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A rare case of pancreatic splenosis and a literature review

Splenosis is a clinical condition characterized by the presence of ectopic splenic tissue in the abdominal cavity occurring after abdominal trauma or surgical procedure involving the spleen. We present a case of an 80-year-old Caucasian male who underwent splenectomy at the age of 36, with a prior diagnosis of cancer of the left kidney associated with a pancreatic neoformation. This study compare our case with similar published and reviev the Literature.

KEY WORDS: Accessory spleen, Differential diagnosis, Kidney cancer,Laparotomy, Nephrectomy, Pancreas, Pancreatic splenosis, Splenosis

Introduction

Splenosis is a clinical condition characterized by the presence of ectopic splenic tissue in the abdominal cavity occurring after abdominal trauma or surgical procedure involving the spleen. Pancreatic splenosis is usually asymptomatic, but can be incidentally discovered during imaging studies; when mimicking pancreatic neoplasm, differential diagnosis may be difficult. We present a rare case of an 80-year-old with prior diagnosis of cancer of the left kidney and pancreatic isthmic splenosis mimicking a primary or secondary pancreatic cancer. We present the first case of pancreatic splenosis involving the isthmum. In this study, we compared our case with similar published cases of pancreatic splenosis.

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Case Report

An 80-year-old Caucasian male was admitted to our Institution with a urinary test positive for atypical cells. He complained of abdominal pain and macroscopic hematuria. His medical history included 15 pack-year smoking history, splenectomy at the age of 36 for a severe splenic injury, ischemic heart disease, abdominal aortic aneurism and hypertension. Physical examination was unremarkable and laboratory findings were normal except for hyponatremia. A thoracic-abdominal CT and Uro-CT revealed an abnormal left upper renal calyx, due to the presence of neoplasm. Multiple nodules of different sizes were also described in the abdominal and pelvic areas, in the splenic loggia, liver and mesorectal fat. Furthermore, an irregular hypovascular area was observed in isthmic region of the pancreas (Fig. 1). Abdominal magnetic resonance (MRI) confirmed the presence of a solid mass (26x18 mm) in the left kidney, presumed to be a primary urothelial cancer, and an undefined lesion within the pancreatic isthmus.

Operative Procedure: an open laparotomy was performed: surgical abdominal exploration identified numerous nodular formations spread throughout the abdomen,

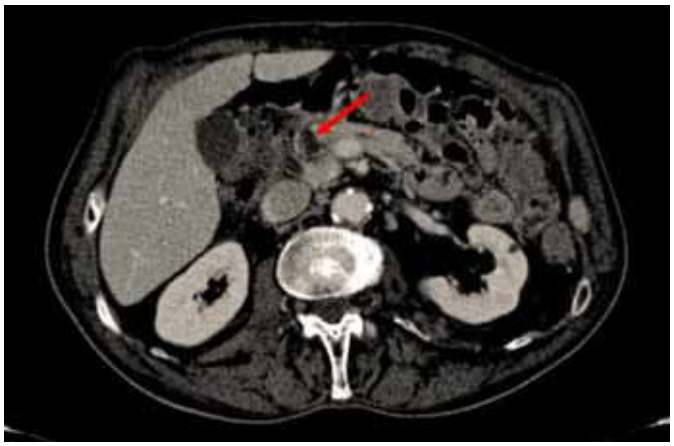

Fig. 1: Arterial CT scan irregular hypovascular area (arrow) in the isthmic region of the pancreas.

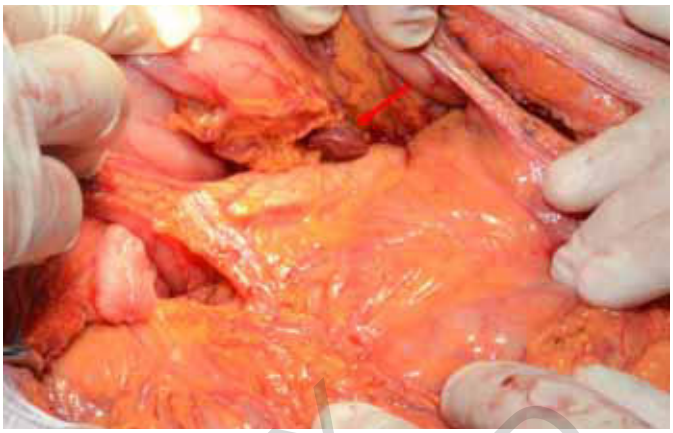

Fig. 2: Intraoperative visualization of the isthmic pancreatic lesion.

which resembled accessory spleens. An additional implant was noted growing in the lesser omentum lying on the superior margin of the pancreatic isthmus, therefore appearing as pancreatic neoplasm (Fig. 2). This lesion and few nodular formations were excised and sent for pathological examination. The operative procedure was completed performing a radical left nephrectomy. The post-operative course was uneventful and the patient was discharged in the tenth post-operative (P.O.) day.
Postoperative treatment consisted of anticoagulation (seleparine 0.6 ml/day for the course of the patient's hospitalization), antibiotic therapy (cephalosporine 2g/day for three days), and proton pump inhibitors (pantoprazole 40mg/day for the course of the patient's hospitalization). The pathological report described a high grade urothelial papillary carcinoma of the renal pelvis, which invaded into the muscular coat of the pelvis but did not involve the parenchyma and adipose tissue of the kidney (sec. UICC 2009: pT1 pN0 – G3 R0; Stage I), and confirmed the diagnosis of splenosis in all the removed lesions.

Review of the Literature

Scopus (www.scopus.com), Embase (www.embase.com), and Medline (www.ncbi.nlm.nih.gov/pubmed) databases were used for the international literature review. No language restrictions were applied. Manual searching of reference lists for relevant studies and previous reviews was also performed. The primary search was conducted for any combination of the words "pancreas" and "splenosis". Studies were included if they contained adequate information regarding symptoms, pancreatic splenosis characteristics, and the type of procedures performed. In the case of duplicate publications, the latest or the most

Table I - *Pancreatic splenosis: review of the literature*

Authors	Sex Age	Size (Cm)	Pancreatic Site	Previous Splenic Trauma	Time From Trauma*	Symptoms	Diagnosis
Fiamingo et al.[29], 2004	M, 52	3	Body	Splenectomy	31	Asymptomatic	Diagnostic laparoscopy
Rogers et al.[28], 2011	M, 64	5	Tail	Splenectomy	47	Asymptomatic	CEUSSPECT CT
Lèon et al.[26], 2012	M, 33	8	Tail	Splenectomy	4	Abd. Pain Nausea Vomiting Weight loss Inappetence	Open distal pancreasectomy
Priola et al.[25], 2013	M, 76	1.5	Tail	Splenectomy	2	Asymptomatic	^{99}mTC-sulfur colloid scintigraphy
De Robertis et al.[24], 2014	M, 53	2.8	Head	Splenectomy	20	Asymptomatic	CEUS + US guided FNA
Present Report	M, 80	1.6	Isthmus	Splenectomy	44	Asymptomatic	CT (thoracic-abdominal) Biopsy during laparotomy

* years

complete study was included. Studies that dealt with splenosis occurring at locations other than the pancreas were excluded. We identified 28 papers dealing with patients affected with splenosis. However, 14[1-14] papers were excluded because splenosis involved other structures without pancreatic involvement; 4[15-17,20] papers were excluded because they described cases of accessory spleen and not acquired splenosis, and 5[18,19,21-23] papers were excluded because the information were incomplete. To our knowledge, only 5[24-26,28,29] papers reported patients with similar clinical presentation to our own case (Table I).

Discussion

Splenosis is defined as an acquired condition characterized by heterotopic autotrasplantation of splenic tissue secondary to traumatic or iatrogenic splenic rupture[30]. A previous abdominal trauma with splenectomy was observed in nearly 93% of splenosis cases[25,26]. A differential diagnosis must be made between splenosis and accessory spleen. Whereas splenosis is defined as an acquired condition, accessory spleen refers to a congenital condition of ectopic splenic tissue[39], which has been reported in as many as 10% of the general population[27]. Fremont et al.[31] reported a variable interval between splenic trauma and splenosis diagnosis; the average time was 10 years with a range from 5 months to 47 years[28,34-35]. The most common abdominal localizations of splenosis are in the omentum, the mesentery and the peritoneum. Extraperitoneal implants in the thoracic cavity, subcutaneous tissue and even in the brain are rare, but have also been described.[11, 29,31-32,38] Accessory spleen, however, is most commonly localized in the splenic perihilar region (80%), followed by the pancreatic tail (17%)[27]. Accessory spleens are generally few in number, very rarely totaling more than six, while lesions of splenosis may occur in greater numbers and with greater variability in size, number and form[28]. An anatomical difference between splenosis nodules and accessory spleen may be noted in their vascularization: for the former, the blood supply generally derives from nearest blood vessels, whereas the latter, instead, are usually vascularized from a branch of the splenic artery[28]. Normally asymptomatic, splenosis can cause abdominal symptoms such as pain due to intestinal infarction or obstruction, hemorrhage, infection, and ureteral compression[24,31,36]. Furthermore, pancreatic localization may mimic primary neoplasia of the pancreas or metastases, usually from renal cell carcinoma. [25, 37] At CT scans, primary carcinoma of the pancreas as well as most metastases in the pancreas show a poor blood supply, however renal cell carcinoma metastases, which represent 4.5% of all pancreatic tumors, appear as hypervascular[37]. Pancreatic splenosis is a rare condition and, even when incidentally discovered during radiological examination, preoperative diagnosis is difficult[25,34]. Ultrasound (U.S.) can be a useful tool for the differential diagnosis of pancreatic lesions. Contrast enhanced CT, in the event of pancreatic nodules of splenosis, show nodular implants with homogeneous enhancement during arterial and portal phases. ^{99m}TC scintigraphy with heat-damaged erythrocytes and Indium 111-labeled platelets represents the radiological gold standard to confirm splenosis; at the same time this technique avoids invasive diagnostic procedures, such as open biopsy or surgical excision [25,31]. In the case reports published in the literature, the preoperative diagnosis of splenosis was possible in three patients, while in the remaining two cases the diagnosis was accomplished only by pathological examination after surgery (Table I). Priola et al.[25] recommend ^{99m}Tc-sulfur colloid scintigraphy as a second exam to evaluate pancreatic lesions of new onset in patients with recent splenectomy. When diagnostic imaging is unclear, Ardengh et al.[19] investigated the usefulness of endoscopic ultrasound-guided fine-needle aspiration (FNA) to confirm the diagnosis of neuroendocrine tumor or pancreatic splenosis. Contrast-enhanced ultrasound (CEUS) represents another non-invasive radiological method for the differential diagnosis between accessory spleen and splenosis: poorly developed capsule and vascularization independent of the splenic artery are both characteristics of splenosis[28]. Rogers et al.[28] used CEUS as the method of choice for the diagnosis of splenosis in conjunction with ^{99m}TC scintigraphy with heat-damaged erythrocytes and SPECT; the author has proposed using CEUS to replace nuclear imaging. De Robertis at al.[24], in a diagnostic study, demonstrated that CEUS shows a characteristic pattern of uniform enhancement in the late phase in cases of pancreatic splenosis, but still utilized US guided FNA to confirm diagnosis. Cytology of splenosis is characterized by an organized, polymorph, lymphoid cellular population without atypia. When splenosis is confirmed in asymptomatic cases, no further treatment is necessary [31]. On the other hand, in symptomatic cases surgical excision of the mass is the treatment of choice [26]. León et al. [26] reported a rare case of symptomatic pancreatic splenosis, presenting with abdominal pain, weight loss, inappetence, nausea and vomiting. In this case, due to the suspicion of a neuroendocrine pancreatic tumor, CT imaging study was acquired, supplemented with endoscopic US and FNA; an open distal pancreasectomy was performed, which resulted in the resolution of symptoms. Later pathological examination confirmed diagnosis as pancreatic splenosis. In our own patient, due to the concurrent presence of a kidney cancer, an operative procedure was mandatory and allowed an accurate differential diagnosis of the pancreatic lesion and the evaluation of the several abdominal implants.

Conclusions

Pancreatic splenosis is a very rare condition. Nevertheless, this report confirms the necessity of considering spleno-

sis in the differential diagnosis of a pancreatic mass in a patient with a history of splenic trauma. CEUS should be considered as a diagnostic option in such cases, however further studies are necessary to demonstrate whether or not this examination is accurate enough to replace ^{99m}TC scintigraphy with heat-damaged erythrocytes, Indium 111-labeled platelets or histological examination to confirm diagnosis.

Riassunto

La splenosi addominale è una condizione clinica rara, caratterizzata dalla presenza di impianti ectopici di tessuto splenico all'interno cavità addominale in seguito lesioni spleniche traumatiche o post-chirurgiche.
in questo articolo presentiamo il caso di un maschio caucasico di 80 anni, precedentemente sottoposto a splenectomia per trauma, che si presenta alla nostra osservazione in seguito a riscontro di una neoplasia del rene sinistro associata ad una neoformazione pancreatica sospetta per cancro.
Discussione: la splenosi è una condizione benigna, generalmente asintomatica.
I noduli di splenosi si distinguono dalle milze accessorie per l'assenza dell'ilo vascolare. La splenosi pancreatica può mimare la presenza di neoplasie o impianti metastatici a livello del pancreas e l'esame TC, permette di differenziare noduli di splenosi da lesioni neoplastiche pancreatiche; nel caso proposto, la lesione pancreatica istmica presentava un comportamento TC riferibile a lesione primitiva del pancreas.
Nel sospetto di splenosi, la scintigrafia con piastrine marcate con Tecnezio 111 e eritrociti marcati con Tecnezio 99 rappresentano il gold standard nella diagnosi preoperatoria. Nel nostro caso si è stato dirimente l'esame istologico intraoperatorio in corso di nefrectomia che ha dimostrato la localizzazione pancreatica di tessuto splenico.

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