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Scientific letters

# Presentation of pancreatic sarcoidosis as a retroperitoneal infiltrative mass☆

## Presentación de sarcoidosis pancreática como masa infiltrativa retroperitoneal

Sarcoidosis is a chronic granulomatous inflammatory process of unknown etiology that can develop in any organ of the body[1]. Gastrointestinal involvement occurs in less than 1% of patients diagnosed with sarcoidosis, and pancreatic involvement is unusual[2]. The symptoms of pancreatic invasion can be confused with episodes of pancreatitis or pancreatic carcinoma, and its preoperative diagnosis is complicated[1,2]. We present the case of a pancreatic mass with no clear preoperative diagnosis and a final result of pancreatic sarcoidosis.

The patient is a 78-year-old woman with an allergy to non-steroidal anti-inflammatory drugs and a history of hypertension, type 2 diabetes mellitus and atrial fibrillation. She reported increasing abdominal pain over the previous 3 months located in the epigastrium and left hypochondrium, which did not subside with habitual analgesia and was associated with anorexia, nausea, vomiting and a weight loss of 4 kg. Lab work-up showed no relevant alterations. Thoracoabdominal CT scan revealed a 60 mm solid vascularized lesion in the portal phase between the greater curvature of the stomach and the splenic hilum, which it displaced without infiltrating. The lesion presented small satellite and periceliac lymphadenopathies. The findings indicated a possible stromal tumor (GIST) without ruling out association with the pancreas (Fig. 1). Endoscopic ultrasound-guided fine-needle aspiration provided evidence of lymphoid tissue with aggregates of epithelioid histiocytes, CD68+ and negative GIST markers (chronic inflammatory reaction with non-caseating histiocytic granulomas). Given the uncertain diagnosis, the patient's symptoms and the impossibility to rule out malignancy, we decided to operate after presenting the case to a multidisciplinary committee. The initial laparoscopic approach was converted to a bilateral subcostal laparotomy due to hemodynamic instability and episodes of sustained ventricular tachycardia. We observed diffuse retroperitoneal granulomatous infiltrate and 3.5 L of ascites. Intraoperative peritoneal biopsies were negative for malignancy and reported as fibroconnective tissue with chronic inflammation, lymphoid and presence of granulomas with multinucleated giant cells. Distal pancreatectomy and splenectomy were performed.

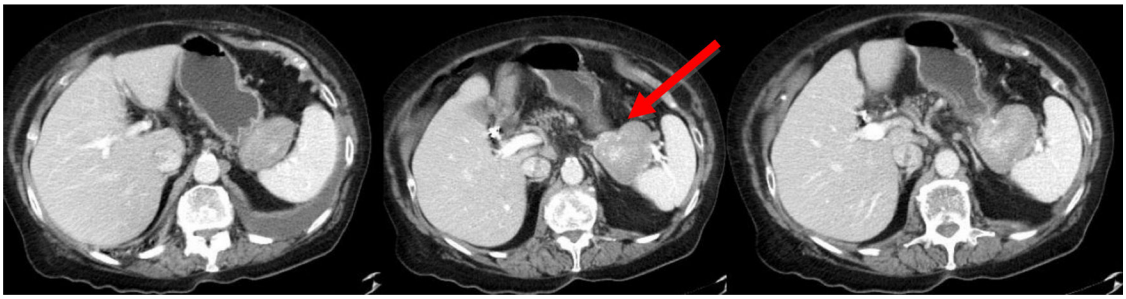

**Figure 1 – A) Abdominal CT with intravenous contrast showing periduodenal collection and intra-abdominal free fluid. The**

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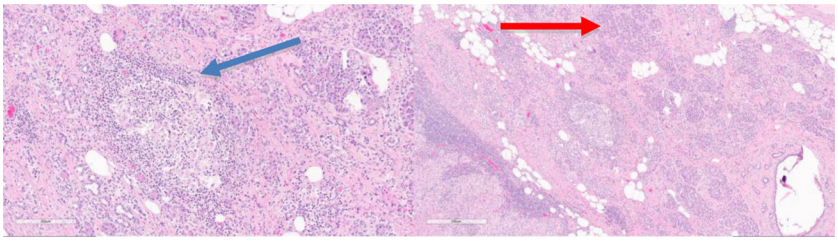

**Figure 2 – Diagram of the surgery performed: division of the neck of the pancreas, suture of the proximal pancreas, and end-to-side duct-to-mucosa Roux-en-Y pancreaticojejunostomy.**

During postoperative ICU care, the patient required vasoactive drugs and amiodarone infusion due to an episode of atrial fibrillation. After favorable evolution, the patient was moved to the ward on the 3rd postoperative day and discharged on the 7th day without further incidents (Clavien-Dindo IVa). The histology study showed a smooth tumor measuring 60 × 50 mm dependent on the pancreas, with a focus of intratumoral calcification. Microscopically, the peripancreatic lymph nodes were observed to have granulomas rich in epithelioid cells, multinucleated giant cells, and chronic inflammation without necrosis. The morphology of the lesion indicated sarcoidosis (Fig. 2). The extending diagnostic tests of the histochemical study for acid-fast bacilli (Ziehl-Neelsen and Fite-Faraco) were both negative, which confirmed the final diagnosis of sarcoidosis. Subsequent follow-up visits were favorable, with no surgical or endocrine/metabolic complications.

Sarcoidosis is a chronic granulomatous disease of unknown cause characterized by epithelioid non-caseous granulomas with accumulation of mononuclear phagocytes and T-lymphocytes. It can appear systemically or with involvement of specific organs: lungs, lymph nodes, eyes, skin, nervous system and heart. In 1937, Nickerson published the first case of pancreatic involvement after the autopsy of a patient with systemic sarcoidosis[3]. Pancreatic localization of sarcoidosis is uncommon and estimated at 1%–4% of patients with systemic sarcoidosis and autopsy after death, with slightly higher figures in black women aged 50–70 years. Less than 50 cases with exclusively pancreatic involvement have been described in living patients[4,5]. Symptomatic pancreatic involvement is unusual and is usually due to either infiltration of the parenchyma or ductal involvement, causing clinical symptoms of acute pancreatitis, obstructive jaundice and hepatic biochemical alterations[1,2,5]. The presentation is varied, ranging from simple inflammatory processes or a nodular lesion to a pancreatic mass, which could sometimes mask a pancreatic carcinoma[6]. In more than 50%, they are located in the head of the pancreas in association with abdominal lymphadenopathies[7].

Despite not having a definite preoperative histopathological diagnosis, abdominal CT scan, PET/CT or MRI, combined with ultrasound-guided FNA, help direct the diagnosis[1,2,8]. Possible analytical alterations include hyperbilirubinemia with a direct component, elevated amylase, lipase and hypercalcemia due to increased calcitriol levels in the macrophages of sarcoid granulomas and due to increased PTH-related proteins, which favors the conversion of vitamin D to active calcitriol[9]. Because of the differential diagnosis with pancreatic neoplasms, many patients are definitively diagnosed after surgery[1,2]. Noncaseating granulomas can be found in patients with malignant neoplasms like lymphomas and carcinomas, representing an immune reaction to a tumor antigen[9].

Although there is controversy regarding treatment, long-term corticosteroid therapy is the treatment of choice as it results in biochemical improvement and reduced size of the tumor mass without improving fibrosis. The use of corticosteroids combined with ursodeoxycholic acid is recommended in symptomatic cases with marked cholestasis and/or high risk of developing liver complications[10]. Surgery is reserved for patients who present a significant intra-abdominal mass and involvement of critical organs, with associated symptoms and systemic repercussions[1,2]. Isolated histological involvement does not require the start of treatment, and asymptomatic patients can be observed.

Thus, although rare and infrequent, pancreatic sarcoidosis must be included in the differential diagnosis of a pancreatic mass. The evaluation of this type of exceptional cases in referral centers and by multidisciplinary committees can avoid errors in diagnosis and unnecessary treatments.

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Miguel Cantalejo Díaz[a,*], Ana Palomares Cano[a], Carlos Hörndler Algarate[b], Luis Antonio Ligorred Padilla[a], Mario Serradilla Martín[c]

[a]*Servicio de Cirugía General y del Aparato Digestivo, Hospital Universitario Miguel Servet, Zaragoza, Spain*
[b]*Servicio de Anatomía Patológica, Hospital Universitario Miguel Servet, Zaragoza, Spain*
[c]*Instituto de Investigación Sanitaria Aragón, Servicio de Cirugía General y del Aparato Digestivo, Hospital Universitario Miguel Servet, Zaragoza, Spain*

*Corresponding author: miguelcd_10@hotmail.com (M. Cantalejo Díaz).

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# Doege-Potter syndrome due to a hepatic solitary fibrous tumour☆

## Síndrome de Doege-Potter secundario a tumor fibroso solitario hepático

Solitary fibrous tumours (SFT) of the liver are uncommon neoplastic lesions with benign histological characteristics that develop in the mesenchymal tissue[1,2]. This type of tumour develops most frequently in locations such as the pleura, peritoneum, thymus or meninges[1]. Their intrahepatic growth is extremely rare, with only a few dozen cases published in the literature[1,2].

This type of tumour has a 5-year survival rate of around 85%, with a 5-year and 10-year risk of metastasis of 26% and 46%, respectively[3]. The main risk factors for a poor prognosis are tumour size and high mitotic indices.

In 5% of cases, these tumours appear in association with a paraneoplastic syndrome known as Doege-Potter syndrome[3], which presents with severe hypoglycaemia and was first described in 1930[4]. This syndrome is more frequent in patients between the ages of 60 and 80[4] and is associated with a worse prognosis, even more so if the location of the tumour is extrapleural[3,4].

This paraneoplastic syndrome presents with severe hypoglycaemia and decreased levels of C-peptide, insulin and IGF-I in the blood due to the secretion of IGF-II by the tumour, which binds to IGF-I receptors[4]. This union not only causes hypoglycaemia but is also associated with an increase in the number of mitoses in the tumour and its malignant transformation.

We present the case of an 83-year-old patient with a history of dyslipidaemia and altered baseline fasting glucose who came to the emergency room after a routine blood glucose test of 33 mg/dL and a large indurated epigastric mass. A thoracoabdominal CT scan revealed a mass suggestive of gastric GIST in the gastrohepatic ligament measuring 16 × 12 × 14 cm that was hypervascular and had interior

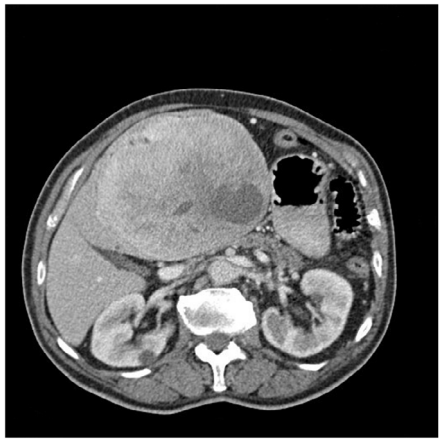

**Fig. 1 – Sagittal CT scan, where the lesion is observed in close contact with the gastric wall.**

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