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Polysplenia Syndrome Detected after Chest Symptoms in Two Adult Patients: Case Report and Review of Literature

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	<h3>Summary</h3>
Background:	Polisplenia syndrome (PSS) is a rare subtype of heterotaxy syndrome and means ambiguous location of the major thoracic and abdominal organs with vascular anomalies and multiple spleens. We reported on the findings of computed tomography (CT) of PSS in adults, detected incidentally.
Case Report:	Two woman underwent a CT examination of the thorax for different thoracic pathologies. There were common abnormalities such as hyparterial bronchi and absence of middle lobe fissure on CTscans suggesting heterotaxy syndrome. Therefore, the abdominal CTs were performed to detect the accompanying abdominal anomalies. Our two cases defined as PSS were diagnosed with multiple spleens in the normal location in the abdomen. The left-dominant liver and short pancreas with agenesis of the pancreatic tail and lateral part of the body were detected on CT scan. In the first case, the vascular abnormalities were as follows: variant entrance of the main portal vein into the liver and atypically located superior mesenteric vein (SMV) joining with the splenic vein to form the portal vein. In the second case, the preduodenal portal vein and hemiazygos continuation with interruption of the hepatic segment of the inferior vena cava (IVC) were the vascular anomalies. The bowels were malrotated in the second case.
Conclusions:	Although such cases are usually admitted as abdominal emergency, our two cases were detected during examinations for thoracic and cardiac pathologies. The knowledge and awareness of PSS can be helpful to diagnose pathology and plan surgical procedures.
MeSH Keywords:	Heterotaxy Syndrome • Multidetector Computed Tomography • Portal Vein
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Background

The term situs describes the position of atria, tracheo-bronchial tree, pulmonary arteries, thoracic and abdominal viscera [1]. Situs ambiguous or heterotaxy syndrome means malposition and dysmorphism of the thoracic and abdominal organs with vascular anomalies. The two main subtypes of situs ambiguous are situs ambiguous with polysplenia and situs ambiguous with asplenia. Polisplenia syndrome (PSS) is characterized by ambiguous location of the major thoracic and abdominal organs with vascular anomalies and multiple spleens [2,3]. Azygos or hemiazygos continuation of the inferior vena cava (IVC) with absence of the hepatic segment is the most frequent, associated anomaly [4,5]. Situs anomalies are usually detected incidentally in adults during imaging evaluations for other causes such as tuberculosis. We presented the radiological findings

of PSS cases in adults and the current literature related to this syndrome.

Case Report

Case 1

A19-year-old woman with a medical history of bronchiectasis underwent computed tomography (CT) of the thorax. CT findings of tuberculosis, bronchiectasis and mediastinal lymph nodes were present on CT scan at the time of admission. Abnormalities such as hyparterial bronchi and absence of middle lobe fissure were seen on the examination suggesting heterotaxy syndrome. Hence, further abdominal CT examination was performed to show other accompanying anomalies. The abdominal CT revealed a normally positioned left-dominant liver. The pancreas was normally

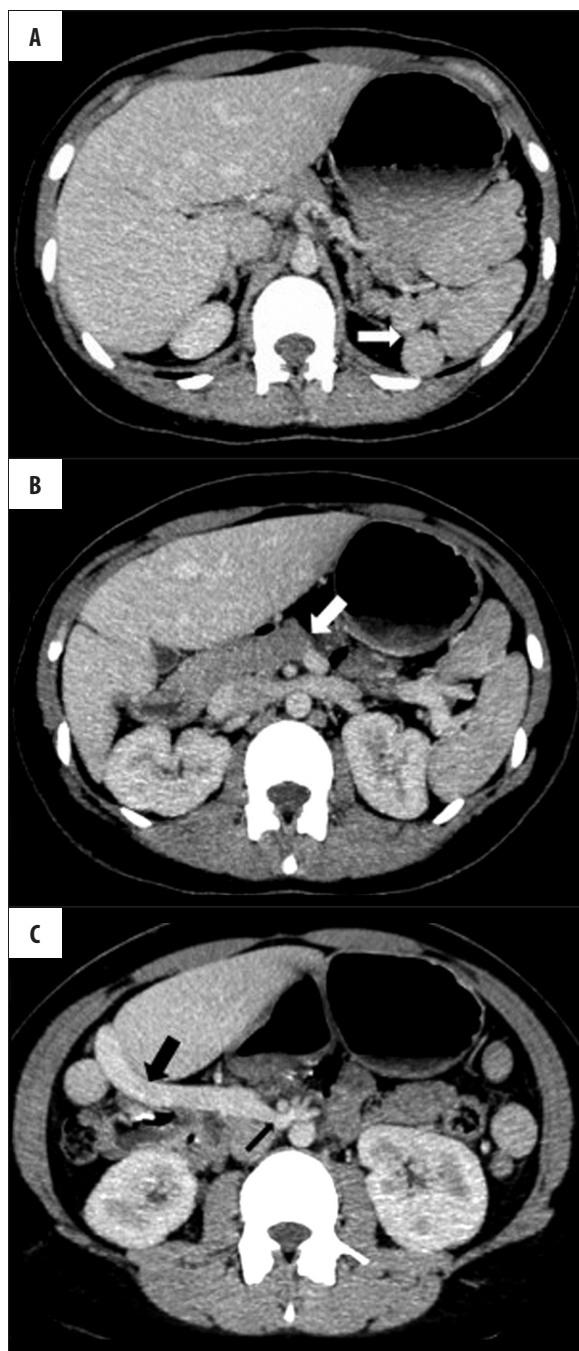


Figure 1. (A) Axial CT scan of the upper abdomen showed multiple spleens in the left upper quadrant (white arrow). (B) Axial CT scan demonstrated a short pancreas with agenesis of the pancreatic tail and lateral part of the body (arrow). (C) Axial CT scan demonstrated the variation in the entrance of the main portal vein to the liver. The main portal vein was transversing the left and hypoplastic right liver lobes horizontally from midline to the lateral contour of the liver (thick black arrow). SMV was joining the splenic vein after passing between the SMA and aorta (thin black arrow).

located, i.e. in the midline, with agenesis of the pancreatic tail and lateral part of the body. There were multiple spleens without positional anomaly on CT scan. The abdominal IVC was in the normal position, right to the midline,

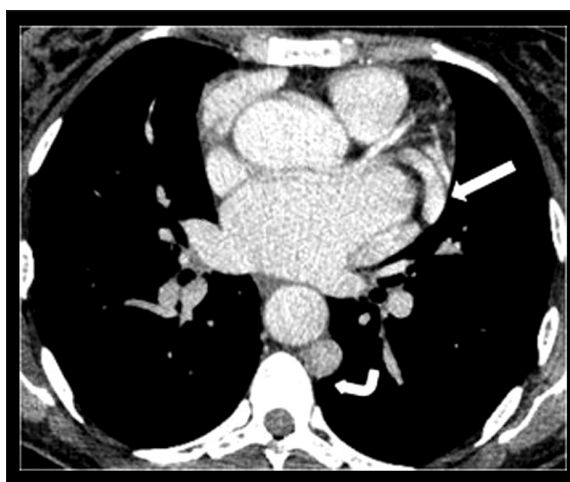


Figure 2. Thorax CT scan revealed that the left-sided IVC (curved arrow) and circumflex coronary artery were draining into dilated coronary sinus (arrow).

and continuous with intrahepatic part draining into the right heart. The superior mesenteric vein (SMV) joined to the splenic vein after passing between the superior mesenteric artery (SMA) and aorta, and then it formed the main portal vein lateral to the SMA. The main portal vein was transversing the left and hypoplastic right liver lobes horizontally from midline to the lateral contour of the liver and entered the left liver lobe after making a u-turn (Figure 1A-1C). The main portal vein was bifurcating in the left-dominant liver.

Case 2

A 46-year-old woman with patent ductus arteriosus presented with shortness of breath. She was examined with CT of the heart to discover cardiovascular anomalies. It was shown that left-sided IVC and circumflex coronary artery were draining into a dilated coronary sinus (Figure 2). Intrahepatic veins were draining into the right atrium. She had bilateral bilobed lungs on CT of the thorax without the minor fissure. The abdominal CT was performed to detect the accompanying vascular anomalies and malrotation in the abdomen. The left-dominant liver was located in the midline of the abdomen. The pancreas revealed agenesis of the pancreatic tail and lateral part of the body. The spleen had four foci in the left-hand side of the abdomen. Hemiazygos continuation with interruption of the hepatic segment of IVC was noticed on the left side of the aorta. Preduodenal portal vein was detected on the abdominal CT scan (Figure 3A, 3B). The stomach was located on the left-hand side in relation to the midline. The bowels were malrotated as small intestines were located in the lower right quadrant of the abdomen. However, the descending and sigmoid colon were normally located, the cecum and the ascending colon were placed in the upper left quadrant of the abdomen (Figure 4).

Those two cases were diagnosed as PSS with morphological and vascular anomalies.

Discussion

The term situs means the position of atria, tracheobronchial tree, pulmonary arteries, thoracic and abdominal



Figure 3. (A) Axial CT scan of the upper abdomen showed the left-dominant liver (thick white arrow) and multiple spleens in the left upper quadrant (thin white arrow). Abdominal IVC was noticed on the left side of the aorta (black arrow). (B) Axial CT scan demonstrated preduodenal portal vein (white arrow) and left-sided abdominal IVC (black arrow).

viscera [1]. Situs anomalies are rare and confusing conditions. Situs solitus is the normal position of organs and vessels within the body. The mirror image position of normal (situs solitus) is called situs inversus. Situs ambiguous or heterotaxy syndrome indicates malposition and dysmorphism of the thoracic and abdominal organs with vascular anomalies. It is subclassified as situs ambiguous with polysplenia and situs ambiguous with asplenia [2,3].

In PSS (double left-sidedness or left isomerism) the patient has bilateral bilobed lungs, bilateral pulmonary atria, abnormal location of abdominal solid organs and malrotation of bowels with multiple spleens. Although there is no single characteristic pathognomic abnormality of PSS, azygos or hemiazygos continuation of the IVC with absence of the hepatic segment is the most frequent associated anomaly. This syndrome is more common in females than males [2,4,5]. The etiology of PSS is not known yet. The causative factors of PSS are thought to be association of embryonic, genetic and teratogenic components [6].



Figure 4. Axial CT scan of the lower abdomen showed small intestines in the right side of the abdomen (arrowhead) and colon in the left side (arrow).

Cardiac anomalies are less common and less confusing in PSS than asplenia syndrome [2]. Coronary sinus is sometimes absent in PSS [1]. However, coronary sinus was dilated and left-sided IVC and circumflex coronary artery were draining into coronary sinus in our second case. PSS is described mainly in childhood due to critical cardiac anatomic malformations or biliary atresia. It is detected incidentally in adults during imaging evaluation for unrelated conditions or in symptomatic cases with anatomical abnormalities [7].

Our two cases were women and diagnosed as PSS on CT examination at the time of investigation for other problems, as mentioned before. Bilateral hyperarterial bronchi and bilateral bilobed lungs were seen in both cases. In our cases, the stomach and multiple small spleens were on the left side of the abdomen. However, there were different variations in those cases. The livers were left-dominant whereas the liver was in the midline in the second case and right-handed in the first case. Hemiazygos continuation with interruption of the hepatic segment of IVC was revealed in our second case, as the most common finding of vascular anomalies.

The preduodenal portal vein is a congenital anomaly of the portal vein that passes in front of the duodenum. The 'S'-shaped normal portal vein results from the loss of caudal and cranial communication in the normal development. The preduodenal portal vein is a result of disturbed process of development [8,9]. In our second case, the preduodenal portal vein was seen with the loss of its 'S' shape. In the first case, the vascular abnormalities concerned the entrance of the main portal vein to the liver and its course after joining the SMV, with its atypical location, i.e. between the SMA and aorta.

Normal pancreas is formed by fusion of the ventral and dorsal pancreatic buds. The uncinata process and the head are formed from the ventral pancreatic bud. The dorsal pancreatic bud whose development occurs in the dorsal mesogastrium together with the spleen gives rise to the body and tail. This explains why the anomalies of pancreas can be expected in patients with PSS [10]. In our two cases, agenesis of the pancreatic tail and lateral part of the body was revealed.

As mentioned before, there is no characteristic abnormality in PSS. We presented the abnormalities of cases with this syndrome.

Conclusions

PSS is a rare syndrome of congenital abnormalities involving the cardiovascular structures and visceral organs in

the abdomen that are diagnosed incidentally in adults. The cases usually present with urgent abdominal complaints at the emergency department. In contrast, our two cases were detected during the examinations for thoracic and cardiac pathologies. The knowledge and recognition of the PSS can be helpful to diagnose pathology and plan surgery.

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