

Congenital agenesis of the right lobe of the liver in situs ambiguous revealed by a colonic obstruction due to a wandering spleen torsion: report of a case

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Citation

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Abstract

Congenital agenesis of the right lobe of the liver in situs ambiguous is rare. This abnormality revealed by a colonic obstruction due to a wandering spleen torsion has never been described. We report the case of a young man admitted for acute colonic obstruction associated with a painful hypogastric mass. Plain abdominal X-ray showed an enormous colonic dilatation with a soft-tissue density imaging in the lower middle quadrant. Laparotomy found a colonic obstruction due to pedicle torsion of a wandering spleen around the sigmoid. The liver was located at the left hypochondrium and presented a congenital agenesis of the right lobe with a retrohepatic gallbladder. The patient underwent splenectomy and sigmoidectomy. Liver and gallbladder were respected. The post-operative course was uneventful. This case presentation highlights the necessity to diagnose liver and other organ abnormalities in situs ambiguous earlier in order to prevent acute complications and to survey hepato-biliary patterns.

INTRODUCTION

Situs ambiguous or heterotaxia is a very rare situation characterized by an abnormal arrangement of organs and vessels without any mirror-image location of the viscera, relative to situs solitus (normal position of the heart and abdominal viscera) [1]. It is commonly asymptomatic and associated with abnormalities of location and morphology of liver and spleen. However, congenital agenesis of the right lobe of a left-sided liver associated with a wandering spleen revealed by an acute abdomen has never been described in situs ambiguous. The objective of this case presentation is to report an exceptional case of congenital agenesis of the right lobe of the liver in situs ambiguous revealed by a colonic obstruction induced by a pedicle torsion of a wandering spleen.

OBSERVATION

A 21-year-old man was referred to our emergency department because of diffuse and continuous abdominal pain, vomiting and constipation related to a 48-hour bowel obstruction. His past medical history was not significant except presence of a very mobile and indolent mass in the lower part of his abdomen. The patient had no history of previous abdominal surgery or trauma. On physical

examination, he did not present jaundice or fever. The peripheral arterial pulses and blood pressure were normal and all vital signs were stable. The patient's abdomen was much distended; moderate tenderness and diffuse tympanic percussion were noted. Hypogastric palpation found a large and firm painful mass. The rest of the physical examination was normal. There was a moderate thrombocytopenia while other laboratory data were normal. Plain abdominal X-ray showed evidence of colon obstruction, with enormous bowel dilatation which reached the diaphragm and two air-fluid levels in left and right flanks (figure 1). A normal liver silhouette was missing in the right upper quadrant. A spleen-like mass silhouette was seen as a soft-tissue density imaging in the lower middle quadrant.

Figure 1

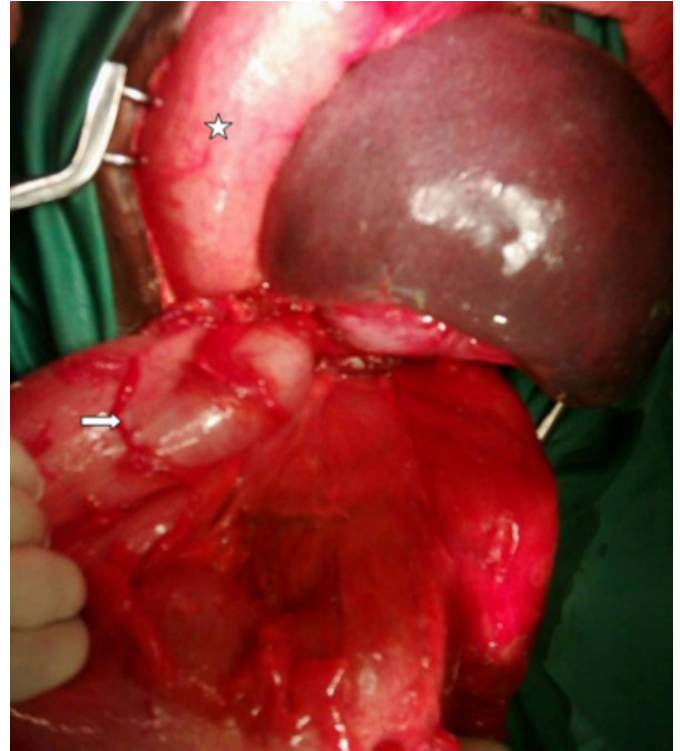
Figure 1: Plain abdominal X-ray. Bowel dilatation reaches the diaphragm (arrow) with air-fluid levels. Absence of liver silhouette in the right upper quadrant. A spleen-like mass silhouette is seen in the lower middle quadrant (star), between the two iliac crests.



With the diagnosis of acute colonic obstruction, the patient underwent an exploratory laparotomy through a middle incision without any other radiographic exam. The operative finding was torsion of an enlarged hypogastric wandering spleen around a very long sigmoid loop which caused a complete obstruction with dilatation of the proximal part of the entire large bowel (figure 2). The enlarged spleen caused a moderate and congestive splenomegaly without any ligamentous attachments. Colon and spleen were viable and well-colored.

Figure 2

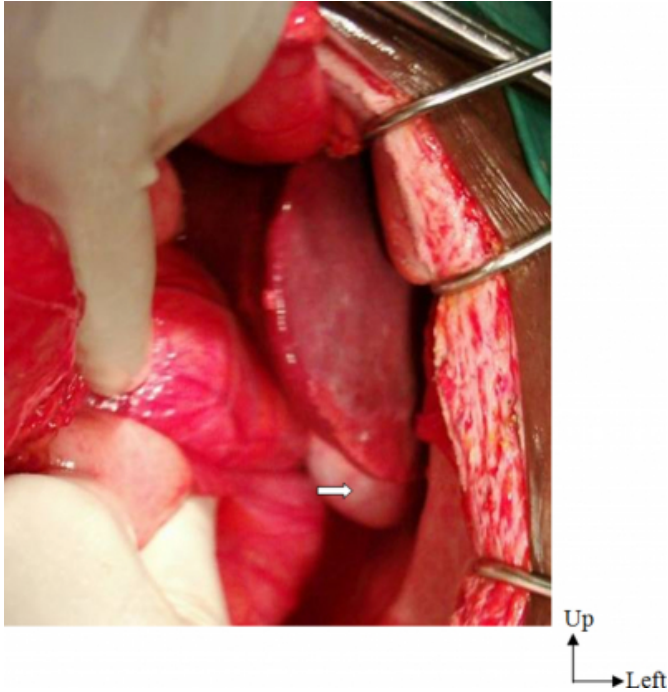
Figure 2: Operative findings. Torsion of the pedicle of a wandering enlarged spleen around the sigmoid (arrow). Dilatation of the proximal part of the colon (star). The spleen has not any ligamentous attachments.



There was a situs ambiguous with the liver in the left hypochondrium. The liver presented a complete agenesis of the right lobe with a compensatory hypertrophy of the left lobe (figure 3). The gallbladder had a normal appearance and was retrohepatic on the lateral segment of the liver, at its extremity. There was not any gallstone. The liver parenchyma presented a homogeneous and normal aspect without any macroscopic sign of fibrosis. There was not any sign of portal hypertension. Except the liver, gallbladder, wandering spleen and the very long sigmoid, all other abdominal organs were normal and located at their usual place.

Figure 3

Figure 3: Situs ambiguous with liver in the left hypochondrium. The liver presents a complete agenesis of the right lobe. The gallbladder (arrow) is retrohepatic, in the extremity of the lateral segment of the liver.



Following detorsion, dimensions and aspects of the splenomegaly were not modified. In view of persistent splenomegaly after detorsion, pre-operative thrombocytopenia and the risk of recurrent spleen torsion after splenectomy, a total splenectomy was performed combined with a sigmoidectomy and Bouilly-Volkman end-colostomy. Liver and gallbladder were respected. The patient received antibiotics, Haemophilus influenzae and pneumococcal vaccines after surgery. The post-operative recovery period was uncomplicated and colonic continuity re-establishment was realized 3 weeks later. Histopathologic examination of the spleen revealed malaria-induced splenomegaly with congestive and fibrosis injuries without any malignancy. A chest X-ray and a computed tomography of the abdomen were performed to study vascular and organ arrangement. Computed tomography confirmed situs ambiguous with left-sided liver in the left upper quadrant and agenesis of the right liver lobe (figure 4). The left lobe was much enlarged related to a compensatory hypertrophy. The caudate lobe was normal. There was an absence of right hepatic artery, right branch of portal vein and right hepatic vein. There was a portal vein which continued on an only well-developed left branch which irrigated the lateral and medial segments. The colon was in the right upper quadrant.

The patient was discharged one week after colonic continuity re-establishment. At the time of writing, the patient remains well, 3 years later.

Figure 4

Figure 4: Postoperative computed tomography. Situs ambiguous with left-sided liver and agenesis of the right lobe of the liver. Compensatory hypertrophy. Well-developed left portal vein which divides in lateral branch (long black arrow) and medial branch (short black arrow). The medial branch gives an anterior branch (long white arrow) and a posterior branch (short white arrow). Hepatic flexure of colon in the right upper quadrant.



DISCUSSION

In absence of associated congenital heart diseases or bowel obstruction due to malrotation, situs ambiguous and agenesis of the right liver are usually asymptomatic and are often discovered incidentally or during unrelated abdominal emergencies [1, 2]. In this indexed case, the circumstance of diagnosis was a double complication due to bowel obstruction induced by pedicle torsion of a wandering spleen. To our knowledge, this is the first case of congenital agenesis of right lobe of the liver discovered in situs ambiguous in an adult, revealed by an acute torsion of a wandering spleen which realizes a colon obstruction; large-bowel obstruction or gastric volvulus due to wandering spleen had been described in situs solitus [3, 4]. Congenital agenesis of right lobe of the liver, which has been described first by Heller in 1870, is a very rare malformation with only 45 cases reported in the literature; the anomaly affecting more often the left than the right hepatic lobe [5, 6, 7]. In the literature, hepatic abnormalities usually described in situs

ambiguous are: left-sided whole liver, middle-located whole liver and symmetric liver (isomeric liver) [1, 8]. The pathogenesis of congenital agenesis of right lobe of the liver is not fully explained but it is thought to be an impaired mutual induction between the endodermal diverticulum and the diaphragm or abnormal development and thrombosis of the right portal vein during 2-3th week of embryologic growth [5, 9]. It must be distinguished from severe atrophy of right hepatic lobe in cirrhosis, intrahepatic cholangiocarcinoma and compensatory hypertrophy of left hepatic lobe after a prior surgical resection of the right hepatic lobe. At computed tomography, morphologic aspects of congenital agenesis of the right lobe of the liver are absence of the right lobe, a retrohepatic gallbladder, compensatory hypertrophy of the unique left lobe and posterolateral interposition of the hepatic flexure of the colon; the best signs are the absence of right portal branch and right hepatic vein [10]. This anomaly can be associated with biliary tract diseases like gallstones, congenital bile duct cyst and portal hypertension [7, 11, 12,]. These complications did not exist in this indexed case and the mostly compensatory hypertrophy of the left hepatic lobe with good blood outflow can explain the absence of portal hypertension. Delayed diagnosis of situs ambiguous anomalies can lead to life-threatening complications like wandering spleen torsion, infarction and necrosis. As congenital agenesis of the right lobe of the liver, the pathogenesis of wandering spleen is related to an anomaly of embryologic growth. During fetal development, the dorsal mesogastrium fails to fuse with the posterior peritoneum leading to an absence of formation of spleen-supporting ligaments [13]. Congenital agenesis of the right lobe of the liver in situs ambiguous must be diagnosed early and surveyed to avoid long-term complications like bile duct calculi or cholangiocarcinoma due to poor bile drainage and compression of gallbladder [7, 14]. For these reasons, some authors [7, 14, 15] propose a preventive cholecystectomy. Our indexed case did not undergo cholecystectomy and with a follow-up of three years, we did not find any biliary disease. However, a splenectomy was performed because of the risk of recurrent torsion of a malaria splenomegaly with thrombocytopenia.

CONCLUSION

This case presentation highlights the necessity of early

diagnosis of situs ambiguous abnormalities; herein, congenital agenesis of the right lobe of the liver associated with a wandering spleen. Early diagnosis permits to survey the hepato-biliary pattern including malignancy and to prevent life-threatening complications like wandering spleen torsion and intestine necrosis by splenopexy. Preventive cholecystectomy still remains a matter of debate. This is the first case of congenital agenesis of the right lobe of the liver in situs ambiguous revealed by a wandering spleen torsion.

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