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Coincidence of left isomerism, malposition of cecum, dorsal pancreatic agenesis, and retroaortic left renal vein: A case report

Sol izomerizmim çekum malpozisyonu, dorsal pankreas agenezi ve reroaortik sol renal ven varlığı ile birlikteliği: Olgu sunumu

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Abstract

Polysplenia / heterotaxy syndrome is a very rare condition that occurs as a result of the maldistribution of the thoracic and abdominal organs. It is examined in two groups with different clinical features and anatomical variations: right and left isomerism. This paper reports the case of a 47-year-old female patient who underwent a non-contrast computed tomography (CT) examination due to right flank pain and suspicion of a renal stone and presented with the findings of left isomerism; i.e., accompanied by intraabdominal variations. On abdominal CT examination, stomach and multiple spleen were localized in the right upper quadrant, and the liver had a midline localization. The interruption of the inferior vena cava with azygos continuation was also detected. The hepatic veins were draining directly into the right atrium. In the literature, a retroaortic left renal vein and dorsal pancreatic agenesis have been individually reported to coexist with left isomerism, but no other case of the coincidence of these three anomalies has been described, which makes the current case report significant. Having knowledge of anatomical variations and clinical status can prevent misdiagnosis, and imaging findings are crucial in the planning of a possible surgical procedure.

Keywords: Left isomerism, Computed tomography, Dorsal pancreatic agenesis, Retroaortic left renal vein, Malposition of cecum

Öz

Polispleni / heterotaksi sendromu, torasik ve abdominal organların yanlış dağılımı sonucu olarak ortaya çıkan çok nadir görülen bir durumdur. Farklı klinik özelliklere ve anatomik varyasyonlara sahip iki grupta incelenmiştir; sağ ve sol izomerizm. Sağ yan ağrısı ve böbrek taşı şüphesi nedeniyle kontrastsız bilgisayarlı tomografi (BT) çekimi yapılan 47 yaşındaki kadın hastada, sol izomerizm – polispleni sendromu ve eşlik eden abdominal organ varyasyonları saptandı. Abdominal BT incelemesinde sağ üst kadranda yerleşimli mide ve çok sayıda dalak, orta hat yerleşimli karaciğer tespit edildi. İnferior vena kava devamlılığında kesilme ve azigos devamlılığı tespit edildi. Hepatik venler doğrudan sağ atriyuma drene oluyordu. Tespit edilen bulgular, polispleni-sol izomerizm ile uyumlu olarak değerlendirildi. Retroaortik sol renal ven, çekum malpozisyonu ve dorsal pankreas agenezisine izomerizm eşlik ediyordu. Literatürde ayrı ayrı rapor edilen sol izomerizme eşlike eden retroaortik sol renal ven ve dorsal pankreas agenezisi olgu sunumları mevcuttu. Ancak bu üç anomalinin eşlik ettiği olgu sunumu yoktu ve bu özellikleriyle olgumuz tekdi. Anatomik varyasyonları ve klinik durumu bilmek yanlış tanıyı önleyebilir ve olası bir cerrahi işlemin planlanmasında görüntüleme bulgularını bilmek çok önemlidir.

Anahtar kelimeler: Sol izomerizm, Bilgisayarlı tomografi, Dorsal pankreatik agenezi, Retroaortik sol renal ven, Çekum malpozisyonu

Introduction

Heterotaxy syndrome is a very rare anomaly characterized with an incorrect position of the left and right abdominal and thoracic organs [1,2]. This syndrome has two types as right and left isomerism [2]. Right isomerism presents with a severe form of cyanotic cardiac disease associated with asplenia [3] and is accompanied by anomalies, such as bilateral epiarterial bronchi, bilateral three-lobed lungs, bilateral right atria, and intestinal malrotation. Left isomerism is often seen with the findings of multiple spleens, interruption of the inferior vena cava (IVC) with azygos continuation, non-orthotopic liver, bilateral hyperatrial bronchi, bilateral bi-lobed lungs, and bilateral left atria [4]. On computed tomography (CT), anatomical anomalies can be described in detail and other accompanying conditions can be detected. Although the coexistence of left isomerism with either dorsal pancreatic agenesis or left retroaortic renal venous variation has been reported in the literature, the coincidence of a left retroaortic renal vein, malposition of cecum, and dorsal pancreatic agenesis with left isomerism has not been previously described. In this study, we aimed to present the CT imaging findings of a rare left isomerism case presenting with these three anomalies.

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Case presentation

A 47-year-old female patient underwent an abdominal CT scan using a 128-slice CT device (GE, Revolution EVO, USA) due to right flank pain and suspicion of a renal stone. A 6-mm renal stone was found at the lower pole of the right kidney and there were also heterotaxy findings of intraabdominal organs. The liver was located in the midline (Figure 1, Figure 2). While the normal appearance of the stomach and the spleen was not observed in the left upper quadrant, five spleen tissues of different sizes were detected instead of a normal spleen in the right upper quadrant (Figure 1). Duodenum and ileum were located on the left side of the abdomen, and stomach and jejunum were on the right. The vena cava inferior was located on the right but interrupted in the intrahepatic segment with azygos continuation. The azygos was dilated (Figure 2). The hepatic veins were draining directly into the right atrium. All these findings were consistent with left isomerism; i.e., polysplenia syndrome. In addition, the pancreatic head and uncinate process were normal, but the body and tail parts were not observed. The pancreas was short and had a vertical orientation. There was also dorsal pancreatic agenesis (Figure 3). CT showed retroaortic left renal vein variation (Figure 1). While the colon was not visible in the right half of the abdomen, the cecum and transverse column were located on the left half of the abdomen (Figure 4). To our knowledge, the coexistence of left isomerism with the malposition of cecum, dorsal pancreatic agenesis, and a retroaortic left renal vein has not been previously reported in the literature. Therefore, the current case is significant due to the rarity of observing these three anatomical anomalies in a patient with left isomerism. Written and informed consent was obtained from the patient for publishing this case report.

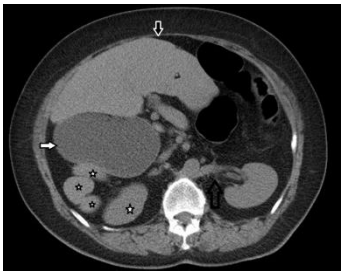

Figure 1: Axial section non-contrast CT showing the midline-located liver (black arrow), multiple spleens (stars), right-sided stomach (white arrow), and retroaortic left renal vein (empty arrow)

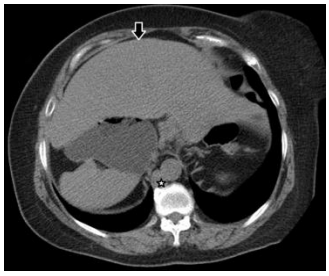

Figure 2: Axial section non-contrast CT showing the midline-located liver (black arrow) and dilated azygos vein (star)

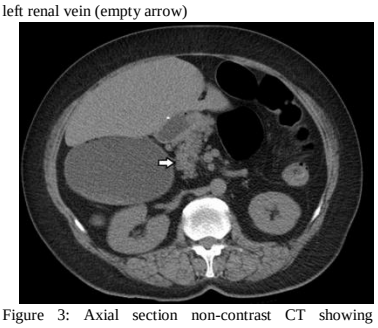

Figure 3: Axial section non-contrast CT showing vertically oriented short pancreas (white arrow)

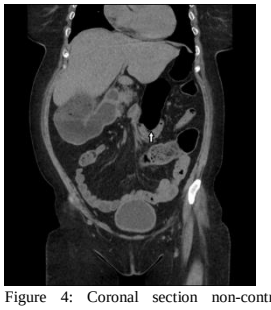

Figure 4: Coronal section non-contrast CT showing the cecum and transverse colon in the left upper quadrant (arrow)

Discussion

Situs solitus refers to the intraabdominal and thoracic organs being located in the normal anatomic position [5] while situs inversus is the opposite localization of these organs, which appears as a mirror image of the normal position [5,6]. Situs ambiguous is an incomplete mirror image anomaly, which describes the state between these two conditions [2,7] and it is divided into right and left isomerism. Right isomerism has severe cardiac anomalies with asplenia and is more common in the male gender. Infants with this condition often die in their first year due to both severe cardiac problems and severe infections caused by asplenia [3]. Left isomerism is more common in females, and it presents with milder, non-cyanotic cardiac anomalies. Immunodeficiency is not an expected finding because of functional spleen parenchyma. For this reason, it can be incidentally diagnosed in adulthood [4,8]. In our case, a 47-year-old female patient was diagnosed incidentally on CT.

Although the actual cause of polysplenia is unknown, many factors of embryological, congenital and teratogenic origin have been reported to play a role in its etiology [8]. Genetic defects in metalloproteinases (e.g., MMP21) that regulate normal right-left asymmetry and the failure of developmental regulatory transcription factors (e.g., FOX A2) may be responsible for the impairment of normal development at embryological stages [9]. Polysplenia is very rare with an incidence of 2.5/100,000.

The spectrum of polysplenia findings is very diverse and variable. Apart from typical IVC anomalies seen in polysplenia, there may be many accompanying vascular variations [1,4]. One of the most common accompanying vascular malformations is a preduodenal portal vein [4,7,8]. In addition, a retroaortic renal vein is common in both normal populations and patients with polysplenia, but it is not a specific finding or a diagnostic criterion for polysplenia. Thus, we consider that the presence of this finding was probably coincidental in our case, as well as in the limited number of previously reported polysplenia cases.

The pancreas may be completely normal in patients with polysplenia, but a short pancreas is a common pancreatic anomaly in this syndrome [1-3,9]. The head of the pancreas has a normal size, but the neck and tail of the pancreas are absent. In our case, while the pancreas had no tail or neck, a normal-size pancreatic head was seen with a vertical course.

In polysplenia syndrome, the stomach can be on the right or left side. Upper intestinal non-rotation/malrotation occurs frequently [2,9]. In our case, the stomach and jejunum were on the right side, and the ileum was located mostly on the left and partially on the right side. Position anomalies of the colon, as in many other organs, are expected findings in polysplenia syndrome. However, in this syndrome, there is no definitive finding of the colon position in the literature. In our case, there was evidence of the malposition of cecum. The cecal and ileocecal valves were located in the left upper quadrant of the abdomen.

Our case had typical features of polysplenia syndrome, but the interesting finding was the coincidence of dorsal pancreatic agenesis, a retroaortic left renal vein, and colonic malposition.

Conclusions

Polysplenia syndrome has a broad spectrum of findings, which are often combinations of various anomalies [1-4,7,8]. Besides the specific findings of polysplenia, common anomalies, such as a short pancreas, or completely incidental variations; e.g., a left retroaortic renal vein can be detected, as in our case.

Having knowledge of specific imaging findings is very important in diagnosis. Radiologists should be aware of this syndrome and should not consider these findings as separate entities. Asymptomatic patients with polysplenia syndrome can be diagnosed incidentally. Sometimes the initial examination can be ultrasound, but CT should be undertaken for definitive diagnosis since it is able to demonstrate all the components of associated congenital anomalies. When radiologists diagnose polysplenia syndrome, they should make an evaluation of the existence of other variations and anomalies. Having good knowledge of the whole anatomy is very important to prevent possible complications and decide on the necessity of a surgical procedure.

References

1. Jung JE, Hur JH, Jung MK, Kwon A, Chae HW, Kim DH, et al. Diabetes mellitus due to agenesis of the dorsal pancreas in a patient with heterotaxy syndrome. Ann Pediatr Endocrinol Metab. 2017 Jun;22(2):125-8.
2. Gupta AC, Herts B. Heterotaxia with polysplenia. J Urol. 2015 Sep;194(3):801-2.
3. Yildiz AE, Ariyurek MO, Karcaaltincaba M. Splenic anomalies of shape, size, and location: pictorial essay. Scientific World Journal. 2013 Apr 21;2013:321810.
4. De Wailly P, Metzler P, Sautot-Vial N, Olivier D, Meunier B, Faure JP. Pre-duodenal portal vein in polysplenia syndrome: clinical effects and surgical application. Surg Radiol Anat. 2011 Jul;33(5):451-4.
5. Gupta R, Soni V, Valse PD, Goyal RB, Gupta AK, Mathur P. Neonatal intestinal obstruction associated with situs inversus totalis: two case reports and a review of the literature. J Med Case Rep. 2017 Sep 18;11(1):264.
6. Douard R, Feldman A, Bargy F, Loric S, Delmas V. Anomalies of lateralization in man: a case of total situs inversus. Surg Radiol Anat. 2000;22(5-6):293-7.
7. Varga I, Babala J, Kachlik D. Anatomic variations of the spleen: current state of terminology, classification, and embryological background. Surg Radiol Anat. 2018 Jan;40(1):21-9.
8. Seo HI, Jeon TY, Sim MS, Kim S. Polysplenia syndrome with preduodenal portal vein detected in adults. World J Gastroenterol. 2008 Nov 7;14(41):6418-20.
9. Pagkratis S, Kryeziu S, Lin M, Hoque S, Bucobo JC, Buscaglia JM, et al. Case report of intestinal non-rotation, heterotaxy, and polysplenia in a patient with pancreatic cancer. Medicine (Baltimore). 2017 Dec;96(49):e8599.

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