Duodenal Duplication. Is Ultrasound Appearance Enough to Confirm the Diagnosis?

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Abstract

A case of duodenal duplication is reported. It is a rare anomaly and is commonly discovered during infancy. Symptoms of partial obstruction dominate the clinical picture. Abdominal ultrasound (US) seems to be the best method to confirm the diagnosis especially when the two-layer pattern is present. In our case the radiological evaluation apart from US comprised plain film of the abdomen, upper gastrointestinal series and CT scan. The diagnosis of duodenal duplication made by US examination as well as by gastrointestinal series and CT scan was confirmed surgically. We consider that when at US the sign of two-layer pattern is present there is no need for further radiological evaluation for alimentary tract duplication.

Key words

Duodenal duplication - imaging- congenital anomalies - gastrointestinal tract - ultrasound

Rezumat

Este prezentat un caz de duplicare duodenală. Aceasta este o anomalie rară, de obicei descoperită în copilărie. Simptomele de obstrucție parțială domină tabloul clinic. Ecografia abdominală (US) este cea mai bună metodă de confirmare a diagnosticului, îndeosebi atunci când este prezent aspectul de dublu-strat. In cazul nostru, evaluarea radiologică, în afară de ecografie, include o radiografie abdominală pe gol, bariu pasaj şi scan CT. Diagnosticul de duplicare duodenală care s-a stabilit prin ecografie, pasaj baritat şi CT a fost confirmat chirurgical. Considerăm că atunci când ecografia evidențiază semnul dublului strat, nu este nevoie de examinare radiologică suplimentară pentru a diagnostica duplicarea tractului digestiv.

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Introduction

Gastrointestinal tract duplications are rare congenital abnormalities that may occur anywhere along the alimentary tract from the mouth to the anus. Duodenal duplication cyst is a rare lesion representing 5-12% of all gastrointestinal tract duplications (1). Other sites include the distal ileum (35%), the distal esophagus (20%) and stomach (8%) (1). Because of their rarity duodenum duplications can represent a diagnostic challenge. Duodenal duplication is commonly found during infancy. Symptoms of partial obstruction dominate the clinical picture (2). The most common symptom is vomiting and the most positive sign is a palpable abdominal mass (3). The duplication consists of a smooth muscle wall, continued by the muscle layer of the intestinal wall and an inner mucosal lining. The ultrasound (US) appearance of these two layers as an echogenic inner lining (mucosa) and a surrounding hypoechoic rim (muscular wall) is useful to exclude other cystic masses lacking a mucosal lining (mesenteric or omental cyst, choledochal cyst etc). We report the case of an infant with duodenal duplication in whom abdominal US provided an accurate pre-operative diagnosis. In addition, we also discuss the surgical approach for this condition on the basis of the "two layer" sign.

Case report

A 3-day full term boy was referred to our department with projectible non-bilious vomiting. Laboratory findings were normal. On physical examination an "olive shaped" mass was palpated in the epigastrium. The possible diagnosis was hypertrophic pyloric stenosis. Because of his young age and the unclear clinical findings an abdominal US was requested. On sonography a well-defined cystic structure,

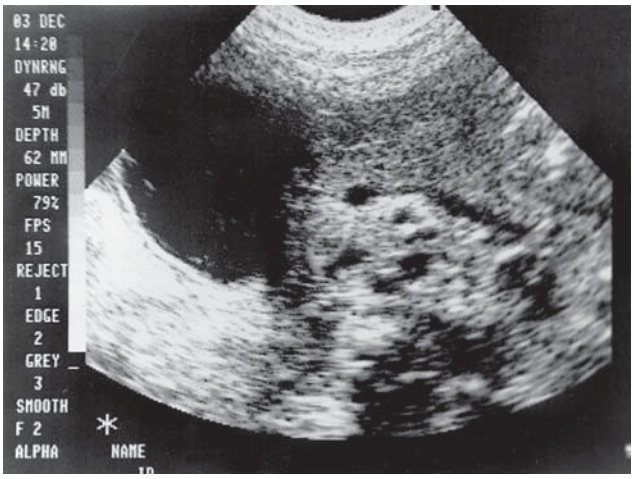

Fig.1 US evaluation: A well-defined transonic cystic structure adjacent to the head of the pancreas. The cyst has a thick wall consisting of two distinct layers (outer hypoechoic, inner echogenic).

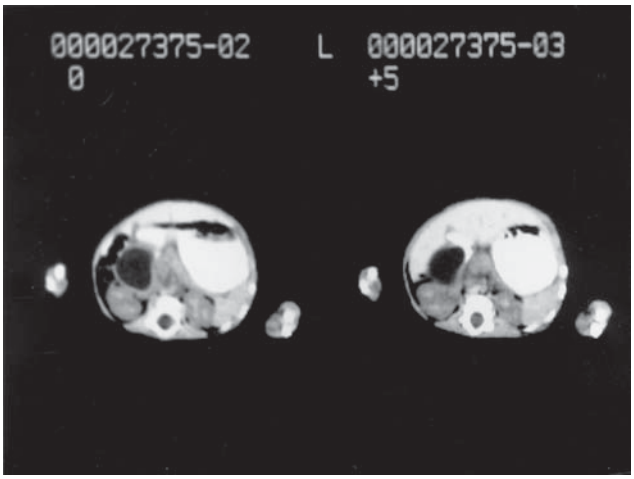

Fig.2 CT evaluation: A cystic mass with a thick wall in the same place as it was described at the US. No enhancement was demonstrated after administration of intravenous contrast material.

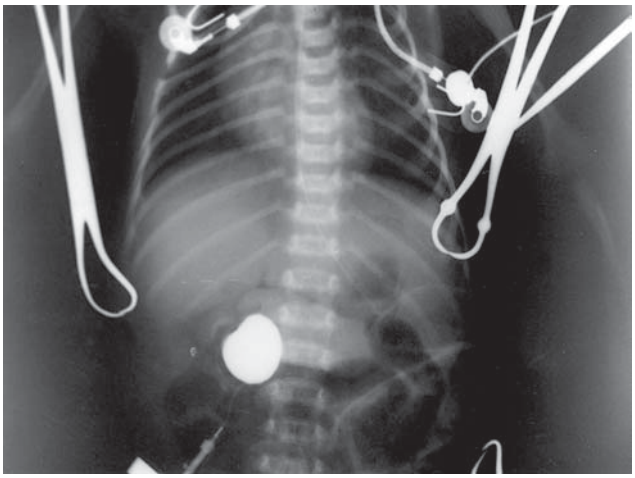

Fig.3 Contrast medium administration in the cyst during the operation. There was no communication of cyst with the duodenal lumen, biliary tree or any other organ.

transonic in the right upper abdomen, medial to the right lobe of the liver, adjacent to the gallbladder and to the head of the pancreas was revealed (Fig.1). The cyst had a thick wall consisting of two layers (outer hypoechoic, inner echogenic). Based on this two-layer appearance the diagnosis of duodenal duplication cyst was made. Because of the fact that a peristaltic activity within the wall was observed during the US examination, the diagnosis was confirmed.

A CT scan of the upper abdomen demonstrated a cystic mass with a thick wall in the same place as described at the US (Fig.2). The content of the mass had an attenuation of 25 HU and no enhancement was demonstrated after administration of intravenous contrast material.

Abdominal CT and especially US features were considered highly suggestive of a duodenal duplication cyst. The relation between the duplication of the duodenum and the adjacent organs was evaluated by abdominal US. The gallbladder and the biliary tree appearance was normal. The size and the echogenicity of the pancreas were normal. Liver, spleen, kidneys and adrenals were also normal.

During the operation a duodenal cyst was revealed. The lesion was catheterized, and after evacuation a contrast medium was administrated (Fig.3). There was no communication of cyst with the duodenal lumen, biliary tree or any other organ as was found at the US examination. The lesion was surgically removed.

Histologically, the duplication consisted of a smooth muscle cell wall and an epithelial layer.

Discussion

Duodenal duplication, which is an uncommon anomaly of the gastrointestinal tract, resulting from dual formation of the organ, is usually adjacent to the involved organ and may communicate with the normal lumen (4). However, it is usually noncommunicative and located along the first and second portions of the duodenum on the mesenteric side (5). Clinically, duodenal duplications manifest with symptoms of obstruction, but due to their location they may cause billiary obstruction and pancreatitis (6).

Abdominal radiography may show a soft-tissue mass within the abdomen. Barium enema examination usually shows a duplication mass extrinsic to the bowel lumen. Contrast material may gain access to such a communication with the lumen (12).

US reveals either a sonolucent mass with good through transmission due to the clear fluid content or an echogenic mass due to haemorrhage and inspissated material within the duplication. If the typical inner echogenic mucosal and outer hypoechoic muscle layers are seen, the diagnosis of duplication can be established (7). It has previously been noted that the wall of an enteric cyst, on US examination, is composed of an echogenic inner mucosal layer and a hypoechoic outer muscular layer (7-10). Also Barr et al considered that the double-layered cyst wall is a

safe sign for the diagnosis of the enteric duplication cyst (100% in a retrospectively review) (11).

In our case the diagnosis of duplication cyst was made on the basis of the "double-layered wall" (or "muscular rim sign") (12). The cyst was situated in the duodenum because of its topography and peristaltic activity within the wall during the US examination. The findings from the adjacent organs were normal. Also there was no difference in cyst appearance in the distension and emptying stages so the diagnosis of noncommunicating duodenal cyst was made.

A CT scan was made to exclude any other duplication sites. There was no more information from CT scan for the morphology of the lesion, for the presence or not of communication, or for the adjacent organs. CT can be used (when US findings are inconclusive) to diagnose the true nature, location, and extent of the lesion, as well as associated vertebral anomalies and possible other duplications (1) (non-existent in our case).

In this case abdominal US demonstrated a cystic mass with a double-layered wall. CT did not add any more information and because diagnosis of enteric duplication is difficult by MR imaging (13), no further evaluation was made and the patient underwent a surgery. Because of the lesion topography (near to pylorus and in anterior-lateral position) and macroscopic aspect (similar to hypertrophic pyloric stenosis) surgeons requested an abdomen plain film after the contrast material administration into the cyst. No communication with the gastrointestinal tract was demonstrated. The cyst was removed surgically without complications.

In conclusion, we believe (based on literature (14-16) and our experience) that when the "double-layered wall" or "muscular rim" sign is seen on ultrasound examination in an abdominal cystic mass, it is not only possibly but rather certainly diagnostic for an enteric duplication cyst.

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