Idiopathic giant pancreatic pseudocyst presenting in emergency with abdominal compartment syndrome and intestinal occlusion: Case report and review of the literature

Giuseppe Evola *, Francesco Ferrara, Mario Scravaglieri, Elio Trusso Zirna, Salvatore Sarà, Luigi Piazza

General and Emergency Surgery Department, Garibaldi Hospital, Piazza Santa Maria di Gesù 5, 95100, Catania, Italy

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A B S T R A C T

INTRODUCTION AND IMPORTANCE: This is the first case of idiopathic giant pancreatic pseudocyst (IGPP) causing intestinal occlusion, intra-abdominal hypertension (IAH) and abdominal compartment syndrome (ACS) reported in the literature. Diagnosis of IGPP in emergency is a challenge because of its rarity and the absence of a history of pancreatitis or pancreatic trauma and specific clinical presentation. Abdominal contrast-enhanced computed tomography (CECT) represents the gold standard in diagnosing pancreatic cyst (PP). Different types of treatment of PP are reported in the literature.

CASE PRESENTATION: A 52-year-old Caucasian female was admitted to the Emergency Department with a three-day history of abdominal pain, inability to pass gas or stool, nausea and vomiting, oliguria and a seven-day history of abdominal swelling and swollen legs. Physical examination revealed abdominal distention, abdominal pain, swelling in the legs. CECT showed a voluminous cystic pancreatic mass suspected of neoplasm. Laboratory tests reported high serum levels of BUN, creatinine and C-reactive protein and neutrophilic leukocytosis. After preoperative diagnosis of ACS, the patient was taken to the operating room for pancreatic resection. The postoperative course was uneventful. Diagnosis of IGPP was made by histopathological examination.

CLINICAL DISCUSSION: IGPP is difficult to diagnose in emergency. Although different types of drainage of IGPP are described in the literature, pancreatic resection represents the treatment of choice when a cystic pancreatic neoplasm cannot be excluded.

CONCLUSION: IGPP is a rare disease that may cause intestinal occlusion, IAH and ACS. Pancreatic resection if necessary is safe and therapeutic with acceptable morbidity and mortality.

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1. Introduction

Pancreatic pseudocyst (PP) is an encapsulated fluid collection surrounded by a non-epithelialized wall, localized in the pancreatic tissue or the adjacent pancreatic space. It mainly represents a complication of pancreatitis or pancreatic trauma [1] but rarely is idiopathic. The size of PP varies from small (<2 cm) to medium (2–6 cm) and large (>6 cm); a PP with the major diameter ≥10 cm is termed giant PP [1]. Few cases of complicated giant pancreatic pseudocysts (PPs) have already been reported in the literature [2] but this is the first case of idiopathic giant pancreatic pseudocyst (IGPP) causing intra-abdominal hypertension (IAH) and abdominal compartment syndrome (ACS). Diagnosis of IGPP in emergency, even with the help of radiological imaging, is a challenge because of its rarity [3], the absence of a history of pancreatitis or pancreatic trauma and specific symptoms and signs. Appropriate treatment of established PP generally requires percutaneous, endoscopic or surgical drainage but in our case report uncertain nature of giant pancreatic cystic mass and the presence of IAH and ACS required in emergency a decompressive laparotomy and a distal pancreaticosplenectomy. A case of IGPP causing IAH, ACS and intestinal occlusion is presented with review of the literature in accordance with SCARE 2020 criteria [4]. The purpose of this case report is to remember that IGPP is a uncommon disease that may require emergency surgery.

2. Presentation of case

A 52-year-old Caucasian female was admitted to the Emergency Department with a three-day history of abdominal pain, inability to pass gas or stool, nausea and vomiting, oliguria and a seven-day history of abdominal swelling and swollen legs. She was pale,
hypotensive, tachycardic and tachypnoic. Vital signs were blood pressure 90/45 mm Hg, pulse 110 bpm, respiratory rate 24 per minute, oxygen saturation 93% in ambient air and temperature of 37.2 °C. The patient wasn’t taking any drug, referred habit on smoking but denied alcohol consumption or abdominal trauma. Her past and familial medical histories were normal. She was employed by profession, married and of medium socio-economic status. Physical examination revealed left sided abdominal distention, generalized abdominal pain without Blumberg’s sign and swelling in the legs. Laboratory tests reported BUN of 150 mg/dL, creatinine of 2.4 mg/dL, neutrophilic leukocytosis (WBC 21,120 10³/μL), C-reactive protein of 296.48 mg/L. The patient was initially managed with oxygen therapy, fluids, intravenous broad-spectrum antibiotics and bowel rest. Abdominal CECT showed a giant predominantly fluid oval shaped and ring-enhancing mass, located in the left hemi-abdomen and measuring 12cm × 21.6cm × 26.5 cm, compressing and displacing the surrounding organs and vessels (including the inferior vena cava) and suspected of pancreatic cystic neoplasm (Fig. 1 A–C). Measurement of intra-abdominal pressure (IAP) with a Foley manometer revealed a IAH of 22 mmHg leading to ACS with acute renal failure; abdominal perfusion pressure (APP) was 38 mmHg. The patient, after understanding the severity of her medical condition and accepting surgery, was taken emergently to the operating room by experienced general surgeons for decompressive laparotomy to improve visceral perfusion and resection of suspected pancreatic neoplasm under general anesthesia. The patient was placed in the supine position on the operating table: intraoperatively the giant fluid mass was found to be originate from the body and tail of the pancreas extending from the epigastrium to the pelvis (Fig. 2), compressing and displacing inferiorly the intestine and the left kidney. Because of the inability to exclude a cystic pancreatic neoplasm a distal pancreatecosplenectomy was performed. The giant cystic pancreatic mass contained about 4 L of turbid fluid, a sample of which was sent for cytology, biochemistry and microscopy culture. Patient was given total parental nutrition for four days, an IV injection of Levofloxacin 500 mg once daily and a SC injection of somatostatin analogue 0.2 mg thrice daily for 5 days. The postoperative course was uneventful without development of
pancreatic fistula or pancreatic endocrine/exocrine insufficiency. IAP decreased to 5 mmHg, mean arterial pressure (MAP) was 80 mmHg and APP 74 mmHg. The patient received pneumococcal, meningococcal and Hemophilus vaccination, was discharged on the 5th postoperative day in a stable condition after removal of abdominal drains and referred to Endocrinology Department. The surgical specimen consisted of a voluminous cystic pancreatic mass of 22 cm of greater diameter originating from pancreatic body and tail, measuring 10 cm and the spleen measuring 16 × 10 × 5 cm (Fig. 3). Pathological examination showed the presence of PP (Figs. 4 and 5). The patient tolerated the advice provided and after a follow-up of six months is asymptomatic.

3. Discussion

This clinical case describes a rare IGPP causing IAH, ACS and intestinal obstruction. PP is a localized fluid collection of pancreatic juice enclosed by a wall of fibrous or granulation tissue that generally arises as consequence of pancreatitis, pancreatic trauma or extrinsic obstruction of the pancreatic duct, although 16% of PPs are idiopathic as in our case report [5]. The incidence of PP is 0.5–1/100,000 adults per year but increases in patients affected by acute pancreatitis (5–16%) or chronic pancreatitis (20–40%) [1]. PPs are the most common pancreatic cystic lesions (75%) [6] and must be differentiated from true cysts and cystic pancreatic neoplasms [7]. While the majority of acute PPs resolves spontaneously over a period of 4–6 weeks, few persisting may require endoscopic, percutaneous or surgical drainage or resection if enlarged, symptomatic or complicated. PP may remain asymptomatic or cause abdominal pain, abdominal swelling, early satiety, nausea, vomiting, weight loss, upper gastrointestinal bleeding, palpable abdominal mass, jaundice and fever. PP can be complicated by rupture, infection, bleeding inside the cyst, esophageal obstruction, gastric or duodenal outlet obstruction, stenosis of common bile duct, cardiac compression, bilateral ureteral obstruction or necrosis, compression of large vessels, ascites or pancreatic-pleural fistula [6,8,9]. PP may also cause splenic rupture [10] as other pathologies [11,12]. In this case report the symptomatic IGPP was compressing and displacing the surrounding organs and vessels including the inferior vena cava leading to intestinal obstruction, IAH and ACS. IAH is a sustained or repeated pathologic elevation of IAP (n.v. 0–5 mmHg) ≥12 mmHg. ACS is a sustained IAP > 20 mmHg (IAH grades III), with or without an APP of <60 mmHg, associated with new organ dysfunction/failure [13]. The earliest manifestation of IAH is oliguria and acute kidney injury. IAH causes reduced perfusion to intra-abdominal and intra-thoracic organs but also affects the intra-thoracic organs and the intracranial pressure [14]. APP is an indicator of visceral perfusion: if less than 60 mmHg predicts the necessity for surgical decompression being a predictor for patient survival [14]. When IAH is not promptly recognized and treated leads to ACS, multiorgan dysfunction syndrome and death. In our case report IAP was 22 mmHg (IAH) causing acute renal failure and ACS, APP was 38 mmHg. Suspcion of PP is confirmed by biochemical and radiological findings. Nowadays there aren’t specific laboratory exams to establish a correct diagnosis, however a persistently elevated concentration of serum amylase and lipase can be observed in up to 50% of patients, like as alterations in liver function and leukocytosis as in our case report. Diagnosis of PP is accomplished often by CECT, ERCP, transcatheter ultrasound, endoscopic ultrasound or magnetic resonance cholangiopancreatography. Abdominal CECT represents the gold standard in diagnosing of PP providing important information about size,
location and relationship of the lesion with adjacent structures as in our clinical case. Management strategies of PP have changed and continue to evolve [15]. Uncomplicated and asymptomatic PP can be managed conservatively with enteral diet and analgesics irrespective of its size, location or extension to neighbouring structures [16] because of PP resolves spontaneously in up to 70% of the cases [17]; complicated or symptomatic PP needs intervention [18]. IGPP includes percutaneous, endoscopic or surgical drainage or resection. Indications for pancreatic resection include multiple or complicated PPs, PPs with associated biliary or pancreatic duct strictures, impossibility or failure of prior internal or external drainage and inability to exclude a cystic pancreatic neoplasm as in our patient [7, 18]. Pancreatic resection, as in our case report, have been used sporadically but persistently in most large series dealing with the treatment of PP: probably the high morality and mortality associated with pancreatic resection had led to uncertainty over the clinical indications for its use [18]. A collective series of 152 patients who underwent pancreatic resection for PP reported an overall 35% morbidity rate, 7% mortality rate and 8% recurrence rate [18]. In a ten-year series of 117 patients operated for a PP, 37 patients (32%) had emergency operation because of a cystic-related complication [19]. Although the majority complicated or symptomatic PPs can be managed by different drainage techniques, few require pancreatic resection performed with acceptable morbidity and mortality.

4. Conclusion

IGPP is an uncommon disease that may lead to intestinal occlusion, IH and ACS. Diagnosis in emergency is a challenge and if IGPP cannot be differentiated from cystic neoplasm it should be treated as malignant neoplasm and need surgical resection. Only few cases of pancreatic resection for PP are reported in the literature and the largest series are of 152 patients with acceptable morbidity and mortality.

Declaration of Competing Interest

All the authors certify that there is no conflict of interest regarding the material discussed in the manuscript.

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Ethical approval

Ethical approval has been exempted by our institution because this is a case report and no new studies or new techniques were carried out.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for the Editor-in-Chief of this journal on request.

Author contribution

Giuseppe Evola: Operated on the patient, drafting the manuscript, literature research.

Francesco Ferrara: Drafting the manuscript and literature research.

Mario Scrvaglieri: Operated on the patient, literature research.

Ezio Trusso Zirna: Operated on the patient, drafting the manuscript.

Salvatore Sarvà: Drafting the manuscript and literature research.

Luigi Piazza: Revising the manuscript.

Registration of research studies

This case report does not require registration as a research study.

Guarantor

The guarantor for this case report is Giuseppe Evola.

Provenance and peer review

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