Pleomorphic sarcoma of the pancreas – case report

Mięsak pleomorficzny trzustki – opis przypadku

Zbigniew Kamocki[1], Andrzej Roszkowski[1], Andrzej Kemona[2], Bogusław Kedra[1]

[1]2nd Department of General and Gastroenterological Surgery, Medical University of Bialystok, Poland
[2]Department of General Pathomorphology, Medical University of Bialystok, Poland

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Słowa kluczowe: pleomorficzny, mięsak, pankreatektomia, trzustka.

Address for correspondence: Andrzej Roszkowski MD, PhD, 2nd Department of General and Gastroenterological Surgery, Medical University of Bialystok, 24 A M. Sklodowskiej-Curie St, Bialystok, Poland, phone: +48 502 374 653, e-mail: andrzejroszko@o2.pl

Abstract

Primary mesenchymal tumors of the pancreas are extremely rare. Sarcoma-type lesions constitute about 1% of all malignant neoplasms. Cases have been described in world literature as: leiomyosarcoma, fibrosarcoma, carcinosarcoma, rhabdomyosarcoma, liposarcoma, hemangioendothelioma, malignant neurilemoma, and malignant fibrous histiocytoma. We report a case of a 75-year-old woman with a tumor described as a postinflammatory cyst, despite lack of acute pancreatitis in anamnesis. The patient was treated operatively. Intraoperatively we found an orange-sized pancreatic tumor, infiltrating the back wall of the stomach and responsible for portal hypertension. The tumor together with the distal 2/3 of the stomach and spleen, pancreatic body and tail was resected. The pathologist diagnosed: pleomorphic sarcoma, probable histiocytic fibrosarcoma.

Streszczenie

Pierwotne nowotwory mezenchymale trzustki występują niezwykle rzadko. Zmiany o typie mięsaka stanowią około 1% wszystkich nowotworów złośliwych. W piśmiennictwie światowym opisano kilkaset przypadków m.in.: *leiomyosarcoma*, *fibrosarcoma*, *carcinosarcoma*, *rhabdomyosarcoma*, *liposarcoma*, *hemangioendothelioma*, *malignant neurilemoma*, *malignant fibrous histiocystoma*. W pracy przedstawiono przypadek 75-letniej kobiety przyjętej z powodu opisywanej w badaniach obrazowych pozapalnej torbieli trzustki. Chorą operowano. Śródoperacyjnie stwierdzono guz trzustki wielkości pomarańczy naciekający tylną ścianę żołądka i powodujący nadciśnienie wrotne. Guz usunięto wraz z dystalną częścią żołądka, śledzioną oraz trzonem i ogonem trzustki. Rozpoznanie patomorfologiczne: *sarcoma pleomorphicum*, *probabiliter fibrosarcoma histiocyticum*.

Introduction

Primary mesenchymal tumors of the pancreas are extremely rare. Sarcoma-type lesions constitute about 1% of all malignant neoplasms. Cases have been described in world literature as: leiomyosarcoma, fibrosarcoma, carcinosarcoma, rhabdomyosarcoma, liposarcoma, hemangioendothelioma, malignant neurilemoma, and malignant fibrous histiocytoma [1-6].

Case report

We report a case of a 75-year-old woman with a history of cholecystectomy and hysterectomy, with arterial hypertension and type 2 diabetes, who was operated on in the 2nd Department of General and Gastroenterological Surgery, Medical University of Bialystok, Poland.

The patient complained of strong paroxysmal epigastric pain lasting for several months, radiating to the spine. The patient felt weak, was subfebrile and had lost about 10 kg within the last 2 months. In the past the patient underwent cholecystectomy, hysterectomy as well as excision of a tumor of the urinary bladder. Additionally she suffered from arterial hypertension and type 2 diabetes. Physical examination at admission revealed tenderness in the middle abdomen and left epigastrium with palpable resistance.

Laboratory analyses: elevated white blood count and C-reactive protein. Tumor markers CA-19-9 and CEA were not increased. Ultrasound scan of the abdomen revealed a hypoechogenic tumor with liquid content (62 mm × 44 mm). The tumor was described as a postin-

flammatory cyst, despite lack of acute pancreatitis in anamnesis. The result of computed tomography (CT) confirmed the liquid lesion located at the junction of the pancreatic body and tail. The capsule of this tumor showed contrast enhancement. The cyst was adjacent to the back wall of the stomach. There were no regional lymph nodes visible and blood vessels seemed not to be infiltrated. The computed tomography result was interpreted probably as a postinflammatory cyst, although malignancy was not excluded. The other abdominal organs were within the normal range.

It was decided to operate on the patient. Intraoperatively we found an orange-sized pancreatic tumor, infiltrating the back wall of the stomach and responsible for portal hypertension. Inside the left lobe of the liver we found a hard walnut-sized tumor – probably a metastasis. The spleen showed features of secondary splenomegaly. The neoplastic infiltration involved the head of the pancreas, portal vein and superior mesenteric vessels. The tumor together with the distal 2/3 of the stomach and spleen, pancreatic body and tail was resected. At the level of the portal vein the infiltration was sharply divided. The pancreatic stump was closed with single sutures. The continuity of the upper alimentary tract was restored according to Hofmeister-Finsterer. The specimen was sent for pathology. The immediate postoperative period was not complicated.

The pathologist diagnosed: pleomorphic sarcoma, probable histiocytic fibrosarcoma G3. T4 N2 M1.

Discussion

Pancreatic tumors remain a significant clinical problem. Carcinomas are the most common pancreatic malignancies, while the frequency of other tumor types

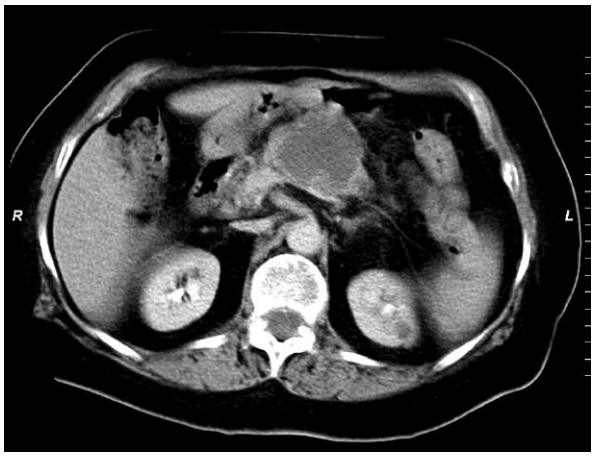

Fig. 1. Huge tumor of the body of the pancreas. Computed tomography scan shows possible neoplastic infiltration of the stomach wall
Ryc. 1. *Olbrzymi guz trzonu trzustki. W obrazie tomografii komputerowej podejrzenie nacieku nowotorowego ściany żołądka*

does not exceed 2%. Pleomorphic sarcoma is a very rare primary pancreatic tumor. Only one case of this malignancy has been reported in the literature. In our patient as well as in the case described by Laverdiere *et al.*, the tumors were detected in women between 60 and 70 years old and were not associated with episodes of acute pancreatitis. Additionally, both patients had a history of laparoscopic cholecystectomy. Intermittent epigastric pain lasting for several months was the initial manifestation reported by both patients. A pancreatic mass, described as a post-inflammatory cyst, was observed on ultrasonography and CT in our patient. Akin to the case reported by Laverdiere *et al.* [4], our patient

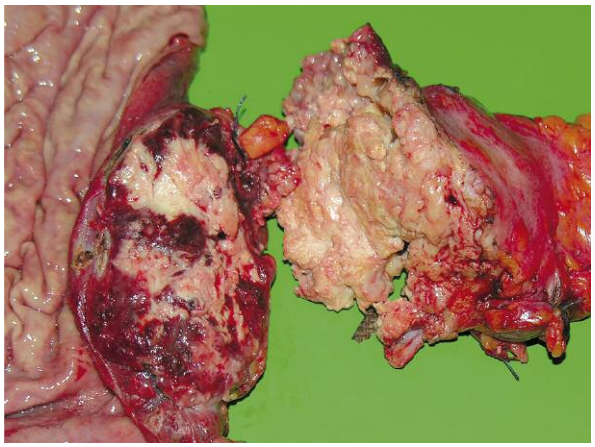

Fig. 2. Postoperative picture of primary pleomorphic sarcoma of pancreas
Ryc. 2. *Preparat pooperacyjny pierwotnego mięsaka trzustki*

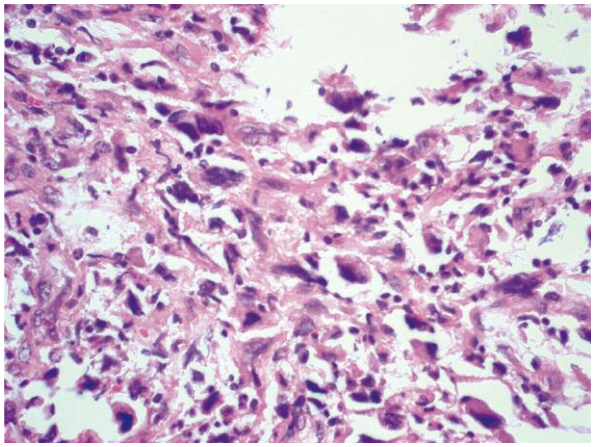

Fig. 3. Sarcoma (H + E stain, field 600×). Remarkable anaplasia of neoplastic cells
Ryc. 3. *Mięsak (barwienie H + E, powiększenie 600×). Uwagę zwraca anaplazja komórek nowotworowych*

underwent surgery because of existing symptoms rather than due to suspected malignancy. Laverdiere *et al.* decided to create a Roux-en-Y cystojejunostomy [4] to facilitate internal drainage of the cyst. Pleomorphic pancreatic sarcoma was revealed by postoperative histopathological examination. The patient was prescribed adjuvant chemotherapy. After 1 year of follow-up and two courses of chemotherapy local recurrence of sarcoma was diagnosed on CT along with liver metastases. The patient died 1 year after the surgery (and diagnosis of pleomorphic sarcoma). In contrast to Laverdiere *et al.*, our patient was qualified for planned laparotomy with peripheral pancreatectomy due to symptoms of compression and despite observed infiltration of the portal system. The tumor was resected radically, along with the infiltrated stomach wall, pancreas body and tail, and the spleen. Despite CT findings the tumor did not compress the portal vein. No morbidity was observed during the postoperative period.

In our opinion, pancreatic lesions that are described as cysts on abdominal CT can mimic sarcoma or other rare malignancies in cases with no history of acute pancreatitis. In suspected cases, laparotomy is a procedure of choice for definitive diagnosis. Only laparotomy followed by histopathological examination provides proper diagnosis and gives an opportunity of successful radical treatment.

In conclusion, we have presented this case because of the very rare occurrence of primary pleomorphic sarcoma of the pancreas.

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