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Polysplenia Syndrome with Commun Mesenter, Reverse Pancreas, IVC Interruption Discovered in an Adult

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Abstract **Case Report**

Polysplenia syndrome is anomaly associated with multiple spleens and anomalies of chest and abdominal organs. We present a case of polysplenia syndrome found in a 27-year-old man. Only those with mild anatomical abnormalities reach adulthood without a diagnosis. Radiological examinations are helpful to the diagnosis by showing the location and aspect of spleen, location of other organs in the abdomen, and identification of other associated anomalies. In this article, we illustrate anomalies of polysplenia syndrome and focus on abdominal organs.

Keywords: Imaging, Polysplenia, Abdomen.

INTRODUCTION

Polysplenia syndrome is a rare congenital subtype of heterotaxia syndrome associated with various visceral and vascular abnormalities. It is defined by the presence of 2 or more spleens and anomalies of other asymmetric organs [1]. During embryogenesis, the spleen plays an important role in the normal development and lateralization of visceral organs. Accordingly, anomalies of the spleen are associated with many congenital anomalies of other organs and show large spectrum of abnormalities in the abdominal organs [2]. Technical advances in sonography, computed tomography, and magnetic resonance imaging have widely enhanced our ability to detect and characterize these anomalies [3].

CASE REPORT

A 27 year old male military officer was admitted to our radiology department with acute abdominal pain. The clinical examination was normal. A computed tomography was performed showing: A median liver .Stomach and multiples spleens (4 in total) were on the right side. Pancreas and duodenum were on the left side. It has also showed a common mesentery. An Inferior vena cava (IVC) interruption with azygous continuation has also been with a suprahepatic veins flow directly to the right atrium. The diagnosis of polysplenia syndrome was made.

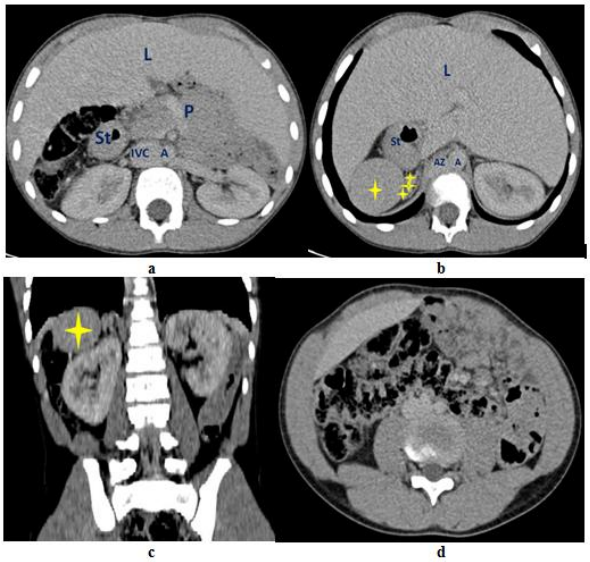

Fig-1: Polysplenia syndrome in an adult with common mesentery, and IVC interruption. (a) Axial CT image of the abdomen at a upper level showing a median liver (L). The Pancreas on the left side. (b) Axial and (c) coronal CT imagesshows the spleens (stars) on the right side, the same side of the stomach (St) and azygos vein (Az) on the right side. (d) Axial CT image of the abdomen showing common mesentery.

DISCUSSION

Polysplenia syndrome characterized by the association of 2 or more multiple spleens with multiple congenital abnormalities in abdomen and chest.

However, some cases of polysplenia syndrome have been described with a single bilobed spleen or a single normal splenic gland. The splenic mass is usually divided into fairly equalized masses, varying in number from 2 to 6 and ranging from 1 to 6 cm in diameter. In all cases, the spleens are still located on the same side of the stomach along the greater curvature due to the development of embryonic splenic tissue in the posterior mesogastrium [4, 7].

It is a rare congenital disease initially described by Helwig in 1929 [3, 5]. Since then, few cases were described in the literature with an incidence of 1/250,000 live births [6].

The pancreas may be normal, short in size, or have a cleft created by traversing vessels [1, 8, 9]. An semiannular pancreas and annular pancreas also have been reported 11. The most common is a short pancreas with agenesis of the dorsal portion [4].

Polysplenia is commonly associated with anomalies of intestinal rotation. Intestinal rotation abnormalities include nonrotation (the small bowel is entirely on the right of the spine and the colon on the left), incomplete rotation (the appearance is between normal and non-rotation), and the rare reversed complete or incomplete rotation [1, 10].

Gastrointestinal tract rotational anomalies may be a rare cause of abdominal pain in adults and may cause bowel obstruction, such as midgut volvulus [10]. Inferior vena cava interruption with azygous continuation is the second most common abnormality observed in polysplenia patients after multiple spleens [1].

A preduodenal portal vein was reported as one of the anomalies in association with polysplenia,this rare anomaly resulting from maldevelopement of portal venous système or malposition of the abdominal vescera [11].

CONCLUSION

Polysplenia syndrome is a complex syndrome with a wide spectrum of abnormalities, the most common of which are multiple spleens and IVC interruption with azygous continuation. A CT-scan with reconstruction is excellent in visualizing these anomalies.

The knowledge of various radiologic features plays an essential role in the diagnosis of this rare syndrome.

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