Case 8719

Fatal ischemic small bowel infarction in anti-phospholipid syndrome

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Section: Abdominal imaging
Area of Interest: Abdomen Liver Lung
Procedure: Diagnostic procedure
Imaging Technique: Ultrasound
Imaging Technique: Conventional radiography
Case Type: Clinical Cases
Patient: 52 years, female

Clinical History:

A 52 year old woman, affected by the anti-phospholipid syndrome (APS), and subjected to dialysis because of right shrunken kidney and left nephrectomy, came to our attention for complaints of generalized abdominal pain and frequent episodes of diarrhoea.

Imaging Findings:

Her medical history included coagulation disorders, thromboembolic events, and positioning of aorto-superior mesenteric, left aorto-renal and aorto-bi-iliac prostheses.

Two months earlier, the patient developed an obstruction of aorto-superior mesenteric bypass with distal revascularization by collateral vessels stemming from gastroduodenal artery and severe subocclusive stenosis of left aorto-renal bypass. After left renal revascularization, nephrectomy was carried out owing to acute intraparenchymal bleeding.

Following the onset of abdominal symptoms, an ultrasound examination, performed in emergency at our unit, revealed: contracted gallbladder with stones and absence of inflammatory signs; colonic meteoric distension, particularly in the right region, with minimal fluid accumulations in the hepatic, splenic, pelvic and left colonic areas. Plain abdominal X-ray showed gastric distension, with evident gas-fluid levels in the small bowel area, while the colonic frame was empty.

CT evaluation confirmed the above findings, showing diffuse fluid distension of small bowel loops, with multiple gas-fluid levels, accentuation of median abdominal wall hernia and minimal free peritoneal fluid in the hepatic, splenic, pelvic and left colonic areas.

One week later, another CT scan showed: inhomogeneous liver parenchyma; an inhomogeneous hypodense area located just below the diaphragm and characterized by indistinct margins and dilatation of intrahepatic bile-ducts, suggestive of hepatic abscess. Moreover, a thoracic CT scan revealed a consolidation area within the lower lobe of right lung in association with scissural thickening.
**Discussion:**

APS is an autoimmune disorder, characterized by association of arterial/venous thrombosis or recurrent pregnancy loss with positivity in at least one of the laboratory tests employed to detect anti-phospholipid (aPL) antibodies, including lupus anticoagulant (LA), anti-cardiolipin (aCL) and anti-B2-glycoprotein I (aB-2GPI) antibodies. aPL antibodies, targeting antigens located on endothelium, platelets, apoptotic cells, oxidized low-density lipoproteins, C protein, antithrombin III and prothrombin, have been also described, but their clinical significance remains uncertain. The exact pathogenic mechanisms underlying thrombosis in APS are not known. However, some hypotheses, such as platelet and endothelial activation or altered coagulation, have been proposed. From a clinical standpoint, APS confers an increased risk of thromboembolic events, including peripheral thrombophlebitis, pulmonary thromboembolism, stroke, retinal artery occlusion, Devic’s syndrome, myocardial infarction, placental thrombosis, Budd syndrome, skin and gastrointestinal disturbances.

Renal involvement is not infrequent in APS, and it comprises different clinical pictures, such as hypertension, renal artery thrombosis/stenosis, renal vein thrombosis, renal infarction, end-stage renal disease and a small-vessel occlusive nephropathy.

In the present case, the patient displayed coagulation and vascular disorders with renal and bowel involvement, indicating a typical systemic development of APS. As soon as the patient began to complain of abdominal pain and the imaging evaluations revealed a picture of diffuse fluid distension of small bowel loops with multiple air-fluid levels, her clinical conditions progressed rapidly toward a systemic organ failure and death.

Autopsy detected a massive intestinal ischemia, which was likely caused by systemic microvascular dysfunction, even if the imaging diagnostic procedures did not reveal the typical signs of intestinal infarction, with particular regard for bowel wall oedema or thickening, pneumatosis intestinalis, irregular wall-enhancement, pneumoperitoneum, mesenteric and/or portal vein gas. Of note, in the setting of intestinal infarction, bowel wall oedema is the most common finding on CT scan. There is consistent evidence in the medical literature to support a pathophysiological relationship between APS and bowel infarction. In this respect, the present case is very peculiar, owing to the lack of positive abdominal imaging, which prevented the diagnosis of a severe condition of intestinal ischemia.

In conclusion, systemic disorders associated with microvascular dysfunctions and high risk of thrombotic events can be complicated by severe organ ischemia, which may escape imaging diagnosis and progress toward extensive infarctions with fatal outcomes.

**Differential Diagnosis List:** Fatal ischemic small bowel infarction in APS.

**Final Diagnosis:** Fatal ischemic small bowel infarction in APS.

**References:**


Description: The supine view (a) displays gastric and small bowel distension, while the colonic frame is gasless. Origin:
Description: Tangential projection (b): evident gas-fluid levels in the small bowel area, no signs of pneumoperitoneum. Origin:
Description: Minimal perihepatic fluid accumulation. Origin:
Description: Inhomogeneous liver parenchyma; inhomogeneous hypodense area located just below the diaphragm and characterized by indistinct margins and dilatation of intrahepatic bile-ducts, suggestive of hepatic abscess. Origin:
Description: Inhomogeneous liver parenchyma; inhomogeneous hypodense area located just below the diaphragm and characterized by indistinct margins and dilatation of intrahepatic bile-ducts, suggestive of hepatic abscess. Origin:
Description: Inhomogeneous liver parenchyma. Origin:
Description: Lung window setting: consolidation area within the lower lobe of right lung in association with scissural thickening. Origin:
Description: Minimal perihepatic and perisplenic fluid accumulations. Origin:
Description: Minimal perihepatic and perisplenic fluid accumulations. Origin:
Description: Minimal perihepatic and perisplenic fluid accumulations. Origin:
Description: Diffuse fluid distension of small bowel loops with multiple gas-fluid levels. Origin:
Description: Regular thickening and contrast enhancement of bowel walls. Origin:
Description: Accentuation of the median abdominal wall hernia. Origin:
Description: Coronal MPR reconstruction confirms diffuse fluid distension of small bowel loops with multiple gas-fluid levels. Origin:
Description: Sagittal MPR reconstruction documents the obstruction of aorto-superior mesenteric bypass. Origin:
Description: MPR reconstruction shows distal revascularization by collateral vessels stemming from the gastroduodenal artery, indicating a chronic occlusion of aorto-superior mesenteric bypass. Origin: