

Disconnected Pancreatic Duct Syndrome in Necrotizing Pancreatitis Managed by Open Surgery: A Case Report

Sardar Mohammad Rezaul Islam¹, Debabroto Paul², Shah Alam Sarkar³, Mushfiqur Rahman⁴

¹Professor and Head, Department of Surgery, Ad-din Women's Medical College Hospital (AWMCH), Dhaka, Bangladesh.

²Associate Professor Department of Surgery, Jahurul Islam Medical college Hospital (JIMCH), Bajitpur, Bangladesh.

³Assistant Professor, Department of Surgery (JIMCH)

⁴Registrar, Sir Salimullah Medical College Hospital, Dhaka, Bangladesh.

Abstract

A 38 years old man was admitted with spontaneous rupture of walled of Necrosis of the pancreas. He had an attack of with acute pancreatitis 4 months ago for which he was hospitalized and treated conservatively. USG and MRCP shows a cavity with necrotic collection in the head area of the pancreas. On emergency laparotomy there was about 500 ml of free fluid in the peritoneal cavity. A cavity was noted in the head area which ruptured to the infracolic compartment through the transverse mesocolon. Pancreatic necrotic material and pus was drained from the cavity. After lavage an external tube drain was placed. Patient stayed in the hospital for 3 weeks on antibiotic, parenteral nutrition and Octerotide and blood transfusion. 150 ml of clear pancreatic secretion continued to drain via the abdominal drain. It was diagnosed as a case of external pancreatic fistula due to disconnected pancreatic duct syndrome (DPDS). After 2 months Roux-Y fistula-jejunostomy was done to close the pancreatic fistula and allow physiological drainage of the pancreatic juice to the jejunum. Patient recovered fully.

Keywords: Acute Pancreatitis, Disconnected pancreatic duct syndrome (DPDS), Walled off necrosis of the pancreas (WONP), Pancreatic fistula, Roux-Y Fistulo-jejunostomy.

INTRODUCTION

Acute pancreatitis is an acute inflammation of the pancreas resulting from an auto-digestion of the gland. In 75–80% of cases acute pancreatitis is a self-limiting disease which subsides spontaneously. But 20–25% of acute pancreatitis are severe, characterized by the development of pancreatic or peri-pancreatic necrosis, resulting in general and local complications responsible for a mortality rate of 8 to 35%^{1, 2}. The most common indication for intervention in acute pancreatitis are local complications such as walled off necrosis (WONP), pancreatic abscess, acute fluid collection, pseudocyst and abdominal compartment syndrome. The recently revised Atlanta classification subdivides acute fluid collections, which occur within four weeks of the onset of acute pancreatitis, into acute peripancreatic fluid collection (APFC) and acute necrotic fluid collection

(ANC). APFC develops to pseudocyst and ANC develops to pancreatic abscess or WONP.⁵ There is general agreement that surgery in severe pancreatitis should be performed as late as possible³. The rationale for late surgery is the ease of identifying well-demarcated necrotic tissue from the viable parenchyma. This approach limits the extent of surgery to the necrotic area only and not to the healthy parenchyma. This decreases the risk of bleeding and minimizes the surgery related loss of viable pancreatic tissue. Hence there is less possibility of surgery-induced endocrine and exocrine pancreatic insufficiency.

Corresponding Author: Sardar Mohammad Rezaul Islam, Professor and Head, Department of Surgery, Ad-din Women's Medical College Hospital (AWMCH), Dhaka, Bangladesh.

Mortality rates of up to 65 % have been described with early surgery in severe pancreatitis ^{2,4} That is why surgical intervention should be avoided within the first two to four weeks after onset of symptoms. Occasionally, pseudocysts or ANC may erode into adjacent hollow viscera of the gastrointestinal tract causing a fistulous connection and spontaneous internal drainage. The pancreatic duct also may be eroded in necrotic pancreatitis leading to pancreatic fistula. This is also called disconnected pancreatic duct syndrome (DPDS). The mechanisms associated with rupture, hemorrhage and erosion is digestion and disruption of the the adjacent wall of a hollow viscus, ducts and blood vessels by lytic enzyme of the pancreas. Compression of adjacent organs may result in local ischemia ^{7,9}. Clinical manifestations associated with DPDS include the development of recurrent peripancreatic fluid collections and persistent external pancreatic fistulae (EPF) after resolution of NP ¹¹⁻¹⁶. Simple internal and external fistulas tend to close spontaneously. Therefore, these fistulae are usually managed conservatively in the beginning using supportive treatment. If leaks do not resolve endoscopic stenting or percutaneous drainage is recommended in the majority of cases. Persistent fistulae require surgery as an alternative treatment option. Permanent internal drainage by Roux-Y cysto-jejunostomy (fistula-jejunostomy) is the ideal option of surgical treatment.

CASE REPORT

A 38 years old man was admitted with sudden upper abdominal pain, vomiting and fever. He was admitted 4 months ago with acute pancreatitis, treated conservatively. On admission, he was hemodynamically stable and temperature was 38.5. He had moderate leukocytosis,

not anemic. Renal and liver functions were within normal limit. Ultrasonography and MRI showed necrotic collection within head of the pancreas. (Fig-1 &2) MRCP shows a cystic structure in the head area of the pancreas with possible connection with the main pancreatic duct (Fig-3).

Emergency exploratory laparotomy was done with upper midline incision. There was about 500 ml of free fluid in the peritoneal cavity. An abscess cavity noted in the head area, which ruptured to the peritoneal cavity via transverse meso-colon to the infra-colic compartment. Pancreatic necrotic material and pus was drained. (Fig-4) The cavity washed with saline and a tube drain was left in the abscess cavity. Post-operatively about 150ml of dirty colored pancreatic secretion drained per day. At one point patient developed intra cystic hemorrhage. That was treated by fresh blood transfusion. The patient was put on Octerotide, prolong antibiotic treatment, fasting and parenteral nutrition. His drain collection gradually became clear pancreatic juice and remain persistent. (fig-5)The collection became around 150 ml per day. He was discharged 3 weeks postoperatively on oral feeding with the external drain tube in situ. It was clear that he developed external pancreatic fistula EPF due to DPDS and will need a second surgery. After 2 months he was re-operated and Roux-Y fistulo-jejunostomy (fig-7&8) was done joining with the fistula opening(Fig-6) on the right side of the transverse mesocolon on the infra-colic compartment. Post operative recovery was uneventful. He was discharged one week after surgery. He was seen after one year and was found doing well. Patient did not develop diabetes which suggests that his tail of pancreas remain functioning despite DPDS.



Figure 1. WONP on USG

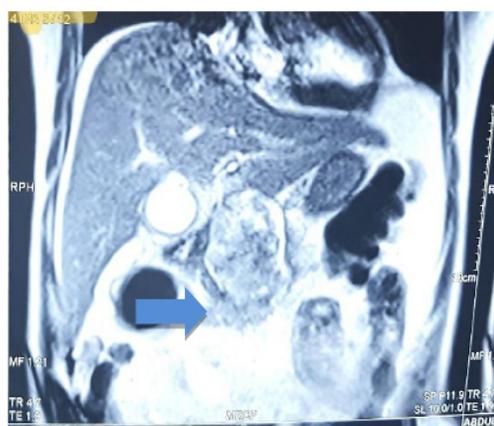


Figure 2. WONP on MRI



Figure 3. DPDS on MRCP

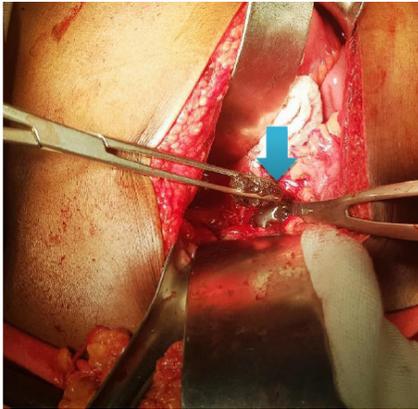


Figure 4. Drainage of pancreatic necrosis

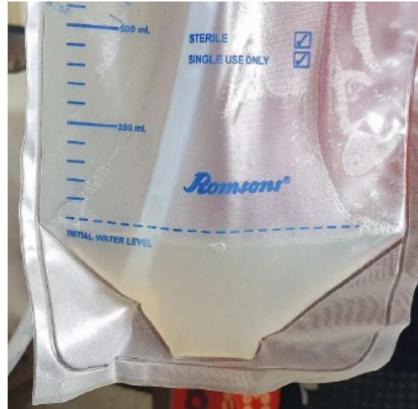


Figure 5. Pancreatic juice collection due to external fistula

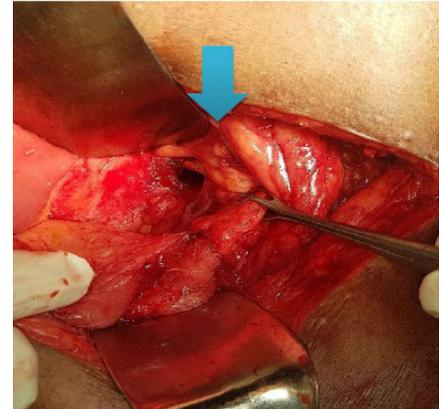


Figure 6. Fistula opening s



Figure 7. Fistulo-jejunostomy

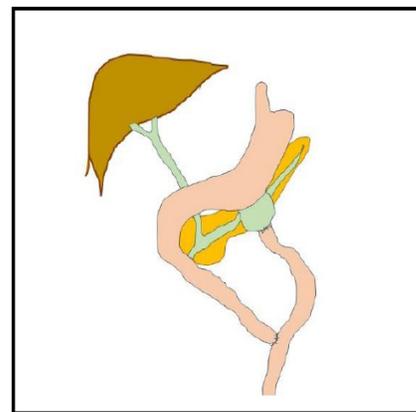


Figure 8. Roux-Y fistulo-jejunostomy(Graphic)

DISCUSSION

In acute necrotizing pancreatitis segmental necrosis of the main pancreatic duct may result in a discontinuity between the left-sided pancreas and the duodenum. Such an interruption in the setting of a viable upstream portion of the gland can give rise to the disconnected pancreatic duct syndrome (DPDS). Bang and colleagues showed that the development of DPDS was associated with the presence of walled-off necrotic collections (WON).¹⁰ By maintaining its secretory function, the disconnected segment may lead to persistent external pancreatic fistulae or recurrent pancreatic fluid collections. Despite the continual evolution of advanced endotherapy, surgery is still needed in a number of patients with DPDS. Surgery results in a high success rate and eventually provides a definite solution. Surgical treatment of persistent DPDS may involve resection of the upstream gland, with or without pancreatic islet cell auto-transplantation to reduce

the risk of diabetes mellitus (DM)¹⁷. If the upstream duct is of adequate size Roux-en-Y pancreato-jejunostomy or cysto-jejunostomy will preserve pancreatic function and will ensure physiological drainage of pancreatic secretions to the digestive tract^{18,19}. In a prospective study by Maatman and colleagues reported 68% of DPDS patients required operative intervention. Symptoms resolution occurred in 89% of patients after definitive surgery.²⁰ The most common surgery performed was internal drainage (46.8%) followed by resection procedures (38.4%), whereas remaining 14.8% patients underwent open debridement of necrosis. Nevertheless, post-operative morbidity is considerably high, and distal pancreatectomy has the highest risk of long-term endocrine and exocrine insufficiency²¹.

This is a case of very late complication of pancreatic necrosis. Here the walled off necrosis (WONP) of the pancreas had spontaneous intraperitoneal rupture causing

peritonitis. The main pancreatic duct was eroded by the long-standing necrotizing pancreatitis causing pancreatic fistula. Fistulo-jejunostomy was done after 8 weeks from the first surgery. This allowed the fistula tract to become very mature to hold anastomosis. It is a safe and effective treatment for persistent pancreatic fistula having the benefit of avoiding a difficult major pancreatic resection. The short- and mid-term outcomes of this procedure in literature are good.²²

CONCLUSION

DPDS is a clinical entity that usually occurs after severe acute pancreatitis and which presents with the appearance of a collection or EPF. Correct diagnosis is essential and it should be distinguished, from pancreatic pseudocyst, partial ductal disruption, WOPN and other post pancreatitis symptoms by CT scan and MRCP. Traditionally treatment was surgical but now it can be endoscopic, using either ERCP or, usually, internal endoscopic ultrasound guided drainage. Endoscopic techniques present low morbimortality but are less successful in the long term than surgical techniques. By-pass or resection surgery is more effective but has greater morbimortality. There are no internationally- agreed therapeutic algorithms but it is increasingly more common to use endoscopic techniques first and if they fail, consider surgery

Fiistulo-jejunostomy is ideal procedure to achieve permanent internal drainage. This not only treats fistula it also helps pancreatic juice take part in digestion when anastomosis is done with jejunum.

REFERENCE

1. Lankisch PG, Brener N, Burns A et al. Natural history of acute pancreatitis: a long-term population-based study. *Am J Gastroenterol* 2009; 104:2797-2805.
2. Yadav D, O'Connell M, Papachriston GJ. Natural history following the first attack of acute pancreatitis. *Am J Gastroenterol* 2012; 107:1096-1103.
3. Uhl W, Warshaw AL, Imrie C, Bassi C, McKay C, Lankisch P, Carter R, DiMaggio E, Banks P, Whitcomb D, Dervenis C, Ulrich C, Satake K, Ghaneh P, Hartwig W, Werner J, Entee G, Neoptolemos J, Büchler MW: IAP Guidelines for the surgical management of acute pancreatitis. *Pancreatology* 2002;175
4. Mier J, Leon E, Castillo A, Robledo F, Blanco R: Early versus late necrosectomy in severe necrotizing pancreatitis. *Am J Surg* 1997; 173:71-75
5. Sarr MG, Banks PA, Bollen TL, et al. Revision of the Atlanta classification of acute pancreatitis. *Acute Pancreatitis Classification Workgroup*. 2008 Apr; Available from: <http://pancreasclub.com/atlanta-classification>. [Google Scholar]
6. Erik G. Pearson, Courtney L. Scaife, Sean J. Mulvihill, Robert E. Glasgow. Roux-en-Y drainage of a pancreatic fistula for disconnected pancreatic duct syndrome after acute necrotizing pancreatitis: *HPB*, Vol-14, Issue-1 2012, <https://doi.org/10.1111/j.1477-2574.2011.00397>.
7. Somani PO, Jain SS, Shah DK, et al. Uncomplicated spontaneous rupture of pancreatic pseudocyst into stomach: A case report. *World J Gastrointest Endosc.* 2013;5(9):461-464. [PMC free article] [PubMed] [Google Scholar]
8. Marta Sandini, Thilo Hackert, Markus W. Büchler .Management of Pancreatic Fistula in Acute Pancreatitis. *The pancreas*: August 2023 <https://doi.org/10.1002/9781119876007.ch36>
9. Okamura K, Ohara M, Kaneko T, et al. Pancreatic pseudocyst ruptured due to acute intracystic hemorrhage. *Case Rep Gastroenterol.* 2017;11:755-762. [PMC free article] [PubMed] [Google Scholar]
10. Bang and colleagues showed that the development of DPDS was associated with the presence of walled-off necrotic collections (WON)
11. Lawrence C, Howell DA, Stefan AM, et al. Disconnected pancreatic tail syndrome: Potential for endoscopic therapy and results of long-term follow-up. *Gastrointest Endosc* 2008;67:673-9.
12. Fischer TD, Gutman DS, Hughes SJ, et al. Disconnected pancreatic duct syndrome: Disease classification and management strategies. *J Am Coll Surg* 2014;219: 704-12.
13. Uomo G, Molino D, Visconti M, et al. The incidence of main pancreatic duct disruption in severe biliary pancreatitis. *Am J Surg* 1998;176:49-52.
14. Fotoohi M, D'Agostino HB, Wollman B, et al. Persistent pancreatocutaneous fistula after percutaneous drainage of pancreatic fluid collections: Role of cause and severity of pancreatitis. *Radiology* 1999;213:573-8.

15. 15. Baron TH, DiMaio CJ, Wang AY, et al. American gastroenterological association clinical practice update: Management of pancreatic necrosis. *Gastroenterology* 2020;158:67-75 e1.
16. 16. Rana SS, Sharma R, Kang M, et al. Natural course of low output external pancreatic fistula in patients with disconnected pancreatic duct syndrome following acute necrotising pancreatitis. *Pancreatology* 2020;20:177-81.
17. 17. Bellin M.D., Freeman M.L., Gelrud A., et al. Total pancreatectomy and islet autotransplantation in chronic pancreatitis: recommendations from PancreasFest. *Pancreatology*. 2014;14(1):27-35. doi: 10.1016/j.pan.2013.10.009. [PMC free article] [PubMed] [CrossRef] [Google Scholar]
18. 18. Sornsins SM, Marx JA. Rupture of a pancreatic pseudocyst following blunt abdominal trauma. *J Emerg Med*. 1988;6(1):29-32. [PubMed] [Google Scholar]
19. 19. Whitman GJ, Pearce WH, Moore EE, Barton B. Free intraperitoneal rupture of a pancreatic pseudocyst: A presentation of acute abdominal trauma. *J Trauma*. 1988;28(4): 555-56. [PubMed] [Google Scholar]
20. 20. Maatman T.K., Roch A.M., Lewellen K.A., et al. Disconnected pancreatic duct syndrome: spectrum of operative management. *J Surg Res*. 2020;247:297-303. doi: 10.1016/j.jss.2019.09.068. [PubMed] [CrossRef] [Google Scholar]
21. 21. van Dijk S.M., Timmerhuis H.C., Verdonk R.C., et al. Treatment of disrupted and disconnected pancreatic duct in necrotizing pancreatitis: a systematic review and meta-analysis. *Pancreatology*. 2019;19(7):905-915. doi: 10.1016/j.pan.2019.08.006.
22. 22. Gunjan Desai, Rajvilas Narkhede, Prasad Pande, Pares Varty, Hitesh Mehta, Dattaprasanna Kulkarni Roux-en-Y fistula-jejunostomy in the management of persistent external pancreatic fistula: is it olde worlde? *Turk J Surg*. 2018 Aug 28;35(1):62-69. doi: 5578/turkjsurg.4110.

Cite this article: Sardar Mohammad Rezaul Islam, Debabroto Paul, et al. *Disconnected Pancreatic Duct Syndrome in Necrotizing Pancreatitis Managed by Open Surgery: A Case Report. International Journal of Research in Medical and Clinical Sciences. 2024;2(1): 47-51.*

Copyright: © 2024. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.