CLINICAL IMAGE

Pancreatic tumor of an autoimmune origin

Michał Holecki, Anna Hawrot-Kawecka, Anna Gasidło, Dorota Strzałkowska, Jan Duława

Department of Internal Medicine and Metabolic Diseases, Medical University of Silesia, Katowice, Poland

A 64-year-old male patient with a pancreatic tumor of unknown origin and retroperitoneal fibrosis was admitted to our department because of severe abdominal pain with ileum symptoms lasting 2 days. The patient's complaints included periodic, progressing abdominal pain, loss of appetite, weight reduction (20 kg within 7 months), and eye dryness. The pancreatic tumor was diagnosed in February 2013 after 5 pancreatic biopsies had been performed. Histopathological findings revealed inflammatory cells but not cancer cells. On admission, the patient presented with cachexia (body mass index, 19 kg/m^2), dehydration, diffuse abdominal pain, and lack of bowel movements. The Blumberg's sign was negative. Abdominal X-rays showed distended intestinal loops with fluid levels. A routine laboratory test revealed a decreased hemoglobin level (8.5 g/dl),

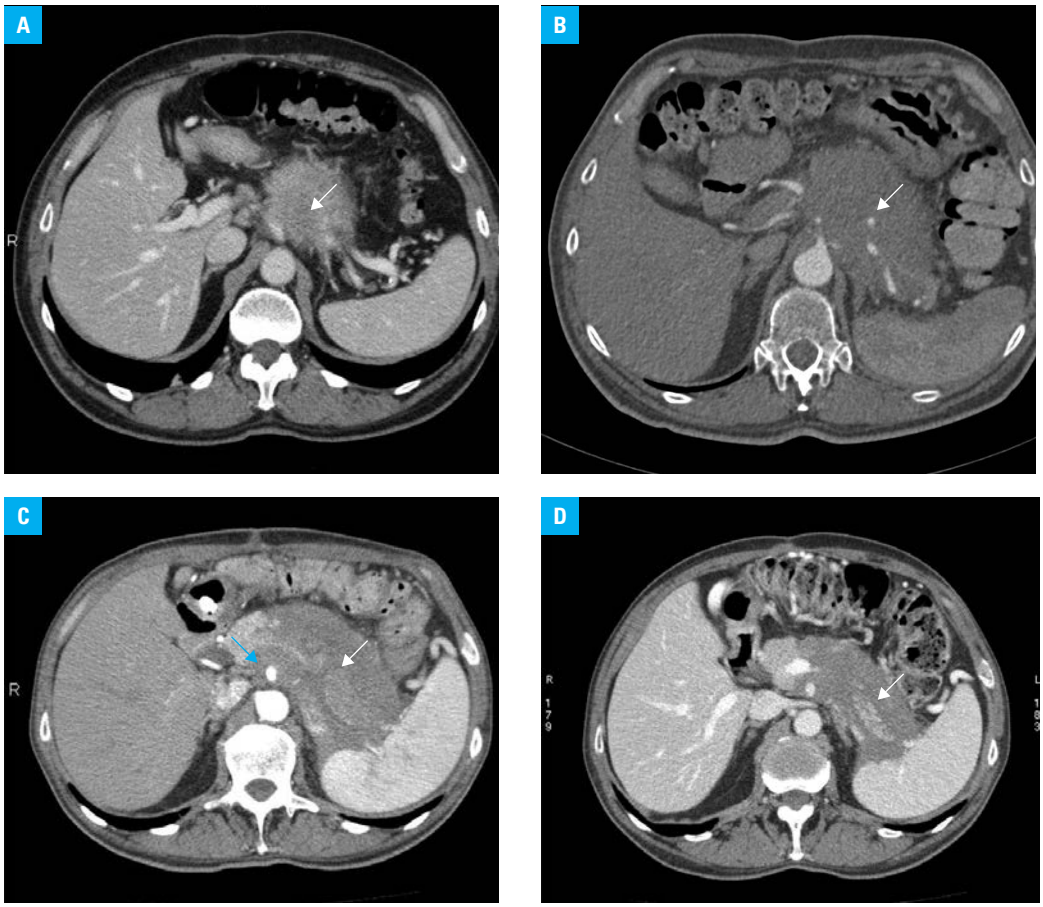

FIGURE 1 **A** – abdominal computed tomography (CT) performed in February 2013: focal pancreatic enlargement (white arrow); **B** – abdominal CT performed in April 2013: diffuse pancreatic enlargement (white arrow); **C** – abdominal CT performed in September 2013: pancreatic enlargement with a capsule-like rim (white arrow), retroperitoneal fibrosis (blue arrow); **D** – abdominal CT performed in November 2013: regression of pancreatic enlargement after 6 weeks of steroid therapy (white arrow)

Correspondence to:
Michał Holecki, MD, PhD,
Klinika Chorób Wewnętrznych
i Metabolicznych, Śląski Uniwersytet
Medyczny, ul. Ziołowa 45/47,
40-635, Katowice, Poland, phone:
+48-32-359-82-23,
fax: +48-32-252-35-93,
e-mail: holomed@gmail.com
Received: December 6, 2013.
Revision accepted:
December 6, 2013.
Published online:
December 20, 2013.
Conflict of interest: none declared.
Pol Arch Med Wewn. 2014;
124 (1-2): 70-71

hypokalemia (3.3 mmol/l), and hyponatremia (133 mmol/l), increased serum C-reactive protein (128 mg/l), γ-glutamyltransferase (290 U/l), and alkaline phosphatase (294 U/l). Serum γ-globulin levels were increased (27%) with elevated immunoglobulin G4 (IgG4) levels (2.84 g/l; normal range, 0.03–2.01). Abdominal computed tomography (CT) performed in February (FIGURE 1A), April (FIGURE 1B), and on admission in September 2013 (FIGURE 1C) showed tumor enlargement (from 51 × 52 × 55 mm to 74 × 70 × 70 mm) with increasing infiltration of the stomach wall, aorta, inferior vena cava, and ureters, dilated intrahepatic bile ducts and retroperitoneal fibrosis. No enlargement of intra-abdominal lymph nodes or focal changes in the liver were observed. Considering the clinical course (duration of the disease, patient's clinical status, laboratory tests, histopathology, and imaging), autoimmune pancreatitis (AIP) was suspected. The patient received steroid therapy (intravenous methylprednisolone, 100 mg/d for 3 consecutive days, followed by oral prednisone, 60 mg/d), antibiotics, analgesics, and spasmolytics. The patient's condition improved within a few days and pain relief and normal bowel movements were observed. After a 6-week course of oral prednisone, the patient reported only rare incidents of weak abdominal pain and weight gain (8 kg). Abdominal CT revealed a decreased size of the pancreatic tumor, regression of infiltration on the vena cava inferior, ureters, and decreased distention of the bile duct (FIGURE 1D). We observed a decrease in C-reactive protein levels (<5 mg/l), increase in hemoglobin levels (12.4 g/dl), and normalization of liver enzymes. The patient's condition markedly improved and AIP was finally diagnosed.

AIP is a rare disorder, which was first described in 1961 in a patient with pancreatitis associated with hypergammaglobulinemia.[1] It is a typical manifestation of an IgG4-related disease but it can mimic pancreatic cancer.[2] The most characteristic radiological feature of AIP is the focal pancreatic enlargement with a capsule-like rim (FIGURE 1C).[3] In some cases, a delayed homogenous enhancement, irregular narrowing of the main pancreatic duct, and a stricture of the common bile duct may be observed.[4]

The diagnosis may be difficult and requires a combination of clinical (obstructive jaundice), serological (raised IgG4 level), morphological (CT), and histopathological (inflammation) features. We described this case to remind about this rare disease, which can be extremely important in a differential diagnosis, treatment, and prognosis of a patient with a pancreatic tumor.

REFERENCES

1 Sarles H, Sarles JC, Muratore R, et al. Chronic inflammatory sclerosis of the pancreas – an autonomous pancreatic disease? Am J Dig Dis. 1961; 6: 688-698.

2 Kamisawa T, Takuma K, Egawa N, et al. Autoimmune pancreatitis and IgG4-related sclerosing disease. Nat Rev Gastroenterol Hepatol. 2010; 7: 401-409.

3 Hiroyuki I, Hiroshi H, Shingo B, et al. Autoimmune pancreatitis: CT and MR characteristics. AJR. 1998; 170: 1323-1327.

4 Sun GF, Zuo CJ, Shao CW, et al. Focal autoimmune pancreatitis: Radiological characteristics help to distinguish from pancreatic cancer. World J Gastroenterol 2013; 19: 3634-3641.