



## An Incidental Findings of Polysplenia Syndrome in an Adult Patient with Multiple Anomalies

**Mehmet Sedat Durmaz\*, Ali Cengiz, Serdar Arslan, Hasan Erdogan, Ismet Tolu and Arzu Cengiz**

Department of Radiology, University of Health Sciences, Konya Education and Research Hospital, Turkey

\*Corresponding author: Mehmet Sedat Durmaz, MD, Specialist, Department of Radiology, University of Health Sciences, Konya Education and Research Hospital, Konya, 42090, Turkey, Tel: +90-332-221-00-00, Fax: +90-332-323-67-23, E mail: [alicengiz7158@gmail.com](mailto:alicengiz7158@gmail.com)

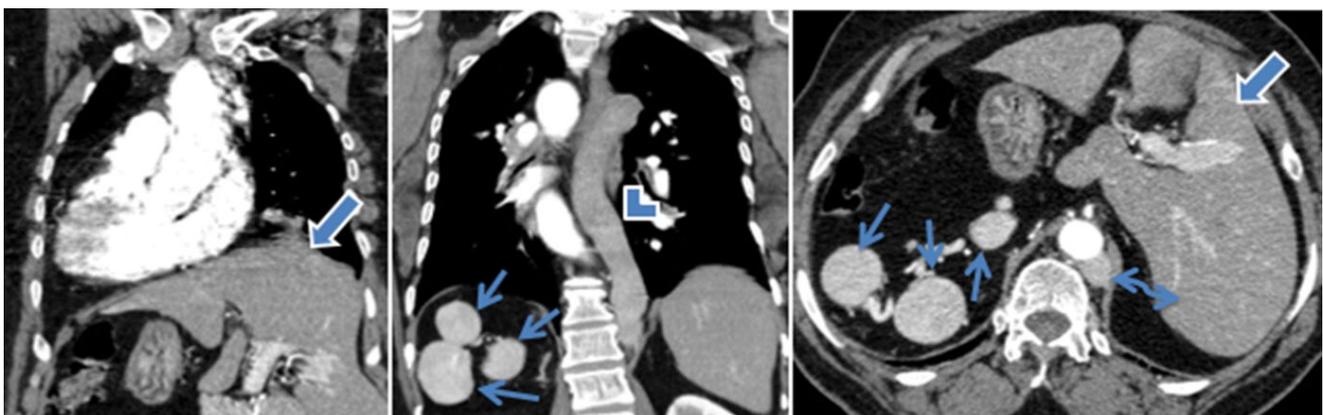
### Introduction

Polysplenia syndrome is a rare congenital subtype of heterotaxy syndrome associated with various visceral and vascular anomalies. Polysplenia syndrome is characterized by presence of two or more spleens and anomalies of other asymmetric organs. It is reported incidence of 1 per 250.000 and more common in females. Here, we report the case of a 53-year-old woman who presented with polysplenia syndrome. The patient had multiple accessory spleens in right sided, dextrocardia, interrupted inferior vena cava with azygos continuation, left-sided liver and congenital short pancreas. Our patient was asymptomatic until now and were diagnosed incidentally.

### Case Report

53-year-old female patient was admitted to our internal medicine clinic with mildly abdominal pain and debility. There was no significant evidence in the patient's medical history. Physical examination showed mild abdominal sensitivity and dextrocardia, other findings were normal. Ultrasound examinations

were performed simultaneously with physical examination. Ultrasound demonstrate the presence of liver in the upper left quadrant of abdomen and multiple nodular spleen structures in the right upper quadrant of abdomen. Abdominal and chest computed tomography (CT) examination were performed to confirm the diagnosis of polysplenia and detect the accompanied additional findings. Initial CT scan shows partial situs inversus and a right-sided heart which is concomitant to right aortic arch. Liver was in the upper left quadrant of abdomen and multiple nodular spleen structures in the right upper quadrant (Figure 1). There was enlarged azygos vein on the left side of descending aorta and vena cava superior was left side of vertebral column (Figure 2). The corpus and tail of pancreas could not be monitored, there was only head of the pancreas in left quadrant (congenital short pancreas) (Figure 3). Vena cava inferior could not be monitored also. Both renal veins drain into azygos vein. Hepatic vein emptying into right atrium (Figure 3). Accordingly, the right lung consists of two lobes and left lung consisted of three lobes. Patient is diagnosed with polysplenia syndrome.

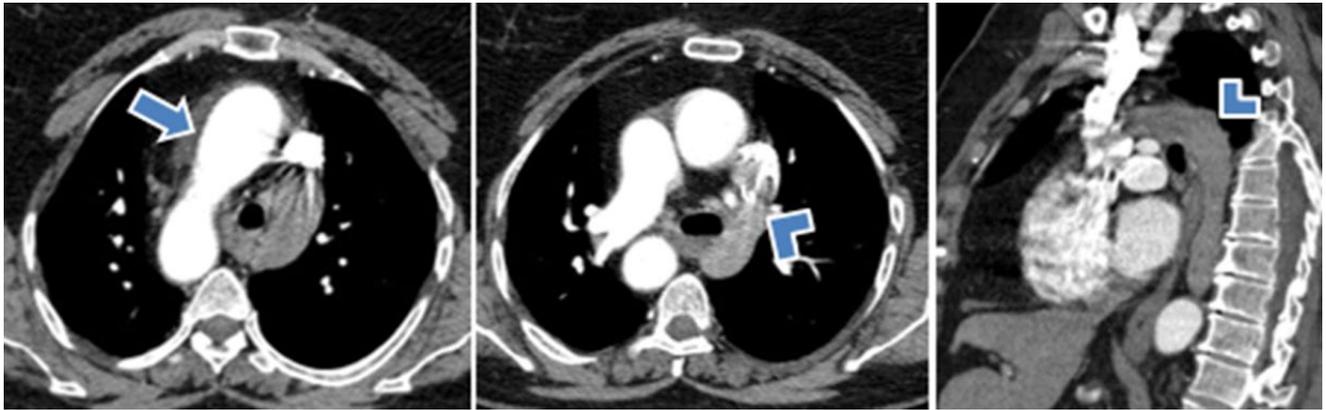


**Figure 1:** The coronal and axial enhanced CT show, heart is in the right hemitorax (dextrocardia), liver is in the left upper quadrant of abdomen (arrow), there are multiple nodular spleen structures in the right upper quadrant (thin arrow). There is enlarged azygos vein on the left side of descending aorta (arrow head) and azygos vein is on the left side of aorta in the abdominal imaging (curved arrow).

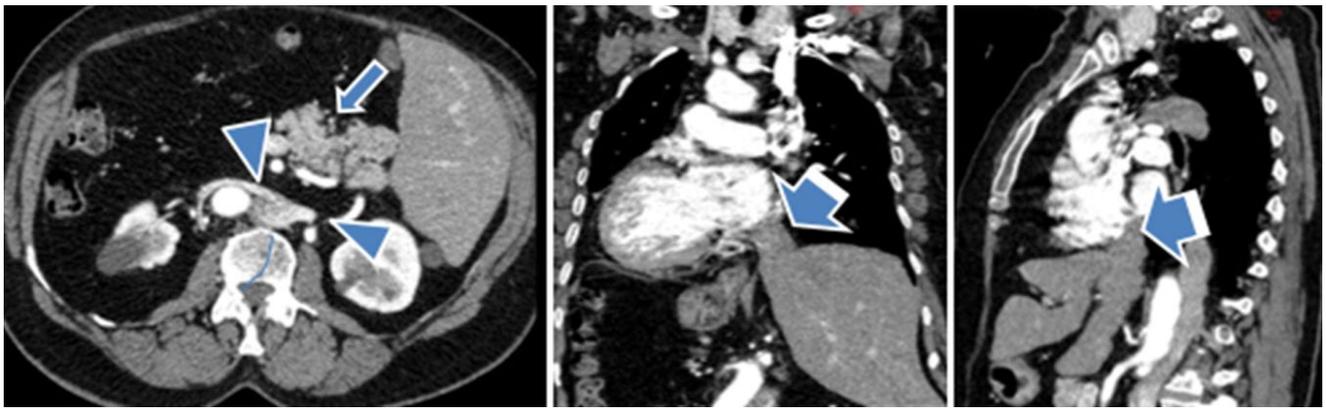
**Citation:** Durmaz MS, Cengiz A, Arslan S, Erdogan H, Tolu I, et al. (2016) An Incidental Findings of Polysplenia Syndrome in an Adult Patient with Multiple Anomalies. Clin Med Rev Case Rep 3:148

**Received:** October 14, 2016; **Accepted:** December 13, 2016; **Published:** December 16, 2016

**Copyright:** © 2016 Durmaz MS, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.



**Figure 2:** Axial and sagittal enhanced CT show right aortic arch (arrow). Vena cava superior is in the left side and enlarged azygos vein drain into the vena cava superior (arrow head).



**Figure 3:** Head of pancreas is in the left upper quadrant (arrow). Renal veins drain into the azygos vein (arrow heads). Vena cava inferior isn't detected and hepatic veins directly drain into the right atrium (thick arrow).

## Discussion

Polysplenia syndrome is a rare congenital disorder, which is characterized by multiple spleens and accompanying cardiopulmonary and abdominal anomalies that is a type of situs ambiguus believed to be left isomerism in most cases. Patients usually do not have a complaint and this congenital anomaly is noticed incidentally during abdominal surgery or radiological examination. Symptomatic patients presented with atrioventricular septal defects and may have anomalous inferior vena cava or other gastrointestinal structural abnormalities including partial or complete agenesis of the dorsal pancreas [1]. These findings may have clinical relevance with a possible increased risk for intestinal volvulus, diabetes mellitus or pancreatitis. The certain underlying etiology of polysplenia is unknown. Embryonic, genetic and teratogenic components have all been implicated as causative factors in polysplenia.

The splenic structures is generally divided into fairly same sized masses, varying in number from 2 to 6 and ranging from 1 to 6 cm in diameter, Total quantity is equal the mass of a normal spleen. The location of the spleens is in either the left or right upper quadrant. The most frequent positional abnormalities of liver and gallbladder can be found in the midline or mirrored. Usually the pancreas is forming part of the head and found in the left upper quadrant. Gastrointestinal malrotation and renal agenesis or hypoplasia may be seen [2]. The pulmonary manifestations include bilateral bilobed lungs [3].

More than 40 percent of reported cases have cardiac anomalies and majority of such children can not survive very long in the first five years of life. The most common cardiac anomalies are atrial septal defect (78%), azygos continuation of the inferior vena cava (65%), ventricular septal defect (63%). Death can occur at an early age especially in case of concomitant cardiovascular abnormalities [4].

In conclusion, polysplenia syndrome is a infrequently seen disease, which may be accompanied by multiple systemic abnormalities. It is important to be familiar with this syndrome especially in order not to overlook accompanying abnormalities. Computerized tomography examinations are very helpful in the evaluation of polysplenia syndrome patients.

## References

1. Matthias M, Wiesner W, Mengiardi B (2007) Annular pancreas and agenesis of the dorsal pancreas in a patient with polysplenia syndrome. *American Journal of Roentgenology* 188: W150-W153.
2. Kapa S, Gleeson FC, Vege SS (2007) Dorsal pancreas agenesis and polysplenia/heterotaxy syndrome: a novel association with aortic coarctation and a review of the literature. *JOP* 8: 433-437.
3. Fukuda K, Onda T, Kimura Y, Miura S, Matsumori R, et al. (2015) An adult case of polysplenia syndrome associated with sinus node dysfunction, dextrocardia, and systemic venous anomalies. *Internal Medicine* 54: 1071-1074.
4. Kumar P, Chandra K, Prajapati N (2014) An Incidental Finding of Polysplenia Syndrome in a Geriatric Patient: A Case Report. *Indian Journal of Clinical Practice* 25.