Groove Pancreatitis: What is its Relevance to Surgeons?

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Obstruction

Groove pancreatitis is a segmental chronic pancreatitis, that affects the anatomical area between the pancreatic head, the duodenum, the common bile duct, referred to as groove area [1,2]. This distinct form of chronic pancreatitis, occurring predominately in and around the duodenal wall has been reported under various names, including cystic dystrophy of heterotrophic pancreas [3,4] pancreatic hamartoma of duodenum [5,6], paraduodenal wall cyst [7,8], myoadenomatosis [9,10], groove pancreatitis (GP) [11,12] and paraduodenal pancreatitis [13,14]. In 1982 Stolte et al. [15] coined the term “groove pancreatitis” for this special form of segmental pancreatitis, characterized by fibrous scars of the anatomic space of groove area and Becker and Mischke classified groove pancreatitis into the pure form and segmental form [11]. The prevalence of GP is difficult to assess, but in the four surgical series, the reported incidence varied from 2.7%, 19.5%, 24.4% to 70% of duodenopancreatectomy specimens obtained from patients in chronic pancreatitis [11,14-16]. GP is often diagnosed in 40 to 50 year old alcoholic [11,15]. The patients usually present with postprandialabdominal pain and subsequently, impaired motility, stenosis of the duodenum and postprandial vomiting often leading to significant weight loss [1-3]. This rare but distinct entity is often confused clinically with ampullary neoplasms, duodenal tumors, cystic tumours of head of pancreas and acute relapsing pancreatitis [1-3]. Jaundice, which is less commonly seen, is found to occur when the inflammatory process involves the terminal common bile duct. Weight loss is occasionally seen and this may make the suspicion of malignancy stronger. A minority of the cases may have acute presentation of gastric outflow obstruction [20]. The duration of the clinical symptoms ranges from a few weeks to more than one year, with most of them presenting within 3 to 6 months of the symptoms [1,2,14,20].

Clinical Symptoms

The predominant symptoms in these patients are, severe abdominal pain and recurrent vomiting [1,2,11,13-15]. Involvement of duodenum will lead to signs related to it, including impaired motility and disordered gastric vomiting, mainly due to duodenal stenosis [1,2]. Jaundice, which is less commonly seen, is found to occur when the inflammatory process involves the terminal common bile duct. Weight loss is occasionally seen and this may make the suspicion of malignancy stronger. A minority of the cases may have acute presentation of gastric outflow obstruction [20]. The duration of the clinical symptoms ranges from a few weeks to more than one year, with most of them presenting within 3 to 6 months of the symptoms [1,2,14,20].

Pathology

The duodenal wall close to the minor papillae often show significant thickening and scarring causing stenosis of the second part of the duodenum [1,2,11,13,15]. Cystic changes in the thickened duodenal wall are characteristic. These can vary in size from 0.2 to 2 cms and are usually found in the submucosa and muscle layer and contain clear fluid and at times granular white material and stones [2,11,13]. These have been reported in 49% of patients in some series [11].

The cyst are lined with columnar epithelium or replaced by granulation tissue [11,21]. The duodenal mucosa often reveals nodular lesions, ulceration and scarring [21]. Microscopy reveals Brunner gland hyperplasia and thickening of the submucosal and muscle layers caused by extensive fibrosis [1,2,11,17,21].

The parenchyma of the pancreatic head shows significant fibrosis and scarring which initially involves the groove area and then the head of pancreas [13,15,17]. Involvement of head may lead to dilatation of pancreatic ducts. Santorini’s duct may become dilated with protein plugs and calcification [11,13,16].

Brunner gland hyperplasia is considered to arise from an increase in cholecystokinin-pancreazymin or gastrin levels [11,14]. This may lead to stasis of pancreatic juice in dorsal pancreas and rupture of the duct. The extravasated mucoprotein material is associated with foreign body giant cell reaction and may lead to pancreatitis in the groove pancreas [14]. In some cases, incomplete involution of the dorsal pancreas at this region is seen as evidenced by the presence of pancreatic tissue at the duodenal wall and is considered as predisposing factor for GP in them [1,2,14,15].

In this editorial, the pathogenesis, diagnostic challenges, imaging modalities, pathological features, management, clinical outcome and the relevance to surgeons of this rare condition is discussed.

Pathogenesis

Majority of these patients are young men and have a history of alcohol abuse [1-3,17]. The pathogenesis of GP remains unclear; however there seems to be a relationship with chronic pancreatitis as indicated by lack of differences in age/ gender distribution or in the prevalence of alcohol consumption between these two conditions. Several factors in the pathogenesis of GP have been reported, including peptic ulcer, gastric hypersecretion, gastric resection, true duodenal wall cyst, pancreatic heterotopia in the duodenum or disturbance of flow of pancreatic juice through the Santorini duct [2,11,15]. The characteristic location of the lesions around the minor papilla suggests the presence of an anatomical or functional predisposing condition at this region [11,15]. In a significant proportion of the patients, absent or narrow Santorini duct or pancreas divisum has been observed [18,19]. This is likely to lead to resistance to outflow of pancreatic juice, along the accessory duct and may render pancreatic tissue vulnerable to alcoholic injury. The viscosity of the pancreatic juice, which changes following excessive alcohol consumption and/ or smoking, may lead to pancreatic duct calcification [1,3,11,14-19]. Moreover
Groove pancreatitis is classified into pure form, which is restricted to the groove area without the involvement of the main pancreatic duct and a segmental form, which encompasses the entire head of the pancreas, with a stenotic and dilated main pancreatic duct [11,13,16,17]. Fibrosis around the inferior bile duct causes its narrowing and jaundice [1,2,17].

**Diagnosis**

Slight elevation of serum pancreatic enzymes (amylase, lipase and elastase1) is often seen in blood tests with occasional rise in serum hepatic enzymes [1,2,16,20]. Isolated elevation in alanine-L-transpeptidase or in alkaline phosphatase has been noted by some [2,20]. Bilirubin elevation has been seen in patients who are clinically jaundiced. Tumour markers, carcinoembryonic antigen and carbohydrate antigen 19-9, when determined, are usually within normal limits [16,20]. Patients with chronic symptoms may develop exocrine pancreatic insufficiency or diabetes [2,14,16,17,20].

Upper gastrointestinal endoscopy shows erosion erythema, stenosis due to edema, fibrosis and polypoid appearance in the descending part of the duodenum [20,22]. Transduodenal biopsy is important to distinguish periampullary cancer from groove pancreatitis [20,22]. However due to sampling errors, the diagnostic value of the biopsy may be limited in number of cases [23]. It is important to be aware that fibrotic changes disclosed by cytology or histopathology do not preclude a neoplasm, since desmoplastic reaction is often found in association with adenocarcinoma, thus mimicking a chronic inflammatory abnormality such as GP [23].

Ultrasonography shows the entire thickness of the duodenal wall and stenosis of the lumen, with a hypoechoic lesion between the pancreas head and the thickened duodenum [1,16,17,20]. Endoscopic ultrasonography has been reported to be accurate in evaluation of the location and demonstration of the extent of GP [24]. It reveals a smooth stenosis of the common bile duct. Santorini’s duct is usually undetectable [24].

Computed tomography (CT) is an excellent means of displaying the spectrum of imaging findings in GP. In the pure form of GP, a poorly enhancing plate like hypodense lesion can be identified on early phase of dynamic CT, between the pancreatic head and the duodenum, near the minor papilla (Figure 1) [25]. The imaging features reflect the histopathological characteristics of the mass. The delayed enhancement observed in a number of cases is due to delayed blood circulation caused by fibrous tissue proliferation, leading to constriction of arteries [17,20]. Also noted are findings of duodenal stenosis accompanied by wall thickening and cyst like lesions in the duodenal wall or in the groove area. These can vary in size from tiny to large. Multilocular cysts are often seen in number of these cases. Multiple cysts located within a thickened duodenal wall exhibiting post contrast enhancement are sometimes associated with duodenal thickening or stenosis [16,17,20]. Patients with chronic symptoms may develop exocrine pancreatic insufficiency or diabetes [2,14,16,17,20].

Differential Diagnosis

Several differential diagnosis are entertained due to the pathology involving the duodenum, CBD and head of pancreas. The differential diagnosis of pure form of GP includes duodenal cancer, cholangiocarcinoma or acute pancreatitis with phlegmon along the groove [1,2,16,17,20-22]. MRCP is useful in differentiating GP from CBD cancer since the former shows a long segmental smooth CBD stenosis or tapering, unlike CBD cancer where it is short and irregular [2,17,20]. The signal characteristics of the sheet like mass or MR images in GP are useful in differentiation from acute pancreatitis, as phlegmon in contrast, exhibits high signal intensity on T2 –weighted images [16,16,20,26]. Chronic pancreatitis presenting with acute episode and pseudocysts in the duodenal wall constitutes an important differential diagnosis. It however differs from GP because of the absence of duodenal wall thickening or stenosis. Moreover in GP, there are true cysts and not pseudocysts in the duodenal wall or in the groove area [1,2].

The significant relevance to the surgeon is in differentiating it from pancreatic adenocarcinoma as it is challenging [17,21,22]. The findings in the imaging that differ from those of pancreatic adenocarcinoma include: GP appears as a sheet like mass unlike a round irregular mass in pancreatic carcinoma [24-26]. In a dynamic CT scan in a patient with GP, the mass in pancreatic head (except for cystic lesions) may enhance, while it will not in malignancy. Stenosis of bile duct is smooth and long in GP, but abrupt, short and irregular in pancreatic cancer [25]. Cystic lesions are more often present in duodenal wall in GP than in pancreatic cancer. Importantly the vessels in cancer of pancreatic head are often encased or infiltrating but are rarely involved in GP.

Because of close resemblance to pancreatic cancer in some patients, surgeons may have to closely follow up of patients with GP particularly if the initial transduodenal biopsy has been inconclusive [14,16,20,21].
Treatment of GP

Conservative management has been recommended in acute phase of GP including bed rest, fasting, analgesia and parental nutrition [1,2,7,27]. The effectiveness of this treatment should be evaluated after 4 to 6 weeks, in terms of clinical symptoms, laboratory data and imaging findings [27]. Conservative management would invariably lead to relief of symptoms in most patients; however the concern is that of relapsing episodes of pancreatitis as a consequence anatomic disturbances of pancreatic outflow and alcohol consumption [27]. In such patients, GP is resistant to medical treatment and may follow a prolonged course. It is prudent that these patients who are resistant to medical treatment and are at a late phase of presentation or present with complications such as duodenal obstruction, would warrant surgical intervention [14,16]. Surgery also plays a role in relieving pancreatic insufficiency (weight loss, steatorrhea or diabetes) and achieves symptomatic relief and adequate weight gain [1,2,16,17,20].

The surgical treatment of choice is a pancreaticoduodenectomy using a Whipple procedure or a pylorus preserving pancreaticoduodenectomy [16,17,20,28]. Apart from surgical procedures that involve removal head, some organ preserving techniques such as pancreaticojugal anastomosis have been advocated in small series; but are generally discouraged as the main site of pathology is retained and outcome is not satisfactory [14]. An alternative treatment option to surgery is the endoscopic drainage of the accessory pancreatic duct via minor papilla [29]. Successful treatment by endoscopic drainage of minor duct has been achieved, where a disturbance in pancreatic juice secretion via minor papilla plays a crucial role. However its long term results are obscure [29]. The long-term outcome is favorable following pancreaticoduodenectomy with complete relief of pain in 75% of the patients [14]. Failure to adopting a change in life style including cessation of alcohol consumption and smoking, leads to a risk of recurrence in 24% of patients [14].

In conclusion this uncommon and interesting condition presents certain diagnostic and management challenges to surgeons and hence deserves attention. It should be included in the differential diagnosis of pancreatic masses with signs of duodenal/periduodenal infiltration. Of outmost importance, is to differentiate it from pancreatic cancer with appropriate use of radiological and endoscopic biopsy options. The role of surgeon is obvious, when complications like duodenal or CBD obstruction sets in. When the focal pancreatic lesion depicted on imaging studies is suggestive of GP, it should influence the surgeon to perform further investigations before radical surgery like pancreaticoduodenectomy with complete relief of pain in 75% of the patients [14]. Failure to adopting a change in life style including cessation of alcohol consumption and smoking, leads to a risk of recurrence in 24% of patients [14].

References