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Hepato-Colonic Vagrancy in Woman with Polycystic Kidney Disease and Suspected Hydatid Disease

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Abstract

Background: The liver is commonly positioned in the upper right abdominal quadrant. However, hepato-colonic vagrancy refers to a rare anatomical anomaly in which liver and colon are abnormally positioned or exhibit excessive mobility, potentially indicating an embryological basis for displacement.

Case Description: A 70-year-old female experienced persistent dyspeptic symptoms, prompting a Contrast Enhanced abdominal CT scan, which revealed an unusual inversion of the liver around its oblique transverse axis. In addition to these findings, the scan showed a mobile caecum, hepatic flexure and ascending colon, resulting in displacement of the liver from its anatomically correct site within the right upper abdominal quadrant. Incidentally, multiple renal and splenic cysts were identified, consistent with Autosomal Dominant Polycystic Kidney Disease (ADPKD). A large hepatic cyst with calcifications was also observed, raising the concerns of hydatid disease, for which hydatid serology was recommended.

Conclusion: Hepatic vagrancy, displacement or malposition is a rare condition, predominantly reported in young adult males. However, this report presents a rare occurrence of concurrent hepatocolonic malposition in an elderly female, with multiple renal and splenic cysts, and a potential hydatid cyst in liver. The findings highlight the importance of recognizing both typical and atypical anatomical variants to ensure accurate diagnostic assessments and guide effective clinical management.

Keywords: Hepato-colonic vagrancy; Liver malrotation; Autosomal dominant polycystic kidney disease (ADPKD); Hydatid disease

Abbreviations: CT: Computed Tomography; CBD: Common Bile Duct; HU: Hounsfield Unit; ADPCKD: Autosomal Dominant Polycystic Kidney Disease; NTU: NovaTec Unit

Introduction

The liver is commonly positioned in the right upper abdominal quadrant, held in place by several anatomical structures, including the coronary, triangular, and falciform ligaments, as well as the connective tissue within the bare area and the inferior vena cava [1]. Hepatic vagrancy, however, refers to the hypermobility of the liver, where it can move transversely or shift from the right abdominal region to the left. This condition is distinct from situs inversus and may or may not be associated with colonic vagrancy, suggesting a possible embryological or developmental link [2].

Hepatic vagrancy is a rare condition, often described using various terms such as wandering liver, floating liver, hepatic torsion, hepatoptosis, hepatic ectopia, and hepar ambulens [3]. The hypermobility of the liver can result from the absence or laxity of the normal hepatic suspensory ligaments, lack of tethering to the inferior vena cava or persistence of the ventral mesentery [4]. This condition can lead to the liver occupying abnormal positions within the abdomen, which can be identified through imaging techniques such as multidetector computed tomography (CT) with multiplanar reformations [5].

This case study documents a fascinating case of hepato-colonic vagrancy, identified in a 70-year-old woman undergoing contrast-enhanced Computed Tomography (CT) of the abdomen due to persistent dyspeptic symptoms.

Case Presentation

A 70-years old female was directed for a Contrast Enhanced CT scan of the abdomen due to dyspepsia and shortness of the breath. She had been diagnosed with Type 2 diabetes a few months earlier and was being managed with oral hypoglycemics. There was no history of abdominal surgery or trauma. On examination, a palpable midabdominal mass was noted. Her laboratory tests were unremarkable. An abdominal Computed Tomography (CT) scan was performed using a liver protocol which included pre-contrast, arterial, portovenous and delayed phases. The scan revealed an abnormal position of the liver; the bulk of liver was located in the mid abdomen, and the porta hepatis was oriented posterosuperiorly to the right side (Figure 1A,1B, 2 & 3).

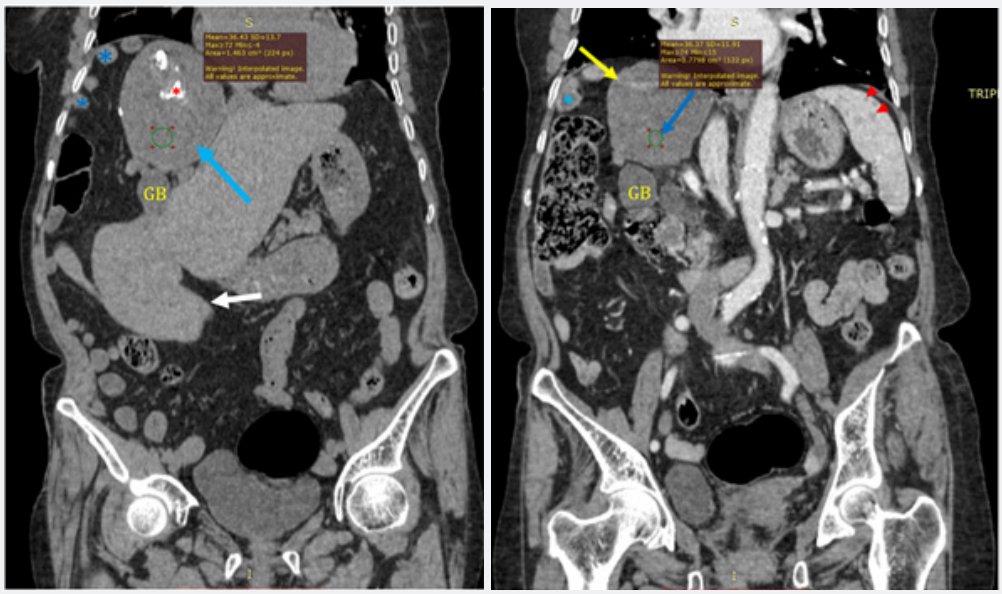

Figure 1A: Coronal non contrast enhanced CT Abdomen & Pelvis. The liver is inverted in the abdomen (white arrow). Ileum is seen in right upper quadrant in the subphrenic space (blue asterisks). Hepatic Cyst=blue arrow, with calcifications (green asterisk), fluid contents are 34HU. Region of Interest (ROI) is placed in cyst contents, showing density of 34HU. GB=Gall Bladder
Figure 1B: Coronal contrast enhanced CT Abdomen & Pelvis, showing enhancement of thin wall of cyst (yellow arrow); contents remain unenhanced (blue arrow) showing ROI. GB=Gall Bladder; ileum=blue asterisks; splenic cysts=red arrowhead.

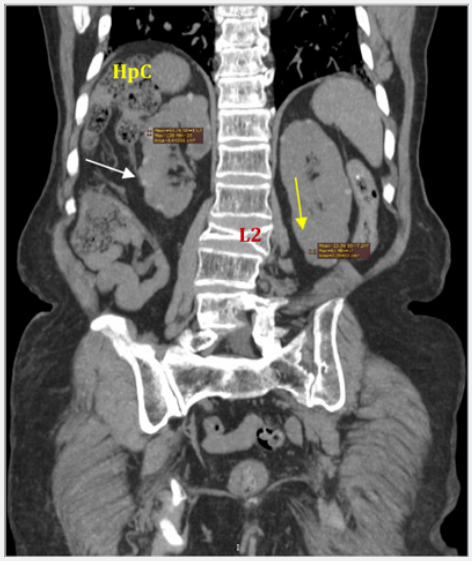

Figure 2: Coronal non-enhanced CT Abdomen & Pelvis (MIP images), showing multiple cysts in bilateral kidneys, some of which are hyperdense.
Hypodense renal cysts=yellow arrow; hyperdense renal cysts=white arrow; HpC=Hepatic flexure of colon is in right subphrenic region. Osteoporotic fracture of L2 vertebra=L2. ROIs are placed within both hypodense and hyperdense renal cysts.

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Delineation of segmental anatomy by hepatic and portal vein branches revealed an apparent inversion or rotation of the liver around the oblique transverse axis through the porta hepatis, reaching the mid abdomen at the level of the superior end plate of L5, with loss of the bare area of liver (Figure 1A, 1B, 3 & 4). Portal vein and common bile duct (CBD) were stretched due to this malrotation; however, normal contrast opacification of the portal vein was observed. CBD exhibited focal dilatation in its midpart, measuring upto 12mm, which was attributed to the stretching of the porta hepatis. No intra-hepatic biliary dilatation was noted (Figure 3 & 4).

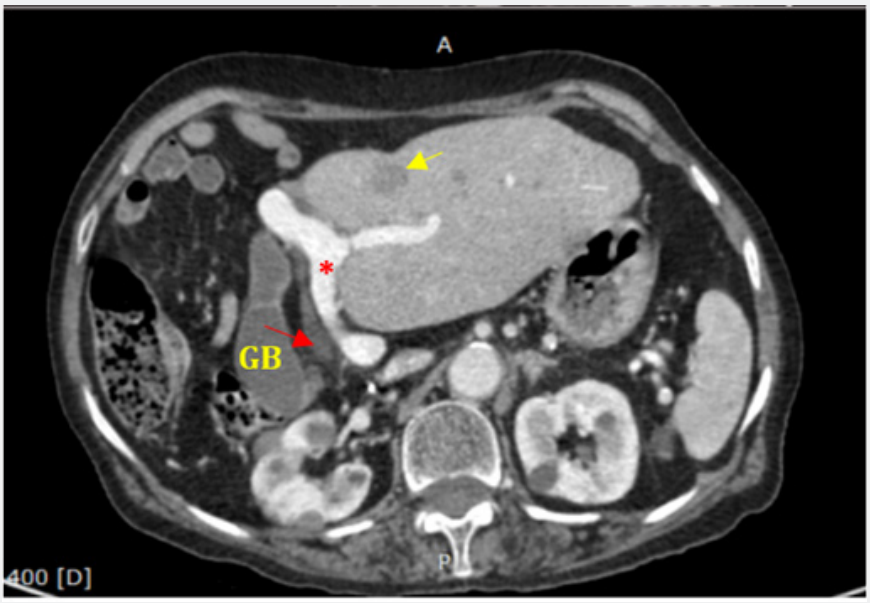

Figure 3: Axial Contrast enhanced CT Abdomen & Pelvis demonstrating stretching of CBD and portal vein. Gall Bladder (GB) is hanging freely posteroinferiorly. Common bile duct (red short arrow) and Portal Vein (red asterisk) are stretched. Right hepatic vein, not yet filled with contrast=yellow arrow.
Bilateral renal cysts are re-observed; enhancement of renal parenchyma while the cysts remain unenhanced.

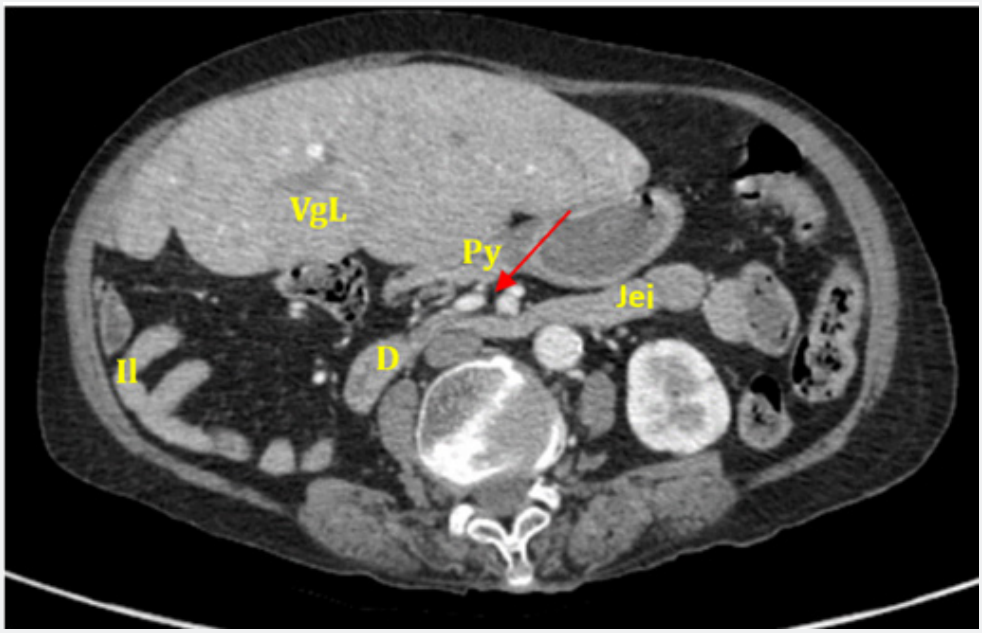

Figure 4: Axial contrast enhanced CT Abdomen & Pelvis, showing pylorus of stomach (Py) compressed between vagrant liver (VagL) and superior mesenteric vessels (red arrow).
Loops of ileum in right upper quadrant=Ile; Du=Duodenum; Jej=Jejunum.

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There was displacement of the small and large gut loops from their normal anatomical positions; however, no malrotation, torsion or volvulus was observed. The superior mesenteric vessels maintained their normal orientation, the superior mesenteric artery to the left of superior mesenteric vein. Ileal loops were seen in the right upper quadrant (Figure 1B, 3 & 4). The ileocaecal junction and caecum had migrated superiorly and were visualized posteromedial to the vagrant liver. The hepatic colon flexure was located in the right subphrenic area, while the transverse colon was pushed inferiorly by the wandering liver. These findings suggested a mobile caecum, ascending colon and hepatic flexure of the colon. Based on these observations, a working diagnosis of hepato-colonic vagrancy was made.

The gall bladder was hanging freely in the right upper quadrant, (Figure 3), directed posteriorly from the porta hepatis, and abutting it along its medial aspect. The stomach was partially distended with fluid, with limited space for the body of the stomach, pylorus and gastroduodenal junction between the wandering liver and the superior mesenteric vessels (Figure 4). The gastro-esophageal and gastro-duodenal junctions, as well as the duodenum, were in their normal orthoptic position. The pancreas, spleen, splenic colon flexure, descending and sigmoid colon, rectum and anal canal were also in their normal anatomical locations and positions. In addition to these findings, a cyst was observed in the right upper quadrant, inseparable from the inverted visceral surface of the liver, measuring 10 x 8 x 7.7cm (Anteroposterior x Transverse x Craniocaudal dimensions). The cyst had a thin capsule, a few thin septae, and multiple coarse calcific foci within it. It contained fluid contents with mean Hounsfield Unit (HU) of 10; delayed enhancement of the cyst wall and thin septations was noted (Figure 1A, 1B & 4).

The kidneys were located in their normal orthoptic positions in retroperitoneum. Multiple cysts were observed in both kidneys, ranging in size from 3mm to 18 mm. The cysts were mostly unilocular and contained low density fluid (10-15HU). A few cysts showed high density non-enhancing fluid contents (50-90HU), suggesting possibility of hemorrhages. Additionally, hypodense, tiny, non-enhancing cysts were seen in the spleen (Figure 1B). These findings supported the diagnosis of co-existent Autosomal Dominant Polycystic Kidney Disease (Figure 2 & 3). An osteoporotic compression fracture of the L2 vertebral body was noted (Figure 2), along with spondylolisthesis at L4-L5, which resulted in development of kyphoscoliosis. Atherosclerotic calcific changes were observed in abdominal aorta and its branches. The large cyst containing calcific foci in the right upper quadrant appeared to be hepatic in origin and didn't seem to be an extension of Autosomal Dominant Polycystic Kidney Disease. Echinococcus was suggested as a possible etiological cause of the hepatic cyst, and further evaluation with hydatid serology was advised.

The patient was offered surgical treatment for hepato-colonic vagrancy and the hepatic (hydatid) cyst, with the potential to alleviate her dyspepsia. However, the patient declined the surgery and was instead advised to make lifestyle and dietary modifications to help manage her symptoms. She was also educated on the warning signs of gut ischemia. Echinococcus IgG serology quantitate levels came out to be 9.4 NTU, which is equiovocal for Hydatid Disease [5]. Albendazole 400 milligrams (mg) twice daily was prescribed for one month as medical treatment for the hepatic cyst. At the 2- and 6-month follow-ups, the patient remained stable, without any complications. No further imaging was performed on patient's request.

Discussion

Hepatic vagrancy, also known as wandering liver, is an exceptionally rare condition, with a mere 25 reported cases in the literature [7]. Most of these cases have been documented in young adult males [8,9]. Over 75% of the reported cases are associated with colonic abnormalities, primarily obstruction and/or volvulus [10].

Liver is positioned in the right hypochondrium, in a subphrenic position in its orthoptic location. Several mechanisms help maintain the liver in a relatively fixed position in the right upper abdominal quadrant. The attachment of the hepatic veins to the inferior vena cava and the presence of suspensory ligaments, including the falciform, coronary, and triangular ligaments, form the main support framework for the liver [11]. Superior and inferior displacements of liver are commonly observed in various abdominal and pulmonary pathologies [12]. However hepatic vagrancy or wandering liver is distinct from these in the context that there is some congenital, developmental or acquired weakness of suspensory ligaments and supporting mechanisms of the liver [13].

In some cases, wandering liver is incidentally discovered when a patient presents with non-specific abdominal pain and undergoes imaging [3]. However, in most instances, hepatic vagrancy is associated with bowel abnormalities, primarily involving the colon. Common colonic abnormalities include sigmoid volvulus, a mobile ascending or descending colon, and/or redundant transverse [9,11]. These findings have led to the proposal of hepato-colonic vagrancy syndrome. This syndrome is characterized by several features, such as liver hypermobility, poor tethering of the liver to inferior vena cava, and persistent midline falciform ligament and the absence of bare area of liver [8].

Researchers have hypothesized that there is abnormal persistence of the ventral mesentery. This persistent undifferentiated ventral mesentery contains hepatic veins, preventing the migration of the liver to the right hypochondrium. The fixed inferior vena cava acts as a fulcrum around which the liver rotates [3]. Embryologically, in a normal fetus, the mesentery develops into the dorsal and the ventral mesentery. The dorsal mesentery gives rise to the mesocolon, while ventral mesentery develops into the lesser omentum and the suspensory ligaments of liver. In patients with hepato-colonic vagrancy who underwent

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laparotomy, redundant mesocolon and the absence of the liver's suspensory ligaments (triangular and coronary ligaments) were frequent findings [10,12]. Although, there is a potential for vascular compromise, reported cases are very rare in literature [13].

To our knowledge, and after a thorough review of literature, our patient is the only female in the senior age bracket presenting with hepato-colonic vagrancy. The plane of liver rotation is oblique transverse, passing through porta hepatis, with the inferior vena cava acting as fulcrum. There were no indications of colonic obstruction, torsion or venous compromise in our patient. Our case is unique, as the patient had co-existent features of Autosomal Dominant Polycystic Kidney Disease, evidenced by multiple renal and splenic cysts. Another distinctive characteristic was the presence of a large right upper quadrant cyst with calcifications, which appeared to be of Echinococcal origin. It was postulated that patient's dyspeptic symptoms were due to the compression of the stomach and pylorus by the vagrant liver (Figure 4). It is also plausible that the hydatid cyst contributed to, or at least aggravated, the development of hepato-colonic vagrancy.

There is no single consensus regarding the management of hepato-colonic vagrancy. If the presenting clinical feature is bowel obstruction or volvulus, surgical intervention to relieve the obstruction should be performed, along with hepatopexy. For patients with mild or unrelated symptoms, non-surgical treatment can be considered, along with patient education on the warning signs of ischemia or obstruction [7,9,14].

Conclusion

In conclusion, hepatic vagrancy or wandering liver, is a rare condition that typically presents in young adults with symptoms of large bowel obstruction or volvulus. During surgical intervention for bowel obstruction, the absence of coronary and triangular ligaments of the liver is often noted, which is attributed to hepatic vagrancy. The frequent association of hepatic vagrancy with colonic obstruction suggests a common embryological and developmental link. Our patient, a senior female, presented with dyspepsia and was further evaluated through cross sectional imaging. Hepato-colonic vagrancy was identified as the principle finding, with no radiological evidence of intestinal obstruction. Additional findings included multiple bilateral renal cysts, multiple splenic cysts and a calcified hepatic cyst, the latter being suggestive of echinococcal origin. This case study underscores the importance of recognizing both normal and abnormal anatomical variations for accurate diagnosis and management. The patient was managed conservatively with lifestyle and dietary modifications and remained stable at 2- and 6-month follow-ups. This case study highlights the need for thorough imaging and evaluation in patients with atypical abdominal symptoms to ensure accurate diagnosis and appropriate management.

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