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Images of Interest / Imagens de Interesse

Complete Pancreatic Fatty Replacement in a Patient with Cystic Fibrosis

Substituição Lipomatosa Pancreática Completa num Doente com Fibrose Quística

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Abstract

The pancreas is the abdominal organ most frequently affected in cystic fibrosis and pancreatic lipomatosis is the most common abnormality seen in these patients. We present a case and images of a young male patient with cystic fibrosis and associated complete adipose replacement of the pancreas.

Keywords

Pancreas; Pancreatic lipomatosis; Cystic fibrosis.

Resumo

O pâncreas é o órgão abdominal mais frequentemente envolvido na fibrose quística, sendo a lipomatose a anomalia pancreática mais comum nestes doentes. Apresentamos um caso clínico e respetivo estudo imagiológico de um jovem adulto com fibrose quística e infiltração adiposa completa do pâncreas.

Palavras-chave

Pâncreas; Lipomatose pancreática; Fibrose quística.

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Case

A 21-year-old male patient with cystic fibrosis (CF) presented to consultation with a history of steatorrhea for four months, without weight loss or abdominal pain. He had been submitted to a lung transplant two years before. The blood tests were unremarkable, except for a normocytic normochromic anemia (hemoglobin level: 12.4 g/dL). The stool test revealed a markedly reduced elastase level (<15μg/g versus a reference level >200 μg/g), in keeping with severe exocrine insufficiency.
Abdominal computed tomography (CT) followed by magnetic resonance with cholangiopancreatography (MRCP) were requested to evaluate the pancreatic parenchyma. The CT (Fig. 1) and MR (Fig. 2) showed an enlarged pancreas, with loss of its lobulated contour and total fatty replacement of the parenchyma. There were no calcifications or other abnormalities of the main pancreatic duct.

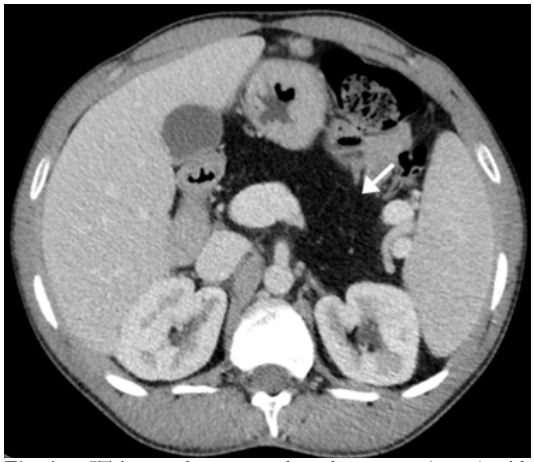


Fig. 1 – CT image shows an enlarged pancreas (arrow) with diffuse fat density. There is also splenomegaly and portal vein enlargement, evocative of portal hypertension.

Discussion

Complete fatty replacement of the pancreas is typically associated with genetic conditions, such as CF, Shwachman-Diamond syndrome and Johanson-Blizzard syndrome. A lower degree of infiltration can be seen more commonly in elderly patients and in those with obesity, diabetes mellitus, steroid intake or chronic pancreatitis.[1]

The pancreas is the abdominal organ more frequently affected in patients with CF.[2] Moreover, fatty replacement of the pancreas can be found in 51 to 75% of the patients with CF and it is the most common pancreatic abnormality seen in this disease.[1] Mutations in the CFTR gene (CF transmembrane conductance regulator) lead to the impaction of thick mucus in the pancreatic ducts, which results in ductal obstruction followed by progressive fat deposition in the acinar cells. In fact, the degree of

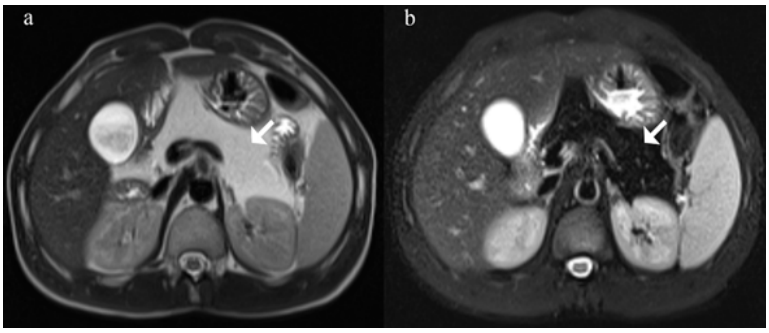


Fig. 2 – MR T2-weighted images without (a) and with fat suppression (b) show pancreatic enlargement (arrows), with loss of the normal pancreatic lobular contour and complete fatty replacement. The pancreatic signal intensity is similar to that of the retroperitoneal fat.

fat infiltration on imaging correlates with the degree of exocrine insufficiency.[2,3]

The fatty replacement of the pancreas in patients with CF can be focal or diffuse. Furthermore, it can be associated with pancreatic atrophy, but a smaller portion of patients may present a pseudohypertrophic appearance of the pancreas,[2] as seen in this case.

Other pancreatic manifestations of CF are chronic pancreatitis, pancreatic fibrosis and pancreatic cystosis.[1]

Exocrine insufficiency management in patients with CF relies on pancreatic enzyme replacement associated with lifestyle modifications.[2]

Ethical disclosures / Divulgações Éticas

Conflicts of interest: The authors have no conflicts of interest to declare.

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Confidentiality of data: The authors declare that they have followed the protocols of their work center on the publication of data from patients.

Confidencialidade dos dados: Os autores declaram ter seguido os protocolos do seu centro de trabalho acerca da publicação dos dados de doentes.

Protection of human and animal subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

Protecção de pessoas e animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pelos responsáveis da Comissão de Investigação Clínica e Ética e de acordo com a Declaração de Helsínquia da Associação Médica Mundial.

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