

Twist of Fate: A Rare Case of Congenital In-situ Hepatic Malrotation with Diaphragmatic Eventration

Case Report

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Abstract

Hepatic malrotation is an uncommon developmental anomaly in which the liver and its associated structures occupy an atypical orientation. We present a 46-year-old man with a rare combination of hepatic malrotation and diaphragmatic eventration, identified incidentally during evaluation for chronic abdominal pain. This case adds to the limited published literature, highlighting the challenges in detection and the implications for radiological interpretation and surgical planning. Recognition of this anomaly in adults is particularly important, as it is often overlooked or misinterpreted.

Keywords: Hepatic Malrotation; Diaphragmatic Eventration; Congenital Anomaly

Introduction

Congenital variations of normal liver anatomy are rare. Liver anomalies can be broadly classified as congenital anomalies or acquired morphological changes. They may also be categorized based on the system involved into anatomic hepatic anomalies, hereditary anomalies, and vascular or biliary system anomalies [1]. Anatomical anomalies—such as situs abnormalities or atypical lobation, including accessory lobes—are most often incidental findings and may be identified in pediatric or adult patients during imaging studies, surgical procedures, or autopsies. One was in a patient with antiphospholipid antibody syndrome as an incidental post-mortem finding by Zhong, [2] and the other in an adult with congenital diaphragmatic hernia [3,4]. We report a rare case of hepatic Malrotation with diaphragmatic eventration in an adult, emphasizing its radiological findings and clinical relevance.

Case Report

A 46-year-old male with a history of HBsAg positivity and pan-gastritis presented to the medicine department with complaints of chronic abdominal pain, not associated with vomiting or loose stools. The patient reported self-medicating with tramadol daily for the past 6 months and had recently increased the dose due to aggravated pain over the last week. On examination, the patient was hemodynamically stable with mild diffuse abdominal tenderness. Laboratory investigations were normal. An ultrasound scan performed elsewhere had reported bilateral renal calculi. A contrast-enhanced CT (CECT) of the abdomen was advised for further evaluation. The CT study demonstrated a malrotated liver with the hepatic hilum oriented anterolaterally and the gallbladder located in an atypical anterior position [Figure 1]. There was moderate to marked elevation of the posterior aspect of the right hemidiaphragm along with upward

displacement of the right hepatic lobe, suggesting diaphragmatic eventration [Figure 2]. Additionally, a right-sided partial duplex collecting system with microliths in the mid-pole of the right kidney was demonstrated. Diffuse fat deposition was noted around the lower esophagus, pericardium, and visualized mediastinum, indicative of mediastinal lipomatosis an incidental finding. No evidence of other associated findings like dextrocardia, polysplenia or asplenia, intestinal malrotation and heterotaxy syndrome was identified. The patient was managed conservatively with antispasmodics and proton pump inhibitors for symptomatic relief, and a psychiatric opinion

was sorted for tramadol addiction but subsequently patient was lost to follow-up.

Discussion

Hepatic malrotation is a very uncommon congenital anomaly in which the liver fails to assume its expected orientation during embryonic development. Anatomical anomaly in the adult liver is a very rare occurrence. Most of them usually involve a part of the liver, a lobe, or segments that present with agenesis or fusion [2].

During the mid-third to early fourth week of embryonic development, the hepatic diverticulum emerges from the distal foregut as an endodermal outgrowth. After penetrating the septum transversum, the liver bud divides into right and left components, which subsequently give rise to the individual lobes. The pars cystica—destined to form the gallbladder and cystic duct—arises as a ventral outpouching from the developing bile ducts. Initially, the bile duct opens ventrally into the developing duodenum; following gut rotation, it assumes a dorsal opening. Anatomical variations of the liver are rare and are usually detected incidentally, with most exerting no significant physiological or functional impact. Among these, lobar or segmental agenesis and fusion represent the predominant forms [3].

Recognizing various anatomical abnormalities of the liver will benefit clinical practice, especially valuable for radiologists and surgeons [2]. Advanced diagnostic imaging modalities play a major role in medical practice. Especially, understanding the normal liver anatomy [1,5] or variants for treating interventionists, surgeons, and interpreting radiologists [1]. This report documents one of the very few adult cases of hepatic malrotation described in the literature, highlighting the importance of recognizing such anomalies to ensure accurate diagnosis and safe clinical management.

Conclusion

This case highlights the critical importance of recognizing rare anatomical variations, such as hepatic malrotation, in clinical practice. Comprehensive preoperative imaging plays a pivotal role in ensuring accurate diagnosis, guiding surgical planning, and preventing intraoperative complications. Awareness of such anomalies is essential for radiologists and surgeons alike to optimize patient care and outcomes.

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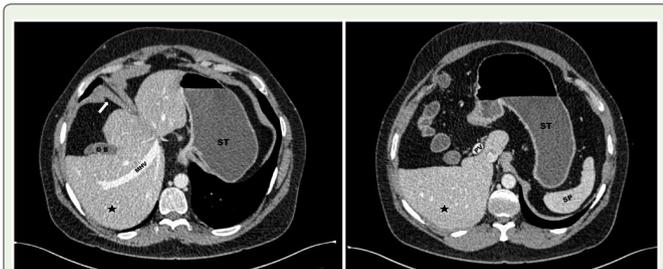


Figure 1: Axial CECT abdomen demonstrates a malrotated liver with the hepatic hilum directed antero-laterally and the gallbladder oriented anteriorly. A thickened anterior diaphragmatic slip is noted (white arrow). The stomach and spleen are in their normal positions. GB – Gallbladder; MHV – Middle hepatic vein; ST – Stomach; SP – Spleen; ★ – Right lobe of liver.



Figure 2: Coronal and sagittal sections demonstrating marked elevation of the posterior right hemidiaphragm (white arrow). The anterior right hemidiaphragm appears thickened, while the posterior dome is markedly thinned, with upward displacement of the right lobe of liver (★), consistent with diaphragmatic eventration.

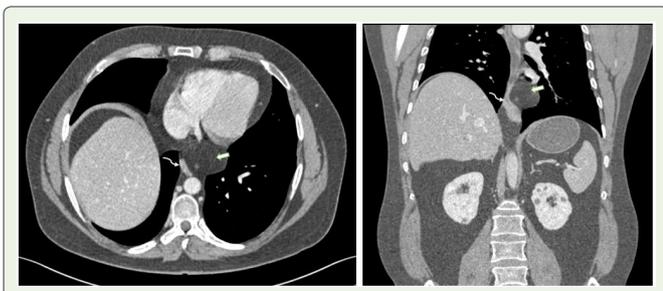


Figure 3: Axial and coronal sections of the abdomen demonstrate diffuse fat deposition surrounding the lower esophagus, pericardium, and visualized mediastinum, consistent with mediastinal lipomatosis, noted as an incidental finding. The curved arrow indicates the esophagus, while the arrowhead denotes the fat deposition.