Case 9519

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Pancreaticoliths with obstructive pancreatic lipomatosis

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Section: Abdominal imaging
Area of Interest: Abdomen
Procedure: Diagnostic procedure

Imaging Technique: CT

Special Focus: Calcifications / Calculi Case Type:

Clinical Cases

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Patient: 50 years, female

Clinical History:

A 50-year-old lady presented with chronic upper abdominal pain with episodes of exacerbation and remission with the use of oral analgesics. She also complained of weight loss and steatorrhoea for the past 6 months. On examination she was found to have elevated blood sugar 280 mg/dl.

Imaging Findings:

CT study of abdomen revealed multiple intraductal calculi in the region of pancreatic head. These were associated with near total lipomatosis of the pancreatic parenchyma. No significant normal pancreatic glandular tissue was seen throughout the pancreatic bed. In light of the clinical details and based on the imaging findings, the diagnosis of pancreaticoliths with obstructive pancreatic lipomatosis was made.

Discussion:

Pancreatic lipomatosis refers to replacement of a part or of nearly all glandular components of the pancreas by lipomatous tissue. It has also been termed as fatty replacement, fatty infiltration, and adipose atrophy of the exocrine pancreas [1]. Pancreatic lipomatosis can be partial or complete.

Partial or incomplete lipomatosis is relatively common and is frequently encountered in diabetic, obese, and elderly patients [1, 3]. At cross-sectional imaging, 4 different types of uneven pancreatic lipomatosis have been described [6]: Type-1a (35% of cases) characterised by fatty replacement of the head with sparing of the uncinate process and peribiliary region; type-1b (36%), replacement of the head, neck, and body, with sparing of the uncinate process and peribiliary region; type-2a (12%), replacement of the head & the uncinate process, and sparing of the peribiliary region; and type-2b (18%), by near-total replacement of the pancreas with sparing of the peribiliary region.

Complete lipomatosis of pancreas is uncommon. It can be associated with congenital syndromes such as cystic fibrosis, Shwachman-Diamond or Johanson-Blizzard syndrome [6]. It has also been reported in patients with obstruction of the pancreatic duct by a tumour or a calculus [1-3, 5]. The exact pathogenesis of pancreatic lipomatosis is still not well established. The majority of the patients remain asymptomatic, however, complete lipomatosis can lead to exocrine insufficiency with resultant malabsorption and steatorrhoea [3]. These patients generally have a poorer outcome and frequently need pancreatic enzyme supplementation in combination with strict dietary modifications. On ultrasound, pancreatic lipomatosis manifests as hyperreflective pancreas, at times making

differentiation difficult from the adjacent retroperitoneal fat. Cross-sectional imaging is particularly useful in confirming pancreatic lipomatosis and also detecting specific aetiologies such as obstructive calculi or neoplasms. The duct in case of obstructive calculi and neoplasm is characteristically stenotic rather than dilated, and typically has a smooth wall [3]. Complete lipomatosis needs to be distinguished from agenesis of dorsal pancreas. Dorsal pancreatic agenesis is characterised by the absence of body and tail of pancreas and frequently the distal pancreatic is filled by stomach or intestine (dependent-stomach or dependent-intestine signs), which abut splenic vein. In contrast, in case of lipomatosis abundant fat tissue is observed anterior to the splenic vein [4].

It is thus imperative for the radiologist to be attentive to this condition and carefully look for any obstructive pancreatic ductal pathology in patient with lipomatosis. The condition, if not timely detected and treated, can result in pancreatic exocrine insufficiency and malabsorption [3].

Differential Diagnosis List: Pancreaticoliths with obstructive pancreatic lipomatosis, Pancreatic atrophy, Dorsal pancreatic agenesis

Final Diagnosis: Pancreaticoliths with obstructive pancreatic lipomatosis

References:

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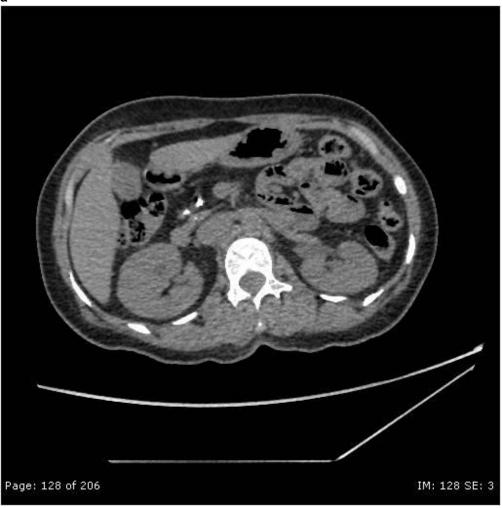


Description: CT scannogram reveals subtle calcific foci overlying the right twelfth rib. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: CT scannogram reveals subtle calcific foci overlying the right twelfth rib (arrow). **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.

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Description: Unenhanced CT image shows multiple calcific foci in the pancreatic head region surrounded by fat. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: Unenhanced CT image shows multiple calcific foci in the pancreatic head region surrounded by fat. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: The normal pancreatic glandular tissue is not discernable. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.

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Description: The pancreatic bed shows fat instead of normal pancreatic glandular tissue. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: Fat-attenuation tissue is seen replacing the pancreatic glandular parenchyma. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.

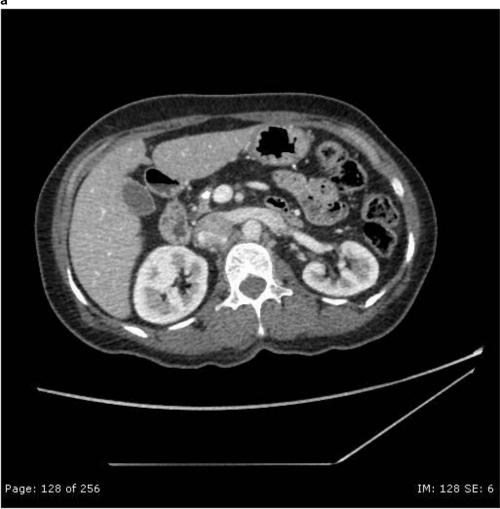


Description: Near total pancreatic lipomatosis is seen except for suggestion of a thin sliver of periductal parenchyma. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.

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Description: Pancreatic bed shows fatty attenuation tissue. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: No normal pancreatic parenchyma is seen in front of portal vein formation. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: The pancreatic bed shows fat lying anterior to the splenic vein and posterior to the stomach. Dependent stomach sign is absent, thus ruling out dorsal agenesis of the pancreas. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.



Description: Dependent stomach sign is negative, thus ruling out dorsal agenesis of the pancreas. **Origin:** Ankur Arora, Radiodiagnosis, ILBS Hospital, New Delhi, India.