Primary angiosarcoma of the pancreas mimicking severe acute pancreatitis - Case report

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Abstract

Primary angiosarcoma of the pancreas is an extremely rare neoplasm that often mimicks severe acute pancreatitis.

A 58-year-old man was admitted with clinical and laboratory signs of severe acute pancreatitis. Contrast enhanced CT scan demonstarted haemorrhagic necrotizing inflammation of the pancreas involving the pancreatic tail, splenic hilum and small bowels with multiple peripancreatic and free abdominal fluid collection. Percutaneous drainage was performed. After 13 days, laparotomy was indicated because of persistent intra-abdominal bleeding, fever and a palpable, rapidly growing mass in the left upper quadrant of the abdomen. During the operation a necrotic, haemorrhagic mass was found in the pancreatic tail; a frozen section showed malignancy, although the tumour was unresectable. Despite all conservative and surgical therapeutic attempts, the patient died within four weeks after diagnosis. Final histology justified primary angiosarcoma of the pancreas.

If a patient with signs of severe acute pancreatitis has fever without elevated PCT, the presence of a malignant tumor of the pancreas should be considered.

Key words: acute pancreatitis, cancer of pancreas, malignant mesenchymal tumour

Introduction

Primary sarcomas of the pancreas are extremely rare, they represent 0.1 % of pancreas malignancies[1]. The most often reported cases are leiomyosarcomas[2]. Only two cases of primary pancreatic angiosarcoma has been reported so far[3,4]. Because of the small number of cases published, primary sarcomas of the pancreas are not well described. They often mimick signs of acute pancreatitis, making differential diagnosis difficult[5].

The authors report a case of a primary angiosarcoma involving the pancreatic tail in a patient admitted for severe acute pancreatitis.

Case report:

A 58-year-old man was admitted to hospital with two weeks' history of upper abdominal pain, nausea and fever after excessive food intake. He denied alcohol or drug abuse, toxic explosion or smoking. He mentioned weakness, weight loss (1-2 kg) within a few weeks and dark stool.

During the physical examination, the patient complained of pain in the left upper quadrant. The abdomen was distended, but there was no tenderness. Slow bowel movement was auscultated and a moderate amount of free abdominal fluid was found during percussion.

Laboratory tests were significant for C- reactive protein (CRP) (87 mg/L), his haemoglobin level was 10.6 g/dL. Other parameters including procalcitonin (PCT), cancer antigen (CA) 19-9, carcinoembryogenic antigen (CEA), alfa-foetoprotein (AFP) were not elevated. Amylase and lipase levels were also norma lat admission, two weeks after the onset of symptoms.

Because of possible gastrointestinal bleeding (melaena), endoscopy (gastro-duodenoscopy and colonoscopy) was performed, which failed to detect the cause of anaemia. Abdominal ultrasonography (US) and contrast enhanced computed tomography (CT) scan demonstarted the signs of acute haemorrhagic necrotizing pancreatitis with multiple peripancreatic fluid collection involving the pancreatic tail, splenic hilum and small bowels. A large amount of free abdominal fluid was detected. [Figures 1. a-c]

Based on the clinical course of the disease and imaging studies, the diagnosis of severe acute pancreatitis was made (haemorrhage of the pancreas being the suspected cause of anaemia). Conventional conservative treatment was started with enteral feeding through a nasojejunal tube. CT-guided percutaneous drainage of intra-abdominal and peripancreatic fluid was performed. The diagnosis of pancreatitis was confirmed by laboratory investigation of the drained abdominal fluid, which showed highly elevated pancreatic enzymes (amylase and lipase).

After 13 days, the amount of the drained, bloody abdominal fluid persisted and the patient required repeated blood transfusions. He had fever, and a rapidly growing palpable mass developed in the left upper quadrant of his abdomen. CRP level increased to 131,8 mg/L, PCT level remained within normal limits (0,26 mg/L).

On laparotomy a large, haemorrhagic mass was found occupying the pancreatic tail and mesocolon. Necrosectomy and peripancreatic drainage were performed. Because of the possible malignancy, lymph nodes from the gastrocolic ligament and necrotic tissue from the panceras were resected and taken for histopathology.

The histological examination of the specimen revealed angiosarcoma with epithelial and vascular differentiation arising from the pancreas. Microscopically neoplastic cells were found immunhistochemically positive for vimentin, CD31, cytokeratin, factor XIII, but negative for CD34 and factor VIII. (Figures 2. a-d)

On the third postoperative day, despite conservative therpy, the patient developed abdominal compartment syndrome with intra-abdominal pressure at 50 mmHg and renal failure, so decompressive laparotomy was indicated. During the operation, a large amount of haemorrhagic ascites and paralytic ileus was found. Two days later, the patient unfortunately died of multiorgan failure (MOF) and sepsis.

Eventually, the patient died within four weeks after the clinical diagnosis of acute pancreatitis. Final histology revealed angiosarcoma arising from the pancreas with the histological signs of acute haemorrhagic pancreatitis. The parenchyma of the pancreas was almost undetectable, it was replaced by tumour mass and necrotic, haemorrhagic tissue. The tumour was complicated by pneumonia, sepsis, respiratory and circulatory insufficiency, which led to the patient's death. No metastasis was confirmed. The tumor grew rapidly.

Discussion

Sarcomas of the pancreas are very aggressive, rapidly growing tumours with a great tendency of metastasis and a very poor prognosis[5-8]. The symptoms are unspecific and might include weight loss, upper quadrant pain, abdominal distension, nausea, vomiting, fever, jaundice, gastrointestinal or intra-abdominal bleeding and paraneoplastic signs[2,5,7-10,14]. Similarly to our case, angiosarcomas may cause life-threatening bleeding or chronic anaemia[3,9,11,14].

Physical examination and laboratory tests are unremarkable, the tumour markers are not elevated. The imaging modalities (US and CT scan) are not specific to sarcomas, endoscopic ultrasound-guided fine-needle aspiration biopsy (EUS-FNAB) can be a possible method of diagnosis[9]. Angiography or Tc 99m-labeled red blood cells can be helpful in detecting of the origin of gastrointestinal bleeding and diagnosing angiosarcomas[12,14].

Because of the rapid growth the tumour is often detected at an advanced stage. The differential diagnosis with carcinomas can be difficult[10,11]. The final diagnosis is based on histological and immunohistochemical staining. Angiosarcomas often show epitheloid cytomorphology[6,10] and have a wide range of histological appearance which is important for diagnosis but not for prognosis[8].

The majority of primary angiosarcomas are reported in the spleen. Only two cases of primary pancreas angiosarcoma have been published so far[3,4].

Surgery has been the only known option to treat localised disease[3,5,7]. In disseminated cases, however, chemotherapy may be worth trying[13].

Survival is very poor with a mean of 3-11 months[6,8,10].

In conclusion angiosarcoma of the pancreas is a very rare entity therefore it is not well described. If a patient presents with signs of severe acute pancreatitis and fever but without elevated PCT, the presence of a malignant tumour in the pancreas should be considered.

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Figs. 1. a-c. Contrast enhanced CT scan showed signs of acute necrotizing pancreatitis with free abdominal and peripancreatic fluid collection.

Figs. 2. a-d. Immunhistochemical staining revealed a.) CD31, b.) cytokeratin, c.) FXIII, d.) vimentin positivity (pictures by Csonka T M.D., Bidiga L M.D., University of Debrecen, Department of Pathology).