DOI: 10.7860/JCDR/2016/21767.9086

Case Report

Surgery Section

Spontaneous Rupture of the Main Pancreatic Duct Synchronous With a Multi-Focal Microscopic Pancreatic Adenocarcinoma: A Case Report

SHIRLY SHOHAT[1], KATERINA SHULMAN[2], BORIS KESSEL[3], OREN GAL[4], ABDEL RAUF ZEINA[5]

ABSTRACT

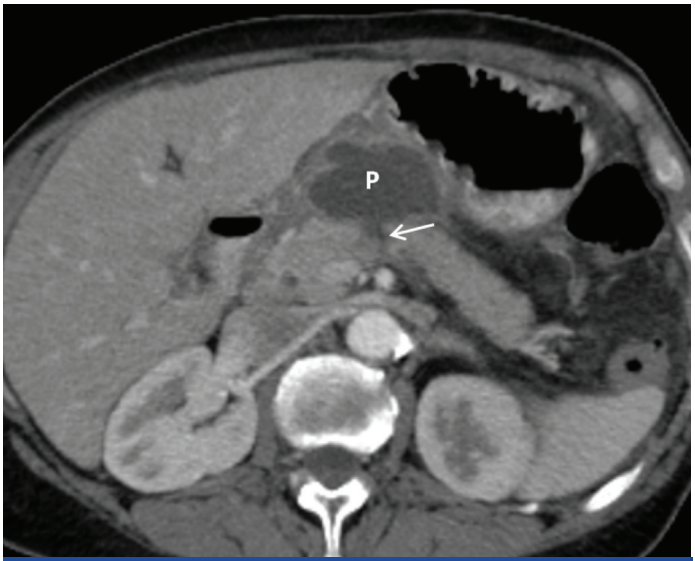
Pancreatic cancer is one of the most lethal types of malignant tumours, commonly diagnosed at an advanced stage. The only curative treatment for this fatal disease is surgery and early diagnosis is the key to a better outcome and prognosis. In this case report we present a 57-year-old woman presenting to the emergency room with abdominal pain and weight loss. Computer Tomography (CT) imaging showed a rupture of the main pancreatic duct and a peri-pancreatic fluid collection with no evidence of a pancreatic mass. An Endoscopic Ultrasound (EUS) guided Fine Needle Aspiration (FNA) did not show any malignant cells and Carcinoembryonic Antigen (CEA) and Carbohydrate Antigen (CA) 19-9 markers were in the normal range. The patient then underwent pancreatectomy that revealed multiple microscopic foci of pancreatic adenocarcinoma with evidence of massive perineural and vascular invasion.

Keywords: Early detection, Pancreatic cancer, Pancreatectomy, Wirsung rupture

CASE REPORT

A 57-year-old woman with a medical history of percutaneous coronary intervention was admitted to the emergency room complaining of a sharp, epigastric abdominal pain, radiating to the back that had been gradually worsening over the previous 3-4 months. She also suffered a 16kg weight loss over that period of time. She denied any abdominal trauma, injury, jaundice, nausea, diarrhea or vomiting. The patient was a smoker for 30 years.

The physical examination was unremarkable except for significant diffuse abdominal tenderness. Vital signs were normal. Laboratory findings revealed a mild leukocytosis of 16×10^3/l, C-Reactive Protein (CRP) elevation (285mg/l) and amylase blood levels were mildly elevated (145mg/l). Liver function tests were normal and both tumour markers, CEA and CA 19-9 were negative. Abdominal CT revealed a peripancreatic fluid collection, most probably as a result of leakage from the main pancreatic duct [Table/Fig-1].



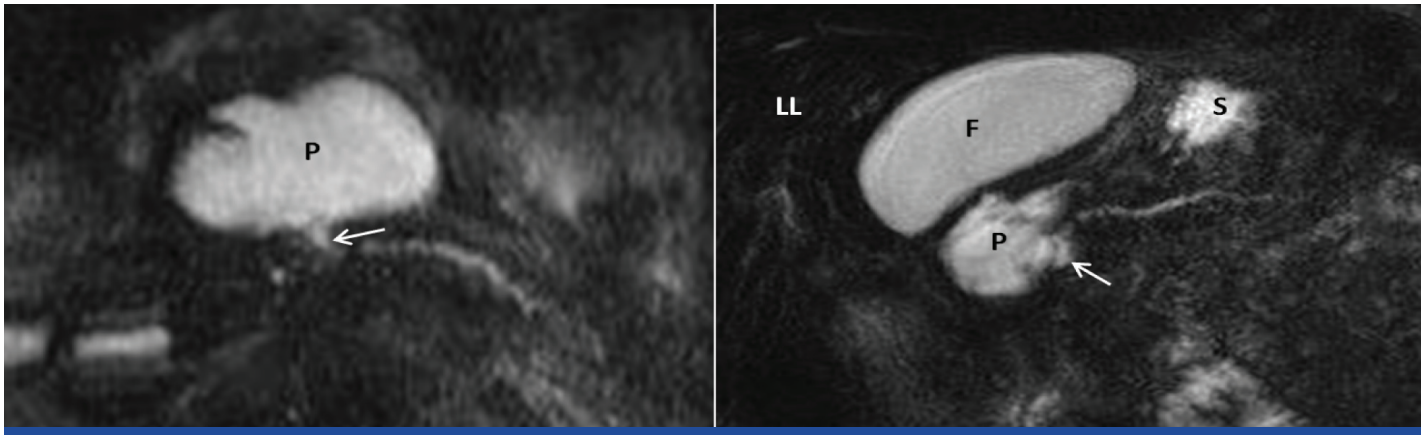
[Table/Fig-1]: Axial contrast-enhanced CT image obtained at the level of the pancreas shows pancreatic spontaneous neck transection (arrow) associated with peripancreatic fluid collection (P) formation.

Due to the unclear clinical and radiological picture, an upper abdomen Magnetic Resonance Imaging (MRI) [Table/Fig-2] and an Endoscopic Ultrasound (EUS) were performed, demonstrating only a slight expansion of the peripancreatic fluid, compared to the CT scan. The differential diagnosis at this stage included ruptured Intraductal Papillary Mucinous Neoplasm (IPMN), pancreatic serous cystadenoma and a pseudocyst. A week later the patient underwent an EUS-FNA of the peripancreatic fluid. The fluid analysis did not reveal any malignant cells and CEA and CA 19-9 markers were normal.

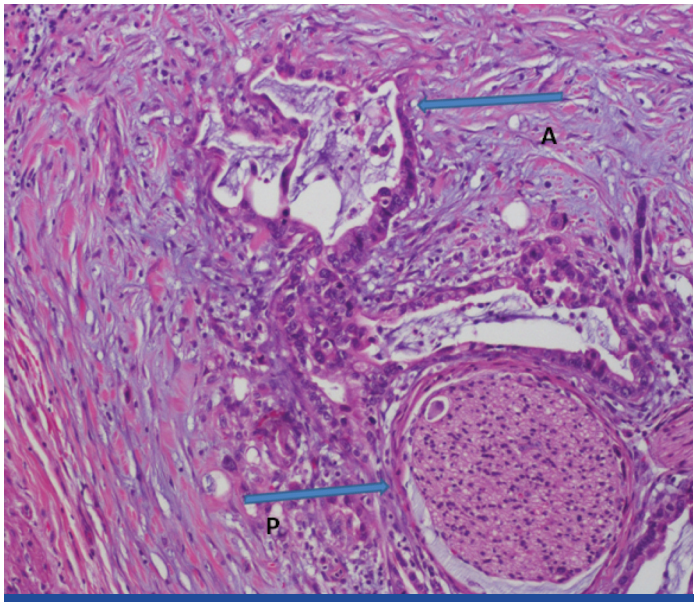
In the absence of improvement, the case was discussed on a multidisciplinary forum and a decision to perform surgery was made. During the surgery, two large peripancreatic fluid collections were found in the liver bed and in the lesser sac, without any palpable pancreatic mass. A distal pancreatectomy and splenectomy were performed and a frozen section showed signs of inflammation alone. The patient was eventually discharged on postoperative day 7.

Examination of the tissue pathology showed multiple microscopic foci of adenocarcinoma spread over a total area of (4X2cm). Most foci presented histologically with an evidence of massive perineural and vascular invasion and several of them reached the surgical margins at the pancreatic neck [Table/Fig-3]. Considering these pathologic results, it was decided to complete the resection and preform a total pancreatectomy.

On the second surgery, no free fluid was found in the abdominal cavity and no mass was palpated in the head of pancreas. Completion of the total pancreatectomy was performed, including a choledochojejunostomy and a gastrojejunostomy. A postoperative complication in the form of wound infection occurred and the patient was discharged on postoperative day 14. The specimen of the pancreatic head showed additional two foci of adenocarcinoma (0.1X0.3cm) and (0.6X0.4cm), with a massive perineural and vascular invasion. Ten lymph nodes were removed with no evidence of malignancy and the surgical margins (R) were negative.



[Table/Fig-2]: Magnetic resonance cholangiopancreatography (MRCP) images (A, B) show the focal disruption of the main pancreatic duct and the leakage site (arrow) with peripancreaticpseudocyst (P) formation. A large fluid collection (F) between the left lobe of the liver (LL) and stomach (S) was also detected.



[Table/Fig-3]: Photomicrograph showing a focus of adenocarcinoma (AC) with perineural invasion (PNI). (Haematoxylin and Eosin, x200)

DISCUSSION

In the United States pancreatic cancer is the fourth leading cause of cancer related death among both men and women. Eighty-five percent of pancreatic tumours are adenocarcinomas arising from the ductal epithelium. In contrast to other malignancies, the incidence and mortality rate for pancreatic cancer has increased during the past decade [1]. Since most symptoms associated with pancreatic cancer are relatively unspecific, the detection of the disease, despite advanced technologies, is still relatively late and at an advanced stage.

Surgical resection is the only potentially curative treatment for these patients, yet only 15 to 20 percent of patients are candidates for pancreatectomy at the time of diagnosis. The importance of early diagnosis also depends on the nodal status of the disease, being a major prognostic factor for the survival rate only 10 percent survive beyond two years with node-positive disease while the survival rate for node-negative disease is 25-30% [2-4].

Present case demonstrates a rare manifestation of pancreatic malignancy in which diffuse microscopic foci were found without any evidence of a mass. Moreover, the initial presentation in this case was the ruptured Wirsung duct, which to the best of our knowledge has not yet been described in the medical literature.

Differential diagnosis of main pancreatic duct rupture, which is usually associated with peripancreatic fluid connection, includes acute pancreatitis, chronic pancreatitis and trauma. When the anamnesis does not fit any of this diagnosis, alternative options may be ruptured cystic neoplasm, serous cystadenoma or IPMN.

The appropriate management in such conditions is always challenging. Abdominal CT scan has high accuracy for the presence of a pancreatic mass [5,6]. However, the diagnostic yield of a CT scan in cases of pancreatic cystic neoplasm remains under evaluation, Fisher WE et al., in his study on 48 patients with cystic lesions of the pancreas found that CT scan was accurate in only 61% of cases in determination of lesion nature [7]. MRI was not found to be superior compared to CT scan in pancreatic tumour diagnosis [8]. However, in cases of cystic lesions MRI and Magnetic Resonance Cholangiopancreatography (MRCP) can add additional anatomy information, mostly regarding communication with the main pancreatic duct and differentiation between pancreatic pseudocyst and cystic neoplasms [9,10]. EUS was found to be inaccurate in distinguishing between benign and potentially malignant pancreatic cystic lesions [11]. Nevertheless, it allows sampling of the pancreatic fluid. However, in case of negative cytology and normal marker levels, the diagnosis of pancreatic cancer cannot be excluded.

During the management of this case, we felt that the diagnosis was unclear. Expanding peripancreatic fluid and increasing patient discomfort encouraged us to use all possible diagnostic modalities. Unfortunately, they were not helpful in differentiating between pancreatic pseudocyst, rupture of any cystic mass or other rare diagnoses. After several multidisciplinary discussions, the indication for the primary surgery was the continuous leakage from the main pancreatic duct and the final pathology report was astounding. Our case demonstrates another option for consideration when an "unexplained" Wirsung rupture happens without presence of pancreatitis or trauma. We assume that a spontaneous rupture could be secondary to one of the cancers' foci growth and can be an early presenting sign of pancreatic cancer. The importance of this case is to increase the awareness of the rare possibility of pancreatic carcinoma which may be initially manifested as a rupture of the Wirsung duct without any evidence of a mass.

The natural history of such a case is unknown. There is no known explanation for the diffuse pattern of the tumour and the effect it has on local recurrence and survival. On one hand, this tumour should be classified as T1N0M0 (according to the American Joint Committee on Cancer, TNM system) so, the early diagnosis and treatment should account for better prognosis [12]. On the other hand, the aggressive features found on the pathology exam, the multi-focal spread, the early massive perineural and vascular invasion and the potential local spread due to the leakage to the abdominal cavity, all suggest worse outcome. Thus, we find it hard to assess the patient's prognosis.

CONCLUSION

Present case may raise awareness for the possibility of pancreatic cancer in any instance of unexplained rupture of the main pancreatic

duct. In addition, such cancer may be initially manifested as a multiple microscopic foci and not as a mass. Such awareness may help in early detection of pancreatic cancer.

REFERENCES

[1] Siegel RL, Miller KD, Jemal A. Cancer statistics, 2016. *CA: A Cancer Journal for Clinicians*. 2016;66(1):7-30.

[2] Benassai G, Mastrorilli M, Quarto G, Cappiello A, Giani U, Mosella G. Survival after pancreaticoduodenectomy for ductal adenocarcinoma of the head of the pancreas. *Chirurgia Italiana*. 2000;52(3):263-70.

[3] Bilimoria KY, Bentrem DJ, Ko CY, Stewart AK, Winchester DP, Talamonti MS. National failure to operate on early stage pancreatic cancer. *Annals of Surgery*. 2007;246(2):173-80.

[4] Cameron JL, Riall TS, Coleman J, Belcher KA. One thousand consecutive pancreaticoduodenectomies. *Annals of Surgery*. 2006;244(1):10-15.

[5] Miura F, Takada T, Amano H, Yoshida M, Furui S, Takeshita K. Diagnosis of pancreatic cancer. HPB : the official journal of the International Hepato Pancreato *Biliary Association*. 2006;8(5):337-42.

[6] Catalano C, Laghi A, Fraioli F, Pediconi F, Napoli A, Danti M, et al. Pancreatic carcinoma: the role of high-resolution multislice spiral CT in the diagnosis and assessment of resectability. *European Radiology*. 2003;13(1):149-56.

[7] Fisher WE, Hodges SE, Yagnik V, Moron FE, Wu MF, Hilsenbeck SG, et al. Accuracy of CT in predicting malignant potential of cystic pancreatic neoplasms. HPB: the official *Journal of the International Hepato Pancreato Biliary Association*. 2008;10(6):483-90.

[8] Lee HJ, Kim MJ, Choi JY, Hong HS, Kim KA. Relative accuracy of CT and MRI in the differentiation of benign from malignant pancreatic cystic lesions. *Clinical radiology*. 2011;66(4):315-21.

[9] Waters JA, Schmidt CM, Pinchot JW, White PB, Cummings OW, Pitt HA, et al. CT vs MRCP: optimal classification of IPMN type and extent. Journal of gastrointestinal surgery : official journal of the Society for Surgery of the Alimentary Tract. 2008;12(1):101-09.

[10] Macari M, Finn ME, Bennett GL, Cho KC, Newman E, Hajdu CH, et al. Differentiating pancreatic cystic neoplasms from pancreatic pseudocysts at MR imaging: value of perceived internal debris. *Radiology*. 2009;251(1):77-84.

[11] Gerke H, Jaffe TA, Mitchell RM, Byrne MF, Stiffler HL, Branch MS, et al. Endoscopic ultrasound and computer tomography are inaccurate methods of classifying cystic pancreatic lesions. Digestive and liver disease: official journal of the Italian Society of Gastroenterology and the Italian Association for the Study of the Liver. 2006;38(1):39-44.

[12] Edge SB, Compton CC. The American Joint Committee on Cancer: the 7th edition of the AJCC cancer staging manual and the future of TNM. *Annals of Surgical Oncology*. 2010;17(6):1471-74.

PARTICULARS OF CONTRIBUTORS:

1. Faculty, Surgery Division, Hillel Yaffe Medical Center, Hadera, Israel.
2. Faculty, Oncology Unit, Hillel Yaffe Medical Center, Hadera, Israel.
3. Faculty, Surgery Division, Hillel Yaffe Medical Center, Hadera, Israel.
4. Faculty, Gastroenterology Department, Hillel Yaffe Medical Center, Hadera, Israel.
5. Faculty, Radiology Department, Hillel Yaffe Medical Center, Hadera, Israel.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Shirly Shohat,
MD, Surgery Division, Hillel Yaffe Medical Center, Affiliated to Rappaport Medical School, Technion, Haifa Hadera-38100, Israel.
E-mail: shirlymitt@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **May 31, 2016**
Date of Peer Review: **Jul 07, 2016**
Date of Acceptance: **Sep 21, 2016**
Date of Publishing: **Dec 01, 2016**