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ORIGINAL ARTICLE

The leaking pancreatic duct in childhood chronic pancreatitis

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Abstract The leaking pancreatic duct in childhood chronic pancreatitis presents with ascites and pleural effusion and is a potentially lethal condition. Seven children with this condition were seen in the period 2003–2006. The correct diagnosis was not entertained till a raised serum amylase was discovered. The diagnosis was confirmed by very high levels of amylase in the aspirated abdominal or pleural fluid. Computerized tomogram was the most useful imaging study and demonstrated a dilated pancreatic duct. All children were operated within 6 days of diagnosis by a Puestow's procedure in six and peripancreatic drainage in one. Six children made a prompt and lasting recovery after a Puestow's procedure while one child, also suffering from metastatic neuroblastoma, died in the immediate post operative period after peripancreatic drainage. We recommend prompt and definitive surgical management of this potentially lethal condition.

Keywords Pancreatic ascites · Pancreatic pleural effusion

Introduction

Pancreatitis is termed 'chronic' when irreversible changes have taken place [1–4]. The radiological hallmarks are calcification and/or ductal dilatation. The serum amylase is not usually elevated and surgery is generally an elective drainage procedure or a resection for chronic intractable pain [2, 5–7]. We present seven children with previously unsuspected chronic pancreatitis who presented with internal pancreatic fistulae, emphasizing the difficulties in diagnosis and the need for prompt surgical treatment.

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Materials and methods

Patient details are outlined in Table 1

Ascites and/or left pleural effusion of unknown origin was the chief presentation. In addition, the children were malnourished, in pain and case no. 7 was in multiorgan failure, needing ventilation and hemodialysis. No previous local or systemic illness or history of trauma was present in any child except in case no. 7 who had received treatment for metastatic neuroblastoma including autologous bone marrow transplantation. The admission diagnoses were invariably wrong and included tuberculous abdomen, tuberculous pleural effusion, empyema thoracis and Budd Chiari syndrome. Hypoalbuminemia was often present and hyperamylasemia indicated the correct diagnosis. Ascitic and pleural fluid amylase levels were astronomical (median = 24,480 IU).

Once pancreatic pathology was suspected, CT scan was preferred as the chief imaging modality (Fig. 1). This showed a prominently visible and often irregular main pancreatic duct with diameter ranging from 1.3 to 5.1 mm. Calcification in the pancreas was seen in two out of the seven patients. In case no. 4 the dilatation was confined to the duct of Santorini in the pancreatic head. CT scan or chest X-ray also showed ascites and/or pleural effusion. ERCP, done only in case no. 1,

Table 1 Clinical and biochemical features of cases 1–7

Case no.	Age (years)	Sex	Presenting symptoms	Examination	Serum amylase	Serum albumin	Ascitic fluid amylase	Follow up serum amylase
1	5	Male	Abdominal pain, abdominal distension	Ascites, abdominal tenderness	547	2.7	4,030	–
2	4	Female	Abdominal distension, Dyspnoea	Ascites, abdominal tenderness	1,820	1.9	7,370	186
3	14	Male	Abdominal pain, vomiting, abdominal distension	Ascites, left sided pleural effusion	4,010	1.5	1,56,200	340
4	2	Male	Abdominal distension, dyspnoea	Left pleural effusion	1,014	3.4	28,950	258
5	15	Male	Abdominal pain, dyspnoea	Ascites, left pleural effusion	2,622	3.6	57,250	535
6	5	Male	Abdominal pain, abdominal distension, vomiting	Ascites	2,910	2.0	24,480	134
7	8	Male	Abdominal distension, dyspnoea, metatstatic neuroblastoma	Tense ascites	933	1.4	7,350	Expired

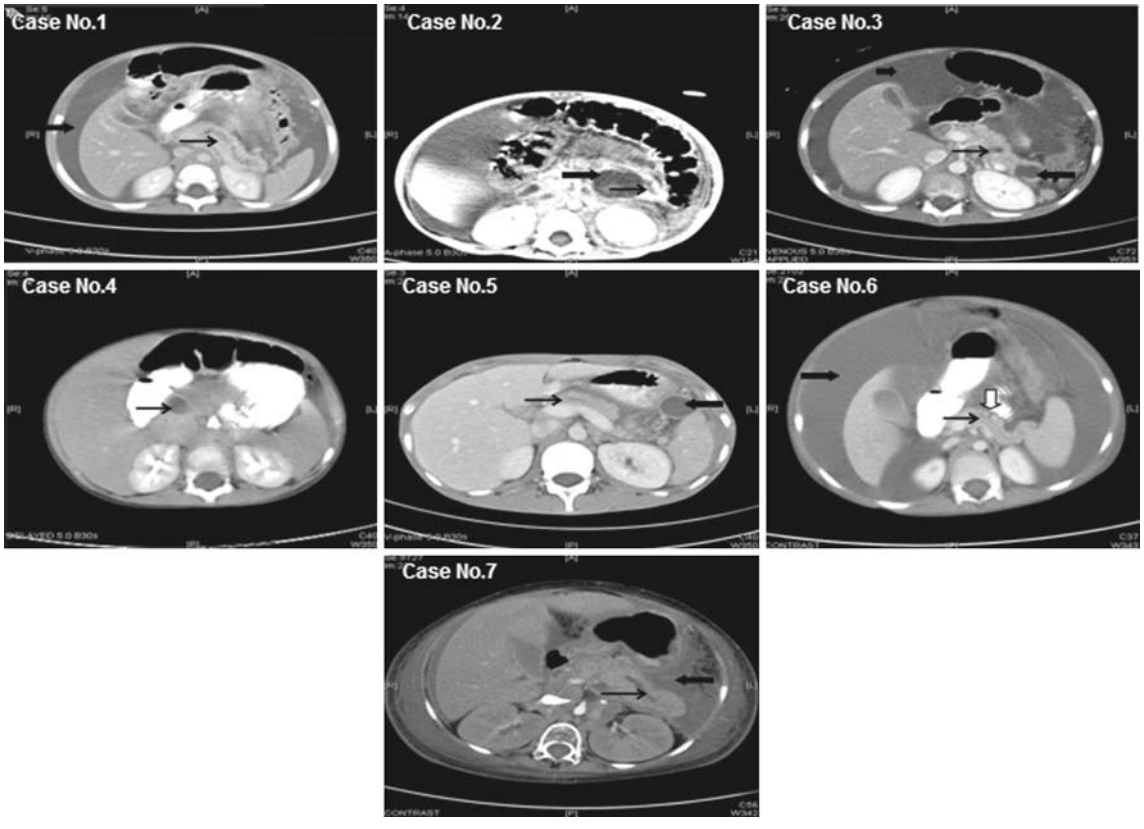

Fig. 1 CT Scan images of cases 1–7 showing dilated pancreatic duct (*black arrows*), leaking side duct in case no. 6 (*white arrow*), ascites and collections (*thick black arrows*)

demonstrated the site of the pancreatic fistula, but this was not considered an essential investigation as the entire dilated pancreatic duct required surgical drainage in the form of a lateral pancreaticojejunostomy, irrespective of the site of the leak.

Surgery was performed within 1–6 days (median = 2 days) of diagnosis of a leaking dilated pancreatic duct and after initial resuscitation including thoracic paracentesis. At laparotomy an anterior duct disruption was found in cases 1–3 and case 6 and a leak

on the postereosuperior surface in cases 4 and 5. The dilated pancreatic duct could be located by palpation or partial division of the pancreatic tissue and aided by an operative pancreaticogram in case 4. In cases 1–6, the dilated duct was laid open along its length and a lateral pancreaticojejunostomy (Puestow's procedure) performed using a roux loop of the jejunum. The duct was always more dilated at surgery than suggested by the CT scan. Peripancreatic drains and a feeding jejunostomy were placed in all these cases. Case no. 7, who was too unstable to undergo a definitive procedure, underwent insertion of peripancreatic drains only.

Results

Postoperatively cases 1–6 made a dramatic recovery and have remained well. Follow up serum amylase (available in 5) ranged from 134 to 535 IU. Pleural effusion or ascites have not recurred. Case no. 7 for whom no definitive procedure was possible because of multiorgan failure and severely deranged coagulation parameters, expired on the third postoperative day. Although this child had metastatic neuroblastoma, this was not immediately life threatening and his death was ascribed to his pancreatic complications.

Discussion

Acute pancreatitis occurs due to pancreatic auto digestion but is a reversible process. In chronic pancreatitis the disease process is irreversible. Inflammatory changes in the pancreas may be associated with varying degrees of pancreatic duct dilatation and progressive or permanent loss of pancreatic function. Pancreatitis is rare in children and chronic pancreatitis is rarer still [1, 4, 8]. Internal pancreatic fistulae following chronic pancreatitis is therefore not a commonly reported condition in children. The child presents with pancreatic ascites (pancreatico-peritoneal fistula) and/or pancreatic pleural effusion (pancreatico-pleural fistula) [9, 10]. Internal pancreatic fistulae are caused by disruption of the pancreatic duct. In acute pancreatitis the inflammatory response walls off the disruption leading to a pseudocyst. The same however does not occur in chronic pancreatitis and ascites or pleural effusion results [11]. Anterior duct disruption leads to pancreatic ascites while posterior disruption causes tracking up of pancreatic secretions in the retroperitoneum into the mediastinum and the resultant pleural effusion [12]. In chronic pancreatitis the serum amylase is generally not raised but a leaking pancreatic duct causes the amylase to be passively absorbed into the blood stream through the pleural and peritoneal surfaces causing a rise in the serum amylase [11]. The preoperative serum amylase in our patients ranged from 547 to 4010 units, though a normal serum amylase does not rule out internal pancreatic fistula [12]. Children with internal pancreatic fistulae are generally anorexic and malnourished with progressive abdominal distension and/or dyspnoea [12]. The serum albumin was less than 3 gm/dl in five of our seven cases.

A high degree of suspicion is necessary to make a diagnosis of a leaking pancreatic duct. The correct diagnosis was initially missed in all our seven cases till serum amylase estimation was done. The diagnosis was confirmed by ascitic and pleural fluid amylase, which was markedly elevated [11, 12]. Chronic pancreatitis was indicated on a CT scan by the presence of a dilated and irregular pancreatic duct and this prompted us to elect for early surgery. We feel that a CT scan to demonstrate a dilated pancreatic duct is more useful and less invasive than ERCP. Though an ERCP would demonstrate the exact site of leakage (case no. 1) this, in our opinion, is relatively unimportant, as the entire dilated duct needs operative drainage in the form of a lateral pancreaticojejunostomy.

Normally the pancreatic duct is seen as a single echogenic line on ultrasonography. The diameter of the normal main pancreatic duct is less than 1 mm [13]. The diameter of the main pancreatic ducts on CT scan in the children in our study ranged from 1.3 to 5.1 mm. This was associated with ductal irregularity (Fig. 1). The marginal dilatation seen in some children was possibly due to leakage of pancreatic secretions from the obstructed duct system. During surgery the ducts were found to be adequately dilated to perform a technically easy Puestow's operation.

Non-operative management of internal pancreatic fistulae has been advocated but the success rate of this line of therapy is reported to be less than 50% [11, 12]. In the setting of chronic pancreatitis, surgery offers the best chance for a definitive cure of the fistula and should not be unduly delayed. A feeding jejunostomy is an invaluable addition to the procedure.

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