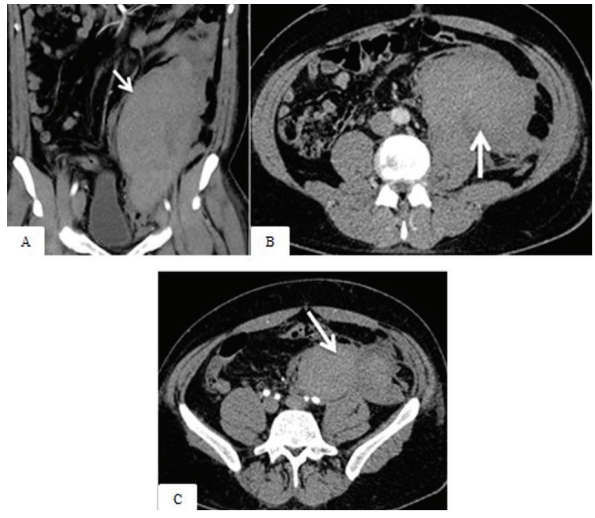

Figure 1. (A-C) Arterial phase IV contrast-enhanced abdominal CT scan showing a massive RPH near the left side of the aorta with mass effect on the left bowel segments and the bladder (arrows)
IV: Inferior vena, CT: Computed tomography, RPH: Retroperitoneal hematoma

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renal veins but attributed to an admixture artifact rather than a thrombus due to the lack of renal parenchymal findings of venous congestion. There was a tubular retroperitoneal aneurysmal lumbar collateral vein, very close to the hematoma, whose rupture was probably caused the large hematoma. DVT of bilateral external iliac veins was also noted (Figure 3). In addition to the continuation of the fluid and erythrocyte replacement, low-molecular-weight heparin was added to the treatment and he was referred to the cardiology department for follow-up. Hemoglobin levels started to increase with the correction of the metabolic acidosis and blood pressure. With the concern of malignant involvement of the veins, especially from testis tumors, scrotal Doppler ultrasonography was performed and did not reveal any pathology. The final diagnosis was spontaneous rupture of the aneurysmal compensatory collateral paraspinal veins resulting in a massive RPH. In conclusion, spontaneous RPH is a very rare clinical entity that typically occurs in patients who are receiving anticoagulation or hemodialysis. It is an extremely rare complication of IVCA. Although the collateral paraspinal veins are important ways for sustaining venous return in these patients, they can become so dilated that they are aneurysmal and ruptured, as in this study.

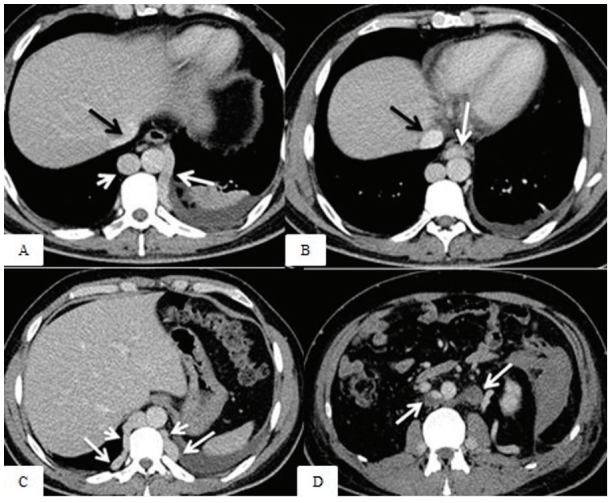

Figure 2. (A-D) The hypoplasic hepatic IVC (A, black arrow) and the normal suprahepatic IVC (B, black arrow) are shown. Note the dilated azygous vein (A, B, short white arrow), dilated accessory hemiazygous vein (A, B, white arrow), and dilated ascending lumbar veins (C, white arrows) which in turn drain into the azygous and hemiazygous system via paravertebral varicoid collateral (C, short white arrows) veins. Thrombosed collateral veins are shown (D, arrows)
IVC: Inferior vena cava

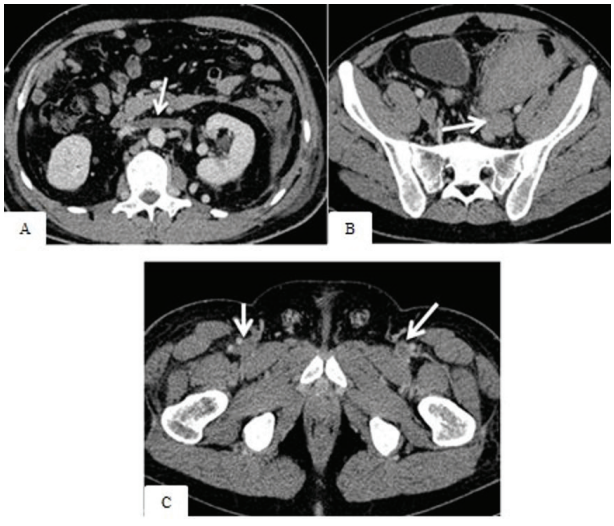

Figure 3. The renal veins draining into the dilated azygous system are shown. The filling defect in the joining part of the renal veins that was attributed to an admixture artifact is visible (A, arrow). A large tubular retroperitoneal structure among the dilated paravertebral collateral vessels interpreted as a likely aneurysmal left lumbar collateral vein whose rupture had caused the large hematoma is shown (B, arrow). DVT of bilateral EIVs was also noted (C, arrows)
DVT: Deep vein thromboses, EIVs: External iliac veins

Ethics

Informed Consent: Verbal informed consent has been taken from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: B.E., H.Ö., Design: B.E., N.N.W., H.Ö., Data Collection or Processing: B.E., Analysis or Interpretation: B.E., N.N.W., H.Ö., Literature Search: B.E., N.N.W., Writing: B.E.

Conflict of Interest: No conflict of interest was declared by the authors.

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