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ARP Case Report Nº 16: Serous Cystadenoma of the Pancreas and Lung Sarcoidosis

Caso Clínico ARP Nº16: Cistoadenoma Seroso do Pâncreas e Sarcoidose Pulmonar

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Abstract

69 year-old female referred to our institution for abdominal discomfort, epigastric pain and weight loss (10Kg in 6 months).
Abdominal e thoracic CT was performed and later on additional abdominal MR.

Keywords

Pancreas; Cystadenoma; Sarcoidosis; Lymphadenopathy.

Resumo

Doente de 69 anos referenciada para a nossa instituição por desconforto abdominal, dor epigástrica e perda ponderal (10kg em 6 meses). Sem antecedentes de patologia ou internamentos prévios.
Realizou TC computadorizada abdominal e torácica. Posteriormente realizou estudo adicional de RM abdominal

Palavras-chave

Pâncreas; Cistadenoma; Sarcoidose; Linfadenopatia.

A 69 year-old female was referred to our institution for vague abdominal discomfort, occasional epigastric pain and marked weight loss.
There were no relevant findings in the patient's medical history or physical examination.
She underwent routine abdominal ultrasound which revealed a heterogeneous mass in the pancreatic body and further examination was warranted.
Abdominal CT (computed tomography) was performed, with pre and post-contrast images demonstrating a lobulated mass in the pancreatic body, measuring 50x51mm (Fig 1a). The lesion has a homogeneous appearance, centrally hypodense, with mainly peripheral enhancement, with central areas of water-density(Fig 1b,1c). No calcified foci are seen.
There are no signs of vascular invasion, with pancreatoduodenal vessels forming a peripheral "mesh" around the pancreatic mass, nor distal parenchymal atrophy.
These findings were suggestive of a serous cystadenoma, confirmed with MR (Fig 2A and B).
Mild splenomegaly and multiple retroperitoneal and retrocrural adenopathy were also seen (Fig 1C). Thus, a thoracic acquisition was added, demonstrating extensive mediastinal and hilar adenopathies as well as irregular perilymphatic nodular thickening with an upper lobe predominance, mainly in the upper left lobe, with multiple centrilobular nodules and fine interlobular septal thickening (Fig 3A). Nodular thickening of the major fissures is also seen (Fig 3B).

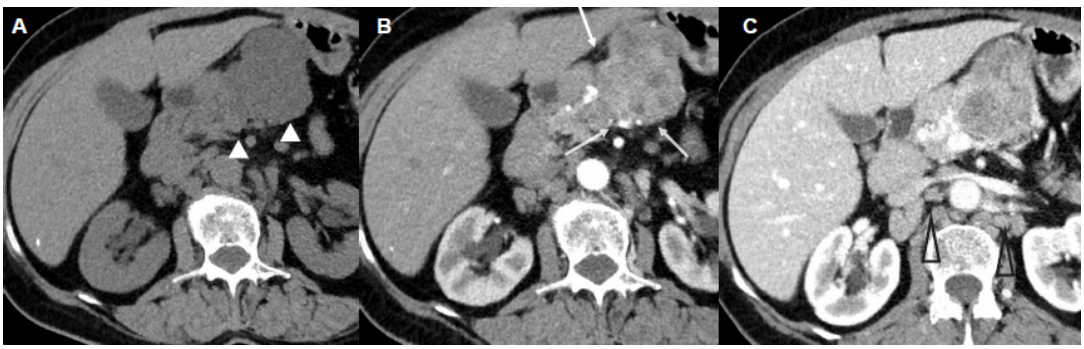

Figure 1 – CT of serous cystoadenoma of the pancreas
A and B - Pre and post-contrast images show a lobulated mass in the pancreatic body with lobulated contours. The lesion is homogeneously hypodense (closed arrows) with marked peripheral enhancement. Due to abundant fibrous septa (straight arrows), the lesion appears solid. C - Multiple retroperitoneal lympadenopathy are seen (black arrowheads), as well as retrocrural (not shown) which prompted additional thoracic imaging.

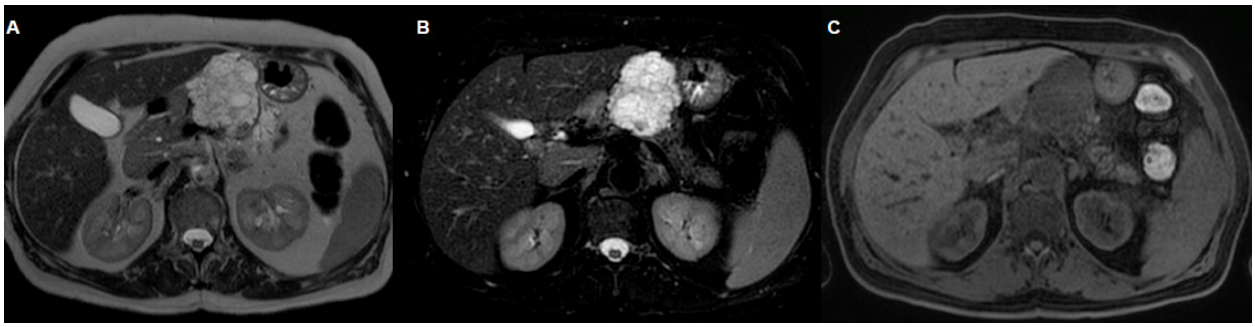

Figure 2 – Abdominal MRI of serous cystadenoma
A and B - Axial T2-weighted HASTE and FATSAT images show that the pancreatic mass has marked T2 hyperintensity and small hypointense septa, demonstrating multiple small cysts with a "cluster" like appearance. There is no visible communication with the pancreatic duct. C - Pre-contrast T1-weighted image shows homogeneous hypointensity of the pancreatic mass.

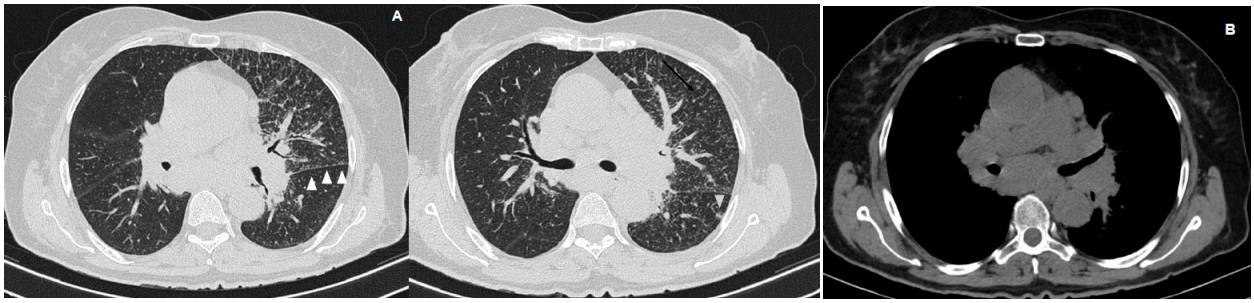

Figure 3 – Lung sarcoidosis on HRCT
A- There is nodular thickening of the major left fissure (white arrowheads) as well as regular thickening of the interlobular septa. There are multiple small centrilobular nodules in the upper left lobe (long black arrow) with a perilymphatic distribution and a ground glass nodular opacity in the upper segment of the left lower lobe (arrowhead). B - Extensive mediastinal and hilar lymphadenopathy.

EUS guided biopsy of the abdominal adenopathies was performed revealing multiple non caseous granulomas, findings suggestive of sarcoidosis. Moreover, high levels of angiotensin conversion enzyme (ACE) on peripheral blood tests was also present.

Discussion

Pancreatic serous cystadenomas are benign neoplasms, typically encountered in older women (60 y.o.). They're composed of multiple small cysts (<1cm) usually arranged in a "honeycomb" like pattern, with a peripheral epithelial lining and central fibrous septae, that can form a central scar, sometimes calcified.
Most lesions demonstrate a slight predominance for the pancreatic head and are usually incidental findings. Rarelly, mass-effect related symptoms can occur due to displacement of surrounding structures. Distal atrophy of the pancreas and ductal dilation, wether of the pancreatic or biliary ducts is uncommon.
Although most cases are sporadic, serous cystadenomas can also occur in the setting of von Hippel-Lindau disease. On imaging, the appearance depends on the number of fibrous septae and degree of enhancement. Lesions are usually lobulated, hypodense on pre-contrast CT, sometimes with central calcification with a stellate pattern. When large numbers of microcysts are present, these lesions may appear solid in nature. Thus, MR imaging can be useful in the diagnosis, demonstrating small clusters of fluid-containing cysts on T2-weighted images.
Due to their benign nature, most lesions are observed without treatment. If symptomatic, surgical resection can be considered and no recurrence has been reported once resected.

In this case, sarcoidosis was an incidental finding. Lung and mediastinal involvement of this non-caseating granulomatous multi-systemic disease occurs in 90% of patients with sarcoidosis, usually between 20-40 years of age , with a slight female predominance.
On high-resolution CT, parenchymal findings of perilymphatic irregular nodular thickening with preferential upper lobe distribution, centrilobular nodules, interlobular septae and fissure thickening is characteristic.
Ancillary findings include miliary and ground-glass opacities. In stage IV disease, findings of pulmonary fibrosis can be seen. Differential diagnosis depends on the disease stage and consequently the different imaging findings, but in this case tuberculosis should be considered. High levels of ACE favor sarcoidosis and the presence of granulomas on biopsy aided the diagnosis. Treatment is reserved for symptomatic patients and the mainstay are corticosteroids, with remission depending on the stage of disease at presentation.