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Disconnected Pancreatic Duct Syndrome: A Neglected Entity

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Author's contribution

The sole author designed, analyzed and interpreted and prepared the manuscript.

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Mini-review Article

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ABSTRACT

Pancreatic fistula is one of the complications of acute necrotizing pancreatitis (ANP), chronic pancreatitis, trauma, surgery, and malignancy. Disconnected pancreatic duct syndrome (DPDS) is one of the potential complications, however, not as well known. The approach to DPDS is clinically different than the approach to a simple pancreatic fistula, with unique diagnostic and therapeutic challenges to minimize morbidity and facilitate a smooth recovery. The aim of this mini-review is to provide clinicians with the most recent updates regarding early detection and to discuss the changing models for its management and methods for treating appropriate pathways.

Keywords: Acute necrotizing pancreatitis; fistula; disconnected pancreatic duct syndrome; endoscopic retrograde cholangiopancreatography; surgery.

1. INTRODUCTION

Disconnected pancreatic duct system (DPDS) is defined as complete necrosis and disruption of the main pancreatic duct, leaving a viable secreting pancreatic segment distal to it, [1,2] and a resulting loss of communication with the gastrointestinal tract.

There is a Scarcity of information in the literature regarding prevalence of DPDS [1].

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Acute necrotizing pancreatitis (ANP) was found to be the major cause of DPDS in 60% of the cases in one study[3], In another study, ANP was identified as a cause in 34%-50% of the cases[1], where chronic pancreatitis was indicated in 31%-35% [3] of the cases and the remaining ones were classified as idiopathic or attributed to trauma.

The mean age of the patients was 53, with 66% of them being male. DPDS has three forms of presentation: The concurrent form, will presents as the same time as the ANP and necrosectomy (56%) and delayed presentation occurs both after ANP with pseudocyst (30%); and as a consequence of chronic pancreatitis (14%) [4].

There are no clear presenting signs and symptoms for DPDS, but there is usually persistence of fluid collection or non-resolving pancreatic fistula, vague abdominal pain, nausea and vomiting, food intolerance and weight loss [5].

2. DIAGNOSTIC CHALLENGES

Most studies show that the majority of cases are diagnosed retrospectively, with delays that characteristically reach up to nine months [3]. This indicates, either a difficulty in diagnosing the condition early for clinical and radiological reasons, or a lack of awareness about this condition on the part of the clinician/radiologist. A missed or delayed diagnosis can often have serious negative impacts on the biopsychosocial welfare of the patients. These effects range from multiple unnecessary radiological investigations to a poor quality of life and serious local and systemic complications, such as persistent or uncontrolled pancreatic fistula or unresolving pseudocyst, with futile prolonged catheter drainage due to reduced chances of achieving spontaneous closure [3]. Therefore, the importance of early detection cannot be overestimated.

The majority of the available data, which was collected from retrospective studies by reviewing the computed tomography (CT) findings of patients with confirmed DPDS, seems to agree on a certain number of criteria for making the diagnosis[2,3]:

1. It has to involve minimum two methods.
2. When a CT scan shows distinct intra-pancreatic fluid collection along the expected course of the main pancreatic duct, and viable pancreatic tissue distal to that collection, the size of the collection is not the key and the location in the neck is considered to be the most common area because of the lack of blood supply [3].
3. If endoscopic retrograde cholangiopan-creatography (ERCP) shows evidence of duct occlusion at the level of the intra-pancreatic fluid collection, with a contrast leak, treatment should be done within a minimum of two weeks after the acute attack [3].
4. One study emphasizes another important radiological sign. Not all fluid collection is an indication of DPDS, and the collection may be compressing the gland to the extent that it appears to be necrotized, while in fact the presence of a thin bridge of viable glandular tissue inferiorly or posteriorly excludes it [2].
5. Secretin-enhanced magnetic resonance cholangiopancreatography (MRCP) was reviewed in one article in terms of its diagnostic efficacy. The article concluded that this is a non-invasive, safe and effective modality even in visualizing leaks, especially three weeks after an acute attack [6].
6. Endoscopic ultrasound (EUS) has correlated features for diagnosing DPDS, and upstream pancreatic parenchyma and ducts were found to terminate within the walled-off necrosis (WON) [7].

3. TREATMENT

The options for treating DPDS based on the latest data available are as follows:

3.1 Embolization with N-butyl-2-cyano-acrylate

An innovative endoscopic technique to seal up pancreatic fistulas by using N-butyl-2-cyanoacrylate is described through a report on a study of 12 patients where the treatment was successful and helped to avoid surgery in eight cases [8].

A report of another four-case study concluded that endoscopic sealing of pancreatic fistulas is safe and effective. [9]

Finally, a case report using the same endoscopic method of sealing for a patient who had been diagnosed with DPDS showed that the patient

remained symptom-free after being followed for one year [10].

3.2 ERCP and Stent

Since the main problem in DPDS is a disrupted duct and the persistence of pancreatic collection, ERCP and stent placement has been advocated and is described in the following reports.

1- In one study, 97 patients were reviewed after stent placement for DPDS (using a 3F), and this was successful in 55% of the cases, with a 36% failure rate [11].
2- Another study of 12 patients with DPDS who were treated endoscopically with two double-pigtail plastic stents (using a 7F and 4 cm) found that long-term follow up with a median time of 28 months showed no recurrence of symptoms [1].
3- In a retrospective review of 33 patients who had recurrent pancreatic fluid collections (PFCs) due to DPDS and were managed by the placement of permanent transmural stents (7F or 10F, 4-cm, double-pigtail polyethylene), eight patients had surgery, one patient passed away due to multi-organ failure and two had lost follow-up. Among the remaining 22, none had PFC recurrence during a median follow-up of 1,026 days, although three patients had stent migration. Unfortunately, the study was not able to identify any distinct risk factors that can anticipate stent migration [12].
4- One randomized trial of 28 patients who were followed over 27 months, tried to answer the question of whether to keep or remove the stents, with 15 patients keeping the stents (group A), and 13 having the stents removed (Group B). The study showed that five patients in group B had recurrence after stent removal, whereas none of the patients in group A had a recurrence, and the study concluded that stent retrieval was associated with a higher recurrence rate [13].

These data, then, collectively show good results for using the stents as treatment but also indicate that migration is a concern and removal of the stents is not recommended because it is associated with higher recurrence rates. This method provides a good option for the older, more fragile patients who are not likely to withstand the major operative resection and for whom there are concerns about exocrine and endocrine functional losses.

3.3 EUS and Drainage

EUS followed by trans-papillary drainage of the retained pancreatic duct represents a reasonable option when the papilla of Vater cannot be accessed or when a catheter cannot be introduced through the papilla because of post-inflammatory or post-operative reactions.

One case report includes EUS-guided trans-gastric drainage of the enlarged pancreatic duct through the insertion of a 10-Fr Amsterdam prosthesis. At a follow-up time of 15 months, no signs of stent blockade or migration were found [14].

Another study involved five patients who had recurrent pancreatic fluid collections of the pancreatic tail that were managed by endoscopic ultrasound-guided drainage into the fourth portion of the duodenum by placement of 7-Fr and 10-Fr double-pigtail plastic stents into the collections. Four patients met the criteria for DPDS, and they were followed for up to 34 months after removal of the stent and had no recurrences. The study concluded that this method of treating pancreatic collection is feasible and appears safe and effective [15].

3.4 Surgery

Two options are available:

1. **Roux-en-Y drainage:** In one study, seven patients were treated with internal drainage procedures, and the median follow-up was 264 days. Only one patient needed distal pancreatectomy, and no patient developed exocrine insufficiency [16].

In a similar study that involved 27 patients, 13 of whom were treated with internal drainage and 14 with distal pancreatectomy, those who had internal drainage needed less operative time, experienced less blood loss, and had fewer transfusion requirements [17].

2. **Distal pancreatectomy:** This procedure is considered to be an option with good results for treating fistula. However, the procedure requires an extensive dissection in an inflamed area, and removal of a major viable segment of pancreas and spleen is not

without potential major morbidity risks of its own [18].

4. CONCLUSION

DPDS is recognized as the sequelae for a number of pancreatic conditions such as sever acute pancreatitis (SAP), pancreatic necrosis and chronic pancreatitis. Delayed diagnosis, resulting mainly from lack of awareness among clinicians and radiologists about the condition and its diagnostic criteria, has led to a higher morbidity rate. In the management of DPDS, both embolization and ERCP along with stenting are appropriate options for older, more fragile patients, while surgical options are better for patients who are young and fit. Internal drainage is considered to be a better surgical option than pancreatic resection.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable

COMPETING INTERESTS

Author has declared that no competing interests exist.

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