Pancreatic Pseudocyst in Federal Medical Centre, Gombe and Review of Literature

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Abstract

Background*: Pseudocyst arises from pancreatic inflammation or ductal disruption and is the commonest cystic lesion of the pancreas. Although spontaneous resolution is common particularly in its early phase, progression of the cyst may culminate in grave consequences for the patient.*

The aim of the study is to highlight the presentation, management and outcome of pseudocyst in an environment where it is reportedly uncommon.

Methodology*: Case series with review of literature.*

Result*: Although pancreatic pseudocyst is not very frequently seen in this environment, it is not entirely rare but is frequently misdiagnosed. Non specific symptoms in the early stage and subsequent confusion with various intra abdominal pathologies are among the reasons for late diagnosis and delayed intervention.*

Conclusion*: Pseudocyst is amenable to surgical treatment. Early recognition is imperative in preventing complications and in defining the true incidence of this disease entity in our locality. Clinical suspicion in every setting of pancreatitis complemented by serum amylase estimations and early ultrasound scan will improve the diagnosis.*

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Introduction

Pseudocyst is the commonest cystic lesion of the pancreas and mostly complicates acute or chronic pancreatitis following alcoholism, cholelithiasis and trauma. It differs from a true pancreatic cyst by lack of an epithelial lining and constitutes about two-thirds of all pancreatic cystic lesions[1]. It is reportedly an uncommon clinical entity in our environment especially in areas where alcoholic beverages are forbidden by religion and culture[2] but the exact incidence is unknown[3]. Pancreatic pseudocyst can occur in various locations in the body and mimic pathology in diverse organs but it is typically found in the lesser sac[4]. Although spontaneous resolution following a non-operative treatment is common particularly in the early stage, progression of the disease may be of grave consequence to the patient. This makes prompt diagnosis with a rapid and efficient intervention very vital to limiting complications from pancreatic pseudocysts.

Case 1.

A.M was a 27years old, multiparous housewife from Jalingo who was referred to us with 3months history of recurrent exacerbation of epigastric pain which radiated to the back and was at presentation associated with progressive abdominal distension, easy satiety, occasional vomiting and weight loss. She had no jaundice. She was previously being managed for unconfirmed peptic ulcer disease dating back to 7 months at a pheripheral hospital and was referred to our centre on account of suspected ovarian mass when her abdominal distension became gross. Examination revealed a young anicteric woman with PR of 72b/m and BP of 130/80mmHg with a non tender, non pulsatile epigastric mass extending to the pelvic region.

USS showed a 14.4 x 13.4cm low level echo cyst with multiple gallbladder tiny echogenic foci casting acoustic shadow with normal wall thickness. Barium meal showed a soft tissue, fairly rounded mass in the lesser sac region with normal duodenal cap and C loop. LFT was normal and RBS was 4.2mmol/L.

A working diagnosis of pancreatic pseudocyst to rule out mesenteric cyst was made and she had laparatomy with findings at operation including: A thick walled, unilocular lesser sac cyst containing debris with adhesion to posterior gastric wall, inferior displacement of the transverse colon and a gallbladder which contained multiple tiny stones. She had cystogastrostomy, excision of cyst wall for biopsy and cholecystectomy. Pseudocyst was confirmed at biopsy. Her post operative period was uneventful and she was discharged on post operative day ten.

Case 2

S.A was a 26year old male, petrol hawker, who sustained blunt injury to the abdomen when he was knocked down by a vehicle 8 weeks earlier. He was

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initially managed in a peripheral hospital for this injury and was subsequently discharged. However, he noticed persistent abdominal pain which became worse 2 weeks after his initial hospital discharge and reported back to the peripheral hospital where he was managed with analgesics and antacids for peptic ulcer disease. This was nonetheless followed by progressive abdominal distension with transient, spontaneously regressed jaundice of 4 days duration. He was referred to our centre on account of suspected chronic liver disease when he developed vomiting, difficulty in lying down/breathing, paedal edema and skin rashes 2 months later. There was no past history of abdominal pain or alcohol ingestion.

USS confirmed the presence of a huge, thick walled, cystic epigastric mass while a barium a meal and follow through excluded intragastric or duodenal lesions. His LFT was normal with Alkaline Phosphatase of 153IU/L and Serum amylase of 41IU/L. An epigastric cystic mass with a downward bulge through the transverse mesocolon was confirmed at laparatomy and he had a Roux en Y Cystojejunostomy done. The postoperative period was uneventful and the histopathology report excluded neoplasm.

CASE 3

HAA was a 27years old male Fulani driver who loss consciousness and sustained blunt abdominal injury in a road traffic accident when his vehicle loss control and hit a tree. He was transferred to us 2 days later after he had regained back his consciousness at the referral hospital where he was initially managed. When seen at the A&E he had abdominal pain and distension. He was in pains, pale, dehydrated but anicteric. His PR and BP were 118b/m and 130/80mmHg respectively. Examination revealed distended abdomen with generalized abdominal tenderness maximal in the epigastrium with resonant percussion notes and hypoactive bowel sounds. An assessment of peritonitis from blunt abdominal injury to rule out perforated viscus was made and patient had emergency exploratory laparatomy. The intraoperative finding were injury to the left lobe of the liver, pancreatic injury with zone 2 retroperitoneal haematoma extending to around the greater curvature of the stomach, the transverse mesocolon and widespread areas of fat necrosis. The liver laceration was repaired, peritoneum was lavage with normal saline and the abdomen closed enmass.

However, the patient re-presented 2 months later, with a huge progressive epigastiric mass associated with discomfort and early satiety.USS revealed an epigastric cystic mass measuring 83cm x123cm x 128cm and a volume of 684cm^3. The RBS was 8.0mmol/L. He had cystogastrostomy following a diagnosis of pancreatic pseudocyst which was confirmed at laparatomy. He developed superficial wound infection and was discharged home 12days post operation on daily wound dressing.

Discussion

Pancreatic pseudocyst is a collection of pancreatic secretions usually rich in digestive enzymes, necrotic tissue, old blood which is walled off by fibrous or granulation tissue[4]. Pseudocysts vary in their sizes and may be single or multiple. They could be located far from the pancreas (extra-pancreatic) but they are typically located in the lesser sac (peri-pancreatic) encased by adjacent viscera such as the stomach, colon, transverse mesocolon, gastrocolic omentum and pancreas[5]. This usually makes mobility exceptional for pseudocysts and was a cause of clinical confusion where it was reported[5]. In this series, all our cases were large unilocular cysts located in the lesser sac.

Pseudocysts are commonly found in the adults due to the incidence of gallstone diseases and alcoholism but could be found in children where it usually follows blunt abdominal injuries[6]. The cases in our report were however all in their third decades of life which may be due to the small number reported. The age range also differs from that of five patients reported by Agbakuru et al in Ile-ife over a 10years period whose age ranged from 23 to 70year and this may be due to the inclusion of patients in whom the aetiology of their cysts was due to alcoholism in their study compared to ours[7].

Most pseudocysts are asymptomatic but can manifest with a wide range of clinical problems depending upon their location, the extent of the fluid collected or the presence of complications such as infection. Nevertheless, the earlier symptoms are usually non-specific, consisting of epigastric pain, nausea, vomiting, fever or weight loss which may make diagnosis difficult particularly where the disease is not been suspected[3]. A pseudocyst typically forms from seepage of fluid into the lesser sac through the posterior layer of the parietal peritoneum which subsequently results into a well circumscribed, sessile epigastric mass immobilized by adhesion to the adjacent structures. Abdominal mass is its most common mode of presentation and this may be associated with pain and tenderness due to inflammation or expansion of the cyst. Diagnosis may however be confounded by confusing this resultant mass with several pathologies which includes aortic aneurysm, panreatic neoplasms, omental/mesenteric

cyst and lesser sac tumors[8,9] in cases of peri-pancreatic pseudocysts. The situation is more challenging in the tropics with inaccessible or few diagnostic facilities in the presence of several disease entities that may mimic the presentation of pseudocysts [10,11]. Two of the patients in this series were erroneously treated for unconfirmed peptic ulcer disease during the early phase of their disease and were also mis-diagnosed and referred as cases of ovarian mass and chronic liver disease without suspecting pancreatic pseudocysts. Early diagnosis of pancreatic pseudocyst will always require a high index of clinical suspicion particularly in every settings of pancreatitis which may be further corroborated by an elevated amylase level. Although an elevated serum amylase is not exclusive to pseudocyst, a persistently elevated level typically above 1000 IU/L will always suggest the possibility of development of pseudocyst as a complication of pancreatitis. Such elevated enzyme level may also arouse early suspicion to search for extra-pancreatic pseudocysts which are formed far away from the pancreas and diagnosis could then be confirmed by demonstrating elevated enzyme level in the aspirates from the extra-pancreatic collections[12,13,14]. Unfortunately, amylase estimation is not quite a regular and routine investigation in most facilities in our locality and only one of our patients had amylase estimation done.

The incidence of abdominal pain in pseudocyst reportedly ranges from 76-100%[7,15] but featured in all the patients in this series. Complications from pancreatic pseudocyst can occur due to compression, invasion or rupture of pseudocyst into surrounding structures besides infection and hemorrhage[16]. The compressive effect on the gastrium, duodenum or the biliary tract may lead to early satiety, gastric outlet obstruction with vomiting or obstructive jaundice[15]. Vascular compression of the inferior venal cava, portal or iliac vessels in gross cases may cause paedal swellings, portal hypertension or organomegally and the aorta may be involved in rare cases[17]. Pancreatic ascites can result from rupture or disruption of the pancreatic duct and there may be pleural or pericardial effusion as well as spontaneous infection of the cyst. Upper gastrointestinal bleeding may arise from vascular erosion into the splenic, gastroduodenal and pancreaticoduodenal arteries or rupturing of a pseudoaneurysm. Bleeding into the pancreatic duct (hemosuccus pancreaticus) may account for the remaining cases of gastrointestinal haemorrhage. Pseudocyst fistulas may occur and the colon, stomach, duodenum and less commonly, the esophagus are the more frequently involved organs with rare biliary tract involvement[18].

Extra-pancreatic pseudocysts collection at various sites is facilitated by proteolytic enzymatic digestion and tracking of fluid along established tissue planes. Pseudocysts could be found in the mediastinum or the neck when the fluid dissects between the crura of the diaphragm[13]. Perirenal cysts may occur when fluid tracks down from the posterior part or tail of the pancreas through the anterior pararenal space en route the lumbar gutters and may finally end up in the inguinal region or the scrotum[14,19]. Intra parenchymal pseudocysts accumulation within the solid organs may occur along their vascular or ligamentous attachment as is the case in splenic and liver parenchyma pseudocysts where the fluid gain access through the splenic hilar vessels and the hepatogastric ligament respectively. Intra pancreatic pseudocysts are more related to chronic pancreatitis and has been reported in heterotopic pancreas[20,21].

Ultrasonography is a cheap, widely available, readily conductible investigative modality with a reasonable degree of sensitivity and remains the initial radiological investigation of choice. It is useful in defining the size of the cyst, its wall thickness, its relationship to the digestive system, the presence of biliary calculi and presence of dilatation of the biliary tract or septae within the cyst. It is a valuable tool for postoperative patient follow up as well as early localisation of pseudocysts. Endoscopic ultrasonography has the ability to demonstrate the presence of vessels or aneurysms in a pseudocyst and is of great value in avoiding hazardous bleeding during endoscopic cystenterostomy. An upper gastrointestinal endoscopy has the ability to differentiate an intragastric pathology from pseudocyst and will also demonstrate extragastric compression from a large pseudocyst. A barium meal serves as an alternative investigation in demonstrating intragastric pathology where endoscopy is not available and can define the relationship of the cyst to the upper digestive tract. Chest X ray will confirm the presence of an accompanying left or bilateral pleural effusion. CT is invaluable in situations where there is limitation with ultrasonography such as in mediastinal pseudocysts and for characterising concurrent or underlying pancreatic lesions. MRI has the ability to demonstrate very well the presence or extension of fistulous tract into the pancreas and is effective in detecting the presence of a solid component to the cyst as well as differentiating between organized necrosis and a pseudocyst[22]. Endoscopic retrograde pancreatography may detect cysts not seen by ultrasound; demonstrate the presence of communications between a pseudocyst and the pancreatic duct which is of important

consideration in the choice of treatment as the risk of pancreaticocutaneous fistulae is lower in endoscopic drainage than percutaneous methods. It is equally efficient in defining pancreatic ductal pathology (such as stricture and obstruction) which may likely prevent spontaneous resolution[21]. MRCP has similar sensitivity to ERCP but has the advantage of providing images of the pancreatic ducts in their natural state by not involving the distension of the ducts by the injection of contrast media[23].When pancreatography is necessary, it is better within a day or two of operation under antibiotic cover to lessen the chance of infection. Angiography is not a routine investigation of pseudocyst but in the setting of pseudoaneurysms resulting from vascular erosion, it could serve both as a diagnostic and therapeutic intervention.

Liver function test, Percutaneous transhepatic cholangiography and other ancillary investigations may complement ultrasonography where there is obstructive jaundice. Microbiology will isolate infective organisms while Cytology or biopsy of the cyst wall will identify neoplastic cysts. Carcinoembryonic antigen (CEA) and carcinoembryonic antigen-125 (CEA-125) tumor marker levels are low in pseudocysts and elevated in tumors. A carcinoembryonic antigen (CEA) level greater than 400 ng/mL within the cyst fluid strongly suggests malignancy. An elevated serum glucose level may herald the development of diabetes mellitus post pancreatic inflammation.

The initially approach to treatment of pseudocysts is usually conservative as cysts may spontaneously resolve when small (less than 6cm in size), are in the acute phase (less than six weeks), have a wall thickness not more than 1cm and when there are no other pancreatic duct abnormality other than the pancreatic duct-pseudocyst communication[24] . Up to 50% of pseudocysts may regress on expectant management[25]. The surgical approach to pancreatic pseudocyst entails drainage of the cyst using various techniques with considerations given to the anatomy of the cyst, the pathology present and the general condition of the patient[26]. Although medical management has been tried and anecdotally reported to be successful, the therapy is usually prolonged[27]

Internal drainage is the traditionally preferred method of treatment and involves an anastomosis between the cyst and the upper digestive tract (stomach, duodenum or jejunum). It has been accomplished by open, endoscopic or laparoscopic techniques[28,29]. Success of internal drainage depends on the presence of a mature cyst wall to hold the stitches in the open technique and this may be infeasible in acute pseudocyst which occurs within 3-4weeks of an attack of acute pancreatitis with a wall of early, immature granulation tissue as oppose to chronic pseudocysts which has persisted for more than 6 weeks and has walls of normal granulation or fibrous tissue [1].Some studies have however demonstrated the feasibility of anastomosis earlier than 6 weeks[30]. The appearance of an isoenzyme of amylase called “old amylase” in serum has been found predictive of cyst wall maturation and was introduced as a biochemical marker and timer for performing a safe internal drainage[21].

The factors guiding the specific drainage site includes: size and location of the cyst, condition of its wall, relation of cyst to the gastrointestine, rapidity of its growth and imminence of rupture , presence of complications (such as rupture, massive suppuration or active haemorrhage of the cyst) and the patients' age/ general condition[5]. Retrogastric anteriorly enlarging cysts are best drained by a posterior cystogastrostomy of Jurasz and those on the pancreatic head abutting on the duodenum by cystoduodenostomy of Kerschner[29]. Large cysts, with inferior enlargement, bulging through the transverse mesocolon especially when they are neither adherent to stomach nor duodenum are best drained by Roux-en-Y cystojejunostomy[29]. The prerequisite for performing a cystoenterostomies include the absence of intra cyst papillary growth, pseudoaneurysm or compartments of the cyst and absent epithelial lining of the cyst wall where a frozen section is carried out[26]. Cystogastrostomy and Roux-en-Y cystojenostomy were employed in treating our patients with satisfactory outcomes.

Endoscopic drainage is a more recent form of internal drainage with all the full compliments of a minimally invasive procedure. It entails an image guided widening to about 2cm of a diathermic puncture made between the digestive tract mucosal and the cyst followed by insertion of nasocystic catheter for a few days. Endoscopic drainage may be achieved by transpapillary stenting when the pseudocyst communicates with the pancreatic duct or by transmural gastric/duodenal wall drainage via the site of maximal extrinsic compression and visible endoscopic 'bulge'[31,32]. Endoscopic drainage has also been extended to extra pancreatic pseudocysts as in mediastinal and intra-hepatic pseudocysts[13,32,33]. Transmural endoscopic cystoduodenostomy and cystojejunostomy are more technically infeasible when the distance between the viscera and the cyst is more

than 1 cm and are hazardous in the presence of pseudoaneurysm or intervening cyst wall vessels which serves as contraindications to the procedures[34,35]. The recurrence rate for all endoscopic procedures for the treatment of pancreatic pseudocysts is less than 5% and the complication rate is approximately10%[36]. The possible complications from this procedure include serious bleeding from unsuspected pseudoaneurysm or intervening vessels in the cyst wall, pseudocyst infection and bowel perforation. The drawbacks to transpapillary stenting particularly include exacerbation of pancreatitis, stent occlusion leading to recurrent duct blow-out and pseudocyst formation or chronic pancreatitis[37].

External drainage provides a rescue treatment where there is a rapidly growing cyst with immature wall and imminent rupture especially in poor risk/ critically ill patients[6]. It involves the drainage of the cyst outside the abdominal cavity either by open or Image guided percutaneous catheter. Sequela of this form of treatment includes infection, fistula formation, skin enzymatic digestion, bowel catheter erosion in prolonged drainage and frequent cyst recurrence[38]. It should consequently not be undertaken in the presence of pancreatic necrosis or a solid non-drainable pancreatic mass, lack of safe access route, active pseudocyst hemorrhage or compartments within the cyst. Serial ultrasound monitoring should be incorporated as part of treatment to ascertain the efficacy of the drainage. Sandostatin, a "somatostatin analogue" which is noted for decreasing the basal and stimulated pancreatic secretion has been used as an adjuvant to catheter drainage to improve its efficacy[39]. Marsupialization is now rarely undertaken as a form of drainage except where the content is massively infected[26].

Complications reported with percutaneous drainage include infection, pneumothorax, pleural effusion, and minor haemorrhage[24]. Image guidance percutaneous catheter drainage is associated with success rate of 90 to 95% but simple aspiration of pseudocyst is associated with recurrence rate of 70% and should be discouraged[24,25,40].

D'Egidio et al has earlier emphasised the need to classify pancreatic pseudocyst to enable the adoption of the most appropriate form of treatment and evolved a clinico-radiographic pseudocyst classification[38].This comprises of patients with acute, 'post-necrotic' pseudocysts having a normal pancreatic duct anatomy and rarely duct-pseudocyst communication in Group I category. Here, Percutaneous drainage is curative in all patients when used. Group II patients include those already suffering from chronic pancreatitis that develops 'post-necrotic' pseudocysts. Here, duct-pseudocyst communication is often present and the pancreatic duct is diseased but not strictured. Surgical internal drainage is usually a successful treatment although percutaneous drainage may be possible, but may have to be prolonged. Group III patients are those with chronic 'retention' pseudocysts who has grossly diseased and strictured pancreatic duct in the presence of duct-pseudocyst communication. In this category, cure demands operative procedures that address the specific underlying ductal pathology and surgical internal drainage has a high recurrence rate while percutaneous drainage is contraindicated.

Haemorrhage is the single most important complication of pseudocyst contributing up to 50% mortality in some series[30]. Managing a pseudocyst with active haemorrhage could be a formidable threat but could be achieved by vascular embolisation prior to surgery, balloon catheterization, suture ligation of the vessel or by pancreatic resection together with the pseudocyst where the haemorrhage is uncontrollable. Equally amenable to pancreatic resections are pseudocysts located at the pancreatic tail[6]. Pancreaticoduodenectomy may suffice where a co-existing pancreatic tumor is discovered or in multiloculated cyst unyielding to other form of drainage. However, this could be a daunting operation with a high morbidity and mortality and is best left to a skilled surgeon in pancreatic surgery. Pancreatic pseudocysts complicated by biliary problems such as gallstones or unresolved ductal compression after drainage of the cyst are treated on their own merits besides the drainage of the pseudocyst. We as a result carried out cholecystectomy in one of our patients with associated gallbladder stone. The mortality of pancreatic pseudocyst is increased in the presence of complications which include: Pancreatic necrosis or abscess, haemorrhage, rupture into cavities and its sequela, intestinal obstruction and fistula formation. Therefore, prompt drainage of the cyst by the most appropriate mean once spontaneous resolution is unlikely as seen in the cases presented in this study remains vital to ensuring a good outcome by eschewing grievous consequences.

In conclusion, although pancreatic pseudocyst is uncommonly seen in our locality, it is not an entirely rare pathology but is frequently misdiagnosed with delayed intervention. The symptoms are non specific for early lesions causing confusion with variable pathologies such as peptic ulcer disease. Furthermore, corroborative diagnostic facilities such as amylase

estimation may not be readily accessed. The diagnosis of pancreatic pseudocysts needs to always be suspected, then investigated and appropriately followed up. The practice of treating peptic ulcer disease without endoscopic confirmation in our environment should be reviewed. Epigastric pain and swelling being among the more definite features of peri-pancreatic pseudocyst should always point to the possibility of pseudocyst when they occur and be followed by an ultrasound scan for a prompt diagnosis.

While late presentations are fraught with grievous consequences, early diagnosis apart from preventing complications will be useful in ascertaining the true incidence of this disease entity in our environment by detecting those cysts that may spontaneously regress and not likely be reckoned with when estimating the incidence of the disease. In our setting, internal drainage provides satisfactory outcome of treatment but is mainly in form of open anastomosis between cyst and the upper digestive tract as there is limited experience with endoscopic drainage in this part of the world. Endoscopy in form of ERCP and MRCP are invaluable for unravelling the underlying ductal pathology and adequate categorisation of pseudocysts. Acquisition of relevant minimally invasive/endoscopic technology and expertise will therefore allow us adopt this approach where it is of superior advantage.

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