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European Journal of Radiology Extra 62 (2007) 19–23

EUROPEAN JOURNAL OF RADIOLOGY EXTRA

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An unusual case of cystic dystrophy of the duodenal wall

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Received 1 September 2006; received in revised form 11 January 2007; accepted 12 January 2007

Abstract

Cystic dystrophy of the duodenal wall (CDDW) is an infrequent disease characterized by a chronic inflammation of the aberrant pancreatic tissue in the duodenal wall. A 60-year-old woman was admitted with several months of upper right abdominal discomfort, vomiting after meals and a 15 kg weight loss. The diagnosis of a CDDW was initially suspected in an abdominal US examination, which revealed an oval solid mass (4.3 cm × 2.7 cm) with a considerable cystic component between the slightly enlarged pancreatic head and the thickened medial wall of the duodenal vertical branch. A contrast-enhanced CT revealed a fibrous parietal thickening and a big ovoid mass of fluid density, divided by the septa, in the space between the pancreatic head and the second portion of the duodenum. The uncinate process and lower part of the head were enlarged, with bumpy contours, non-uniform fibrosis and a great number of calculi in the parenchyma, whereas the body, tail and upper parts of the pancreatic head remained unaffected. The diagnosis of CDDW was confirmed by an endoscopic US and pylorus preserving pancreatoduodenectomy.

The case described is of interest because of an unaffected major papilla and an unusual volume of orthotopic pancreas involvement by chronic inflammation.

Keywords: Cystic dystrophy of the duodenal wall; Computed tomography; Ultrasonography; Endoscopic US; Pylorus preserving pancreatoduodenectomy

1. Introduction

Cystic dystrophy or chronic inflammation of the aberrant pancreatic tissue in the duodenal wall, is a relatively rare disease in the practice of physicians and radiologists [1,2], despite the fact that a heterotopic pancreatic tissue is found in the duodenum in 25–35% of all cases of pancreatic ectopy in the digestive tract [3,4]. The heterotopic pancreas is usually functioning, and the development of acute and chronic pancreatitis in it is even more probable than in the orthotopic gland as a result of an underdeveloped duct system [1]. The progression of ectopic pancreatitis associated with increasing cystic formation could lead to a blockade of the major or minor duodenal papilla and subsequent chronic pancreatitis in the pancreas proper [5]. Furthermore, a malignant transformation of the aberrant pancreas is not a rare occurrence [6–8]. It is essential to carry out a timely and sharp diagnosis of this condition as it often defines the surgical tactics [9–12].

The purpose of this report is to present an unusual case of cystic duodenal dystrophy.

2. Case report

A 60-year-old woman was admitted with several months of upper right abdominal discomfort, periodical nausea and vomiting after meals and a 15 kg weight loss. The patient had no history of alcohol and tobacco abuse. A physical examination revealed no cardiac, respiratory or urinary disorders. The pulse was 78 p/min, the rhythm was sinusoid, and the blood pressure was 100/70 mmHg. There was some tenderness in the right quadrant of the upper abdomen without any distinct palpable mass.

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doi:10.1016/j.ejrex.2007.01.004

Hematological and blood bio-chemical tests did not show any considerable shifts, including serum amylase, bilirubin, alanine aminotransferase and aspartate aminotransferase and gamma GT levels. The urine had a pH 7 and a specific gravity of 1.016. The sediment contained 1–2 white cells per high-power field.

An ultrasonographic examination (including "4D-reconstruction") revealed a liver of normal size and structure, no stones in the gall bladder, non-dilated main pancreatic, intra- and extrahepatic bile ducts. The pancreas was unchanged in size and structure for the most part of the gland, excluding fibro-calculous parenchyma of the uncinate process of the pancreatic head. There was an oval solid mass (4.3 cm × 2.7 cm) with considerable cystic component between the slightly enlarged (3.1 cm) pancreatic head and the medial wall of the duodenal vertical branch, which appeared noticeably thickened. There were no signs of bloodflow in the thin isoechoic internal septa of the cyst (Fig. 1).

An upper GI endoscopy revealed deformation of the second portion of duodenum at the expense of its medial wall (Fig. 2) with superficial erosions. The unchanged main papilla was located above this affection. Biopsy of the duodenal mucosa detected moderate inflammation.

A barium meal did not reveal evacuation disorders.

Contrast-enhanced CT examination revealed fibrous parietal thickening and a big ovoid mass of fluid density, divided by the septa, in the space between the pancreatic head and the second portion of the duodenum. The uncinate process and the lower part of the head were enlarged, with bumpy contours, non-uniform fibrosis and a great number of calculi in the parenchyma (Fig. 3). The body, tail and upper parts of the pancreatic head were unaffected (Fig. 4), and there was a distinct border between the affected and intact aspects of the pancreatic head (Fig. 5). The gastroduodenal artery was shifted notably forwards and to the left (Fig. 6).

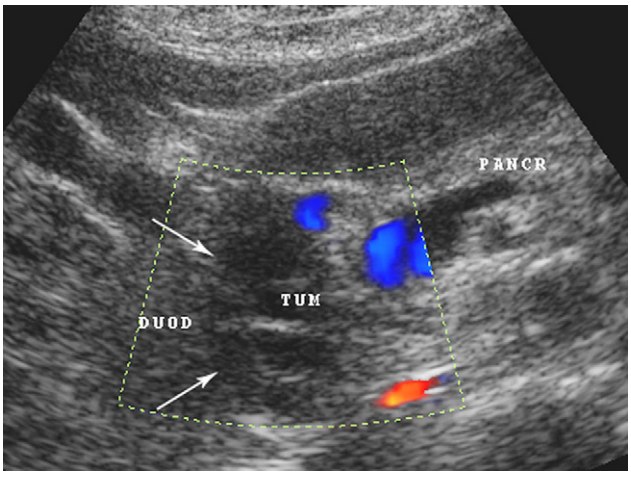

Fig. 1. US with CDI. Cyst in duodenum wall (arrow). There are no signals of bloodflow in the thin isoechoic internal septa of the cyst.

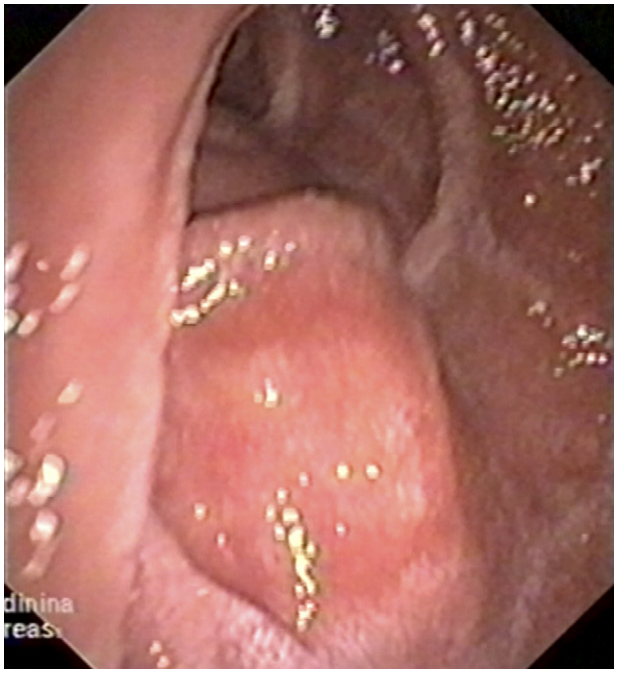

Fig. 2. Endoscopic image of the deformation of the duodenal vertical branch.

The data obtained made the diagnosis of cystic duodenal dystrophy highly suspected, which was confirmed by an endoscopic ultrasound (EUS) examination, which revealed the same size ovoid septated cystic structure located in the submucosa and muscularis propria against the background of infiltration and sclerosis of the second portion of the duodenal wall (Fig. 7). There were no signs of tumor growth in the duodenum, common bile duct, pancreas and peripancreatic lymph nodes.

The patient was successfully treated with pylorus preserving pancreatoduodenectomy (Fig. 8), and the diagnosis of a cystic form of duodenal dystrophy was confirmed (Fig. 9).

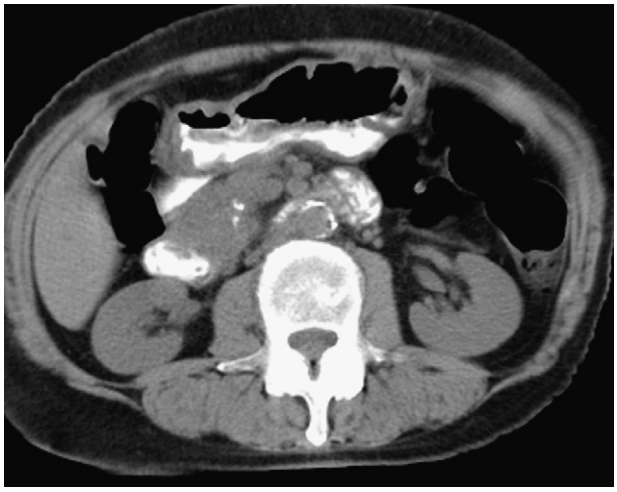

Fig. 3. Spiral CT. Multiple calculi in the uncinate process of pancreatic head.