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Hypoplastic pancreas and ectopic spleen as an abdominal mass: A case report

Hipoplastik pankreas ve karında kitle olarak ektopik dalak: Olgu sunumu

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We report a 35-year-old female patient who presented with pseudotumoral abdominal mass with final diagnosis of ectopic spleen and hypoplastic pancreas. Clinical diagnosis is difficult due to lack of symptoms. Laboratory findings are commonly non-specific; diagnosis can be confirmed by imaging studies. This patient complained only of abdominal painless mass. The laboratory findings were all within normal limits. Diagnostic images revealed ectopic spleen and absence of the dorsal pancreas. This interesting and rare combination has not been reported previously in the literature

Key words: Ectopic spleen, hypoplastic pancreas

Bu olgu sunumunda; karında kitle ile başvuran, ektopik dalak ve hipoplastik pancreas tanısı alan 35 yaşında bir kadın hasta sunulmuştur. Tablonun klinik tanısı semptom olmaması nedeniyle zordur. Laboratuvar sonuçları sıklıkla nonspesifiktir. Görüntüleme yöntemleri ile tanı konabilir. Hastamız karında ağrısız kitle ile başvurmuştur. Tüm laboratuvar değerleri normal sınırlardadır. Görüntüleme yöntemi ile ektopik dalak ve dorsal pancreas yokluğu saptanmıştır. Bu ilginç kombinasyon nadir olup literatürde henüz hiç yayınlanmamıştır.

Anahtar kelimeler: Ektopik dalak, hipoplastik pankreas

INTRODUCTION

The spleen is normally situated in the posterior part of the left upper quadrant of the abdomen with fixation by the splenic peritoneal attachments (gastrosplenic, splenorenal and phrenicocolic ligaments). The spleen is sometimes not confined to its normal location due to congenital deficiency, acquired laxity of these structures or embryologically incomplete fusion of the dorsal mesogastrium, becoming essentially a totally intraperitoneal hypermobile organ (1). Ectopic or wandering spleen is uncommon in clinical practice, usually asymptomatic and generally unsuspected in diagnosis (2). The incidence, based on several large series of splenectomies, is less than 0.5%. It usually occurs at 20 to 40 years of age, and most cases are seen in women (3). Hypoplastic pancreas (partial agenesis of pancreas) is another rare congenital abnormality consisting the parenchyma and ductal system restricted to the head with some residual dorsal tapering (4). This anomaly may be incidental or associated with episodes of recurrent pancreatitis (5). In the literature, the hypoplastic pancreas is frequently characterized by the presence of multiple spleens (polysplenia) and a variable combination of thoracic and visceral anomalies (6).

We report here a patient with ectopic spleen presented with abdominal mass and hypoplastic pancreas. The appearance of these two rare anomalies in the same patient is quite unusual and to our knowledge there is no similar case in the literature.

CASE REPORT

A 35-year-old female patient was admitted to our hospital after recently noticing a palpable abdominal mass located in the right paraumbilical region. She had a history of irritable bowel syndrome, but was otherwise healthy and had experienced no symptoms. She had three children, with no reported complications during pregnancy or childbirth. Her menses were also regular.

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The patient was afebrile with pulse rate of 84 bpm, respiratory rate of 16/min., and blood pressure of 110/70 mmHg. The appearance of umbilicus and abdominal wall were normal. Abdominal examination revealed palpable mass in the right paraumbilical region. The deep palpation detected a 15 cm in diameter, round, mobile, firm, painless and discrete mass with smooth margins and surface. On percussion, dullness of only this area confirmed the presence of the solid structure. The bowel sounds were normal. There was no evidence of hepatomegaly or ascites.

All blood and urine investigations were in normal limits. Ultrasonography (US) showed the absence of a normally positioned spleen, and presence of an enlarged spleen in an ectopic placement as a homogeneous, 18x7x5cm in diameter, isoechoic mass, consistent with spleen. The abdominal computerized tomography (CT- Phillips Tomoscan AV, slice thickness: 10 mm) demonstrated a remarkably enlarged spleen located in the right side of the abdomen and the anterior of the intestine. It displaced from the lower pole of the left kidney downwards and right laterally, towards the bifurcation of the vena cava (Figure 1). CT scan also demonstrated hypoplastic pancreas. Pancreatic tissue in the body and the tail of pancreas was absent (Figure 2). The remainder of the pancreas was normal. According to these clinical and radiologic findings, the patient was diagnosed as hypoplastic pancreas and enlarged ectopic spleen. At three-month follow-up the patient was asymptomatic.

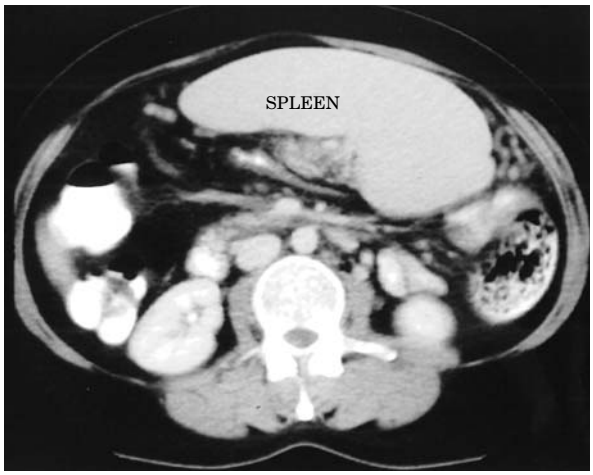

Figure 1. CT scan demonstrating ectopic spleen

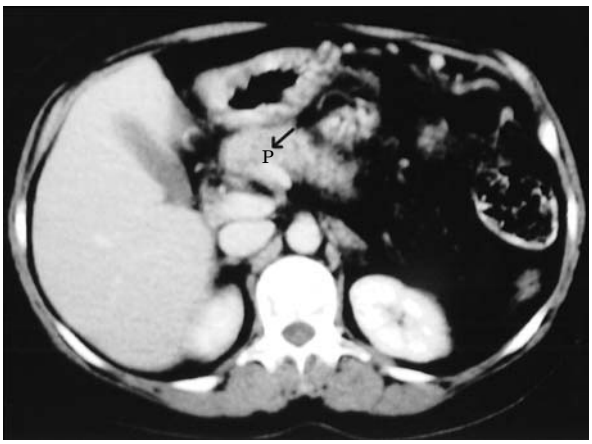

Figure 2. CT scan demonstrating short pancreas

DISCUSSION

Fewer than 500 ectopic spleen cases have been reported in the literature (1). The mechanism of ectopic spleen can be explained by two possible etiologies, acquired and congenital. The acquired form occurs in multiparous women as a result of hormonal changes during pregnancy. This causes a slackening of the abdominal wall and laxity of the ligaments normally attached to the spleen. There have been suggestions that the flaccidity due to pregnancy or connective tissue disease of the abdominal ligaments may also play a role in the development of ectopic spleen, which may explain the higher incidence in women of childbearing age (3). In this case, the patient had been well until this presentation, without clinical complaint or abnormal radiologic images.

The splenic pedicle is formed by the splenic ligaments and contains the splenic artery and vein and the tail of the pancreas. The spleen is connected to the dorsal body wall in the region of the left kidney by the spleno-renal ligament and to the stomach by the gastro-splenic ligament. If there is incomplete fusion of the dorsal mesogastrium, the spleen may remain on a long pedicle and lie in an ectopic intra-abdominal localization. Ectopic spleen localizations may vary, including adnexal, testicular renal or retroperitoneal regions. The weight of a normal adult spleen is between 75-150 g. Most ectopic spleens are enlarged. Splenomegaly is often secondary to twisting or compression of the pedicle and the resultant congestion (3).

Clinical presentation of an ectopic spleen could be acute or chronic. In an extensive review of 133

cases by Buehner and Baker, 76 patients presented with a mass and non-specific abdominal symptoms, 26 patients were asymptomatic, 25 patients presented with acute abdominal pain, and another six cases had an asymptomatic mass (7). The ectopic spleen with elongated pedicle has predisposition to both acute and chronic torsion with possible infarction. Torsion of the spleen with infarction can lead to the development of an "acute abdomen" (8).

The clinical diagnosis is usually easy because non-invasive imaging procedures such as US, nuclear scintigraphy, CT scan and magnetic resonance imaging are usually diagnostic. US may show a solid mobile mass and absence of the spleen in the left upper quadrant. CT of the abdomen is perhaps most often suggestive of the diagnosis. It is the easiest and safest means to diagnose wandering spleen (9). Hematological and biochemical tests are not helpful for diagnosis since they are almost always normal.

Recommendations in the literature for the management of ectopic spleen are varied. The significant risk of post-splenectomy sepsis implicates a conservative approach especially in asymptomatic patients. In the absence of infarction, thrombosis or hypersplenism, in patients presenting with an acute abdomen, detorsion with splenopexy is considered to be the optimal treatment (10, 11). Splenectomy should only be recommended when there is no evidence of splenic blood flow after detorsion of the spleen (12).

Pancreatic anomalies are occasionally reported, but complete agenesis of the dorsal pancreas is extremely rare (5, 6). In ectopic spleen cases, a long splenic pedicle can cause malposition in the pancreas. Agenesis of the dorsal pancreas can be an incidental finding or present with complications such as pancreatitis and diabetes mellitus. Short pancreas is closely related to polysplenia, interruption of the inferior vena cava, azygos or hemiazygos continuation, left-sided vena cava, symmetrical liver, anomalous liver fissure, anomalous liver lobe, median location of the gallbladder, inverted stomach, gallbladder and pancreas, and intestinal malrotation (13-15).

In conclusion, our case had both ectopic spleen and a hypoplastic pancreas without any symptoms. This is a rare case in the literature.

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