

# Acute pancreatitis due to an attack of acute intermittent porphyria

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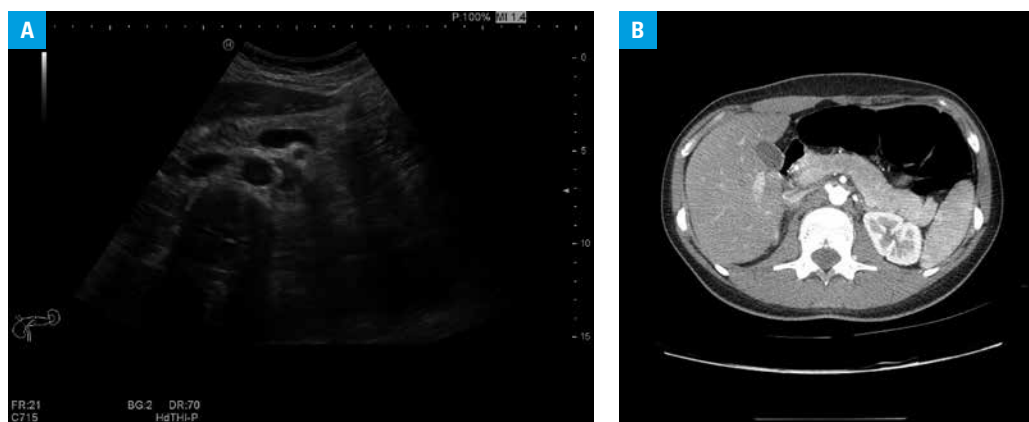
A 25-year-old woman with no chronic diseases was referred to the hospital because of nausea, vomiting, and extremely severe epigastric pain that did not respond to analgesics, even opioids. The symptoms occurred after drinking a glass of red wine 7 days before the admission.

Laboratory tests confirmed elevated levels of pancreatic enzymes: lipase, 410 U/l; serum amylase, 236 U/l; and urine amylase, 1168 U/l. Another abnormality was hyponatremia (sodium level, 124 mmol/l). The inflammatory and cholestatic markers were within the reference range. There was no hypertriglyceridemia or hypercalcemia. The initial diagnosis of acute pancreatitis was established, and the clinical severity of pancreatitis was estimated. There was no evidence of systemic inflammatory response syndrome. The Acute Physiology and Chronic Health Evaluation score (APACHE II) was 2 points.

Imaging studies were performed. An abdominal ultrasound on admission revealed the pancreas of a typical shape, size, and echogenicity. Cholelithiasis or bile duct dilation was not identified, and there was no fluid collection in the pancreatic

region (FIGURE 1A). The normal-appearing pancreas with no peripancreatic abnormalities was confirmed by contrast-enhanced computed tomography (CT) of the abdomen and pelvis (grade A in the Balthazar score, 0 points in the CT severity index, corresponding to mild acute pancreatitis) (FIGURE 1B). Abdominal X-ray revealed colon distension in the region of the splenic flexure (FIGURE 1C). According to the revised Atlanta Classification, acute pancreatitis was evaluated as mild. Cholelithiasis was ultimately excluded by endoscopic ultrasound. There was no evidence of stones in the bile or pancreatic ducts (FIGURE 1D). The normal pancreatic tissue was confirmed by measuring elasticity and strain ratio (0.52% and 1.76, respectively; FIGURE 1E).

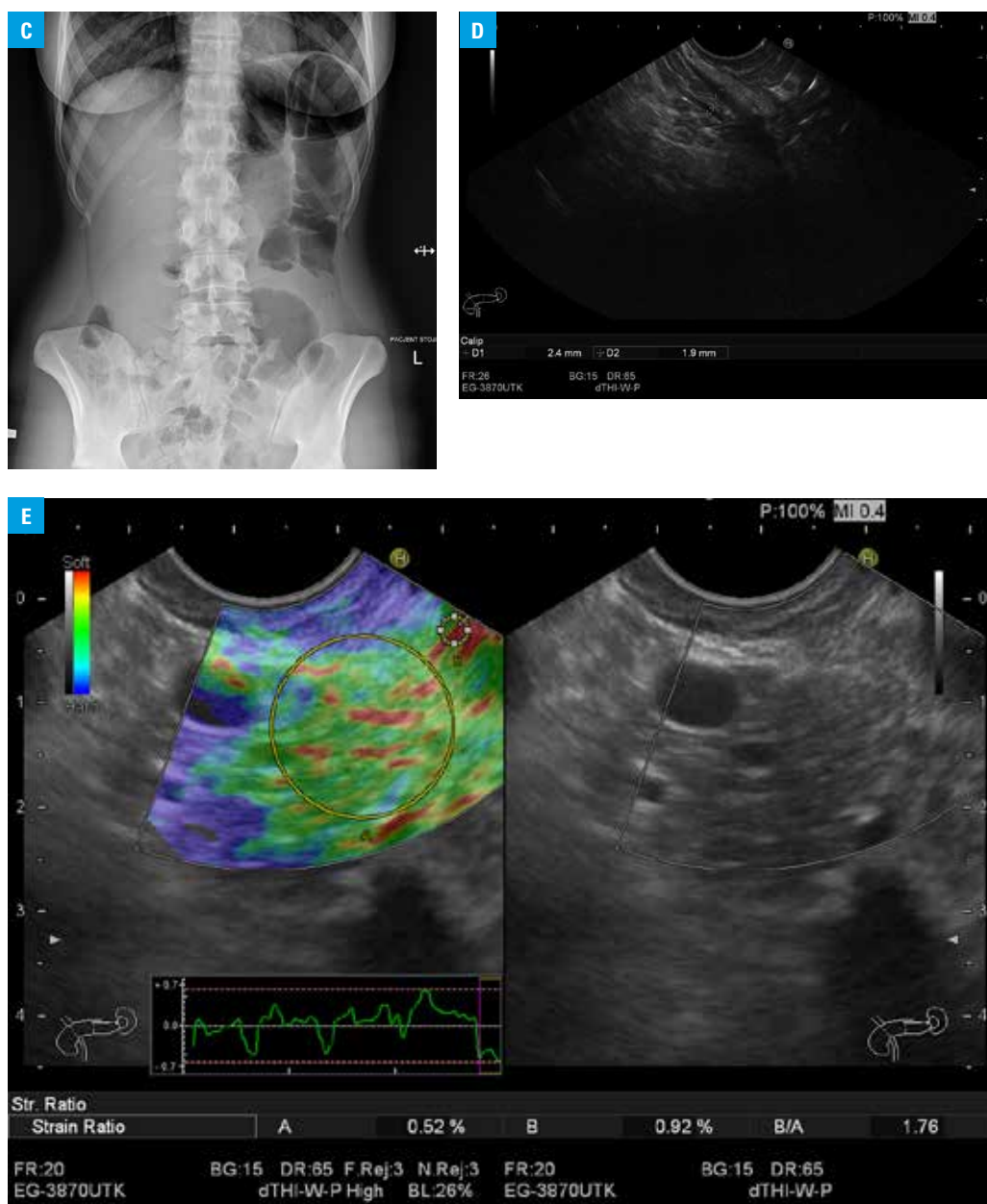
The lack of abnormalities on imaging studies and severe clinical course of the disease with opioid-resistant pain in a young person, accompanied by severe hyponatremia, suggested an attack of acute intermittent porphyria. The laboratory measurements confirmed increased urinary levels of porphobilinogen (PBG) and  $\delta$ -aminolevulinic acid (ALA). A supportive



**FIGURE 1** Imaging of acute pancreatitis due to an attack of acute intermittent porphyria  
**A** – ultrasound image of the pancreas; **B** – contrast-enhanced computed tomography scan of the pancreas

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**FIGURE 1** Imaging of acute pancreatitis due to an attack of acute intermittent porphyria **C** – abdominal X-ray scan showing colon distension due to acute pancreatitis; **D** – endoscopic ultrasound image of the common bile duct (D1) and major pancreatic duct (D2); **E** – elasticity (A) and strain ratio (B/A) of the pancreas



treatment with intravenous glucose solution (10%) and sodium supplementation was introduced, with no effect. Pain relief and normalization of laboratory parameters were achieved after implementation of hemin therapy.

Acute intermittent porphyria is a hereditary defect of the heme production. The enzyme deficiency in the biosynthetic pathway results in the accumulation of the heme precursors in blood (PBG, ALA). Alcohol intake, infection, and hormonal changes in the menstrual cycle may trigger an attack. Its symptoms are caused by neurotoxicity of porphyrin precursors. A typical clinical presentation includes hyponatremia (due to the syndrome of inappropriate antidiuretic hormone secretion) and abdominal pain (due to ALA and PBG toxicity for visceral nerves).

Acute pancreatitis has been described in several case reports. The mild evidence of acute pancreatitis on imaging studies could be explained by the impaired neurological processes related to the activity of porphyrin metabolites.<sup>1-3</sup> Porphyria

should be considered in a differential diagnosis of acute pancreatitis, especially in young people when the cause is undefined. Hyponatremia is an abnormality that can suggest porphyria. The recognition of acute pancreatitis in the course of an attack of acute intermittent porphyria is essential to provide a proper treatment.

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