

Pancreatic mass as an initial presentation of severe Wegener's granulomatosis

Yana Valerieva, Branimir Golemanov, Nadezhda Tzolova, Rumiana Mitova

University Hospital Queen Joanna-ISUL Sofia, Bulgaria

Abstract

Acute pancreatitis or a pancreatic mass is a very rare initial presentation of Wegener's granulomatosis. A 62-year-old woman presented with tumor-like pancreatitis and otitis media. Abdominal ultrasound and magnetic resonance suggested the presence of pancreatic tumor. Ultrasound-guided fine needle aspiration was negative. Distal pancreatic resection and splenectomy were performed and histopathology proved Wegener's vasculitis of the pancreas and spleen. Azathioprine and steroids were subsequently started and six months later the patient was asymptomatic. Involvement of the pancreas could be considered as a presenting symptom of Wegener's vasculitis.

Keywords Wegener's granulomatosis, vasculitis, pancreatic mass, pancreatitis

Ann Gastroenterol 2013; 26 (3): 267-269

Introduction

Wegener's granulomatosis (WG) is ANCA-positive necrotizing granulomatous and pauci-immune vasculitis of the small- and medium-sized blood vessels with a predilection for respiratory tract and kidney involvement. Association of WG with acute pancreatitis or a pancreatic mass, mimicking a tumor is very rare, especially as an initial presentation of severe disease. Diagnosis could be challenging and the disease might potentially lead to a disastrous outcome.

Case report

A 62-year-old woman was admitted to the hospital with ten-day history of acute epigastric pain, radiating to the back and associated with nausea, persistent fever up to 38°C and headache. She had a history of cough and sinusitis a month ago, complicated with otitis media, with insignificant improvement on antibiotic treatment and antitussive drugs. The patient had no other significant past and family history except laparoscopic cholecystectomy for lithiasis 15 years ago. She denied alcohol use, herbal or over-the-counter medications and allergies.

Examination revealed epigastric tenderness without rebound and no palpable organomegaly, peristalsis was normal. There was purulent discharge from the left ear, pain in maxillary sinus and left mastoid. No other pathological findings were present.

The initial laboratory tests showed slight leukocytosis, thrombocytosis, normal ESR, CRP 15.8 mg/dL (normal range <0.6 mg/dL), elevated fibrinogen, ALT and GGT were up to 2xURL (upper reference level) and 3xURL respectively. Serology for HBV and HCV infection was negative. Serum amylase and lipase, cholesterol and triglycerides were within normal range. Albumin was 2.95 g/dL; blood glucose was 148 mg/dL. Serum iron (8.4 µg/dL) and TIBC (38 µmol/L) were low. Hemoglobin was normal at admission to hospital but dropped to 8.8 g/dL within a week without signs of bleeding and prothrombin time also decreased to 53%. Urinalysis showed increased numbers of leukocytes and erythrocytes with proteinuria 0.276 g/L/24h. Blood cultures and urine cultures were sterile. Coombs and Schirmer tests were negative. Serum IgG, IgA, IgM and serum tumor markers (CA 19-9, CEA) were within normal range. RF (rheumatoid factor), ANA (antinuclear antibodies) and anti-MPO (anti-myeloperoxidase) were tested negative, while anti-Pr3 (anti-proteinase 3, c-ANCA) were 15xURL (89.63 U/mL; negative <6 U/mL).

Initial abdominal ultrasound (US) revealed slightly enlarged and hypoechoic pancreatic body and tail with blurry margins, as in acute pancreatitis. Computed tomography of the abdomen also confirmed edema of the pancreatic tail without fluid collections or other abnormal findings. Control US in several days showed blight echoes scattered among the enlarged pancreas, compression of the splenic vein and splenomegaly. Further magnetic resonance was performed and revealed a 3 cm soft-tissue formation in the pancreas tail without pancreatic duct abnormalities, which compressed the

Clinical Centre of Gastroenterology, University Hospital, "Queen Joanna - ISUL", Sofia, Bulgaria

Conflict of Interest: None

Correspondence to: Yana Valerieva, MD, 8 Byalo More Street, Clinical Centre of Gastroenterology, University Hospital "Queen Joanna - ISUL", Sofia, Bulgaria, e-mail: yana_valerieva@abv.bg

Received 11 December 2012; accepted 22 February 2013

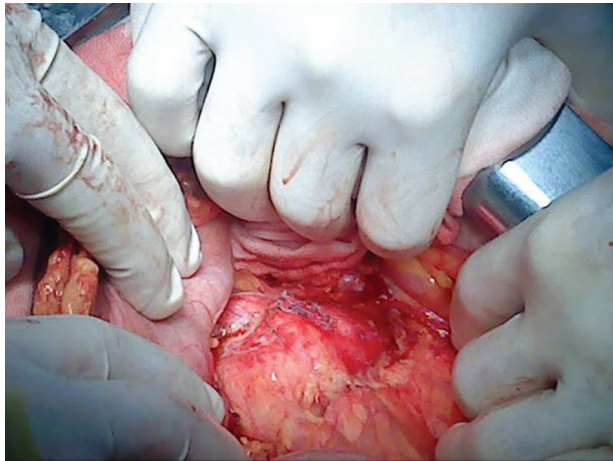


Figure 1 A solid tumor-like formation in the pancreatic body and tail

splenic vein, without infiltration or enlarged regional lymph nodes. Imaging studies of the kidneys showed no alterations and chest X-ray was normal.

The initial diagnosis was acute pancreatitis but normal levels of serum amylase and lipase in the course of the disease, normal biliary tree imaging and lack of pancreatic disease history or alcohol consumption did not support this diagnosis. Normal immunoglobulin levels and absence of pancreatic duct abnormalities ruled out autoimmune pancreatitis, but positive c-ANCA suggested the possibility of other autoimmune diseases. However, pancreatic neoplasm was also suspected because of the presence of pancreatic tail mass. US-guided fine needle aspiration was performed twice but only detritus without atypical cells was established. However malignancy could not be entirely ruled out.

Despite treatment with antibiotics, IV fluids, proton pump inhibitors and analgesia for a week, fever and severe abdominal pain still persisted. On the fifth day of hospitalization the patient suffered phlebothrombosis of left lower

leg and low-molecular-weight heparin was added to therapy. Immunosuppressive treatment was discussed as an option, but the severe intractable abdominal pain and lack of cytological confirmation along with the possibility of pancreatic carcinoma indicated surgery. A solid tumor-like formation in the pancreatic body and tail with well-defined margins was resected and pancreato-jejunoanastomosis a modo Roux and splenectomy were performed (Fig. 1).

The histology from the surgical specimens confirmed WG of the pancreas and spleen – typical vasculitis with fibrinoid necrosis, granulomas and giant cells (Fig. 2).

The patient was further treated with steroids and azathioprine and a clinical remission followed. She was doing well at 6 months of follow up.

Discussion

WG is a rare systemic ANCA-associated vasculitis with a broad spectrum of clinical presentations. Severe disease manifests with lungs, kidneys, nasal mucosa and other organ involvement and has poor prognosis if no appropriate immunosuppressive treatment is constituted [1].

WG is a relatively rare disease, with a slight male predominance, which usually presents at the age of 40-50. ANCA (PR3) is positive in 90% of patients with WG and its level corresponds to the disease activity [2].

Gastrointestinal manifestations and concomitant spleen involvement of WG are less common, announced predominantly in case reports [3]. Pancreatic disease as an initial syndrome of necrotizing vasculitis is even rarer and presents as a pancreatic mass or acute pancreatitis [4]. In most cases diagnosis is significantly delayed and based on histology of other affected organs rather than pancreas. Most patients who presented initially with pancreatic disease had severe course and later developed significant complications. In our case, the diagnosis was even more challenging because of a previous

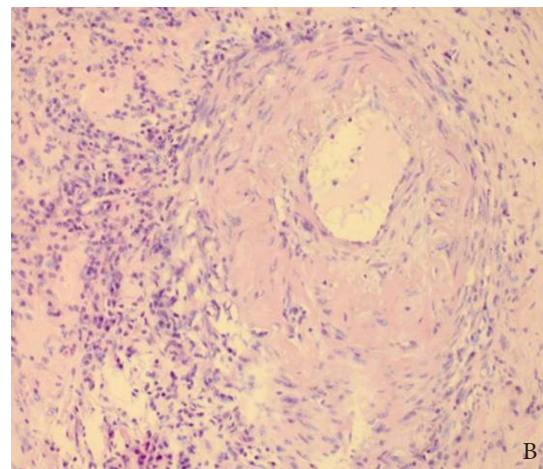
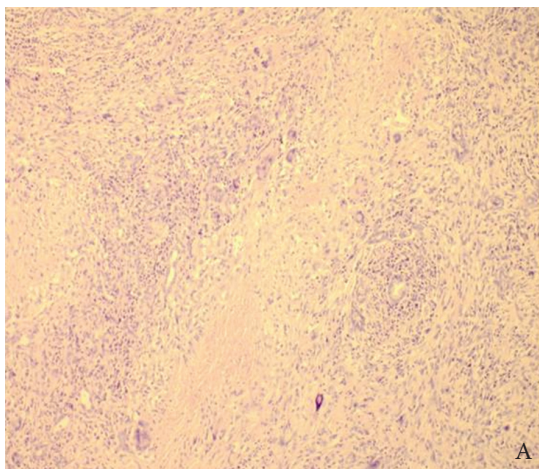


Figure 2 (A) Blood vessel with fibrinoid necrosis (B) Granuloma with giant cells

history of biliary disease and cholecystectomy. Laboratory and imaging studies pointed against acute pancreatitis or may have suspected autoimmune pancreatitis but could not entirely rule out pancreatic neoplasm. Diagnosis was based on clinical suspicion of a systemic autoimmune disorder, positive c-ANCA and histopathological confirmation of granulomatous vasculitis of the pancreas and spleen [5].

This case illustrates the importance of taking into account a possibility of vasculitis in the etiology of pancreatic diseases when most common causes are already ruled out, so that appropriate treatment may be started early in the course of the disease.

References

1. Fauci AS, Haynes BS, Katz P, Wolff SM. Wegener's granulomatosis: prospective clinical and therapeutic experience with 85 patients for 21 years. *Ann Intern Med* 1983;**98**:76-85.
2. Finkelman JD, Lee AS, Hummel AM, et al. ANCA are detectable in nearly all patients with active severe Wegener's granulomatosis. *Am J Med* 2007;**120**:643.e9-e14.
3. Pinkney JH, Clarke G, Fairclough PD. Gastrointestinal involvement in Wegener's granulomatosis. *Gastrointest Endosc* 1991;**37**:411-412.
4. Chawla S, Atten MJ, Attar BM. Acute pancreatitis as a rare initial manifestation of Wegener's granulomatosis. A case based review of literature. *JOP* 2011;**12**:167-169.
5. Leavitt RY, Fauci AS, Bloch DA, et al. The American College of Rheumatology 1990 criteria for the classification of Wegener's granulomatosis. *Arthritis Rheum* 1990;**33**:1101-1107.