SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Incidental Adult Polysplenia, Interrupted Inferior Vena Cava with Azygos Continuation and Semiannular Pancreas

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DOI: 10.36347/sasjm.2024.v10i05.029 | **Received:** 13.04.2024 | **Accepted:** 21.05.2024 | **Published:** 25.05.2024

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Abstract **Case Report**

Polysplenia syndrome is a rare condition (about 2.5:100,000 live births) wherein a person has two or more spleens with accompanying various thoracic and abdominal organ abnormalities. Polysplenia is usually associated with situs ambiguous, while polysplenia with situs inversus is very rare. Herein, we report a rare case of incidentally found polysplenia syndrome in a 40-year-old female who had an interrupted inferior vena cava (IVC) with azygos continuation and also semiannular pancreas, which is a rare finding.

Keywords: Polysplenia; Annular pancreas; CT.

INTRODUCTION

Polysplenia syndrome (PS) is a rare disease (about 2.5:100,000 live births) wherein a person has two or more spleens with accompanying various thoracic and abdominal organ abnormalities. Adult presentation of heterotaxic syndromes and related complexes, approximately 50–90% of polysplenia cases are associated with cardiac abnormalities, and only about 10% of people are expected to live until adulthood without complications [1]. polysplenia is usually associated with the situs ambiguous anomaly [1-3]. When diagnosed in adults, PS is most often an incidental finding on abdominal sonography or computed tomography (CT) performed for other reasons.

Here, we report a rare case of incidentally found polysplenia in a 40-year-old female patient who also had an interrupted inferior vena cava (IVC) with azygos continuation, and also semiannular pancreas, which is an exceedingly rare finding.

CLINICAL CASE

This is the case of a 40-year-old Caucasian female was admitted to the hospital with complaints of generalized abdominal pain, shortness of breath, fatigue, and weight loss. No medical or chirurgical past.

Physical examination demonstrated tenderness to palpation in the right lower quadrant and mid-xiphoid without rebound.

Laboratory findings showed discretely elevated liver transaminases, sedimentation rate, and C-reactive protein.

Abdominal CT examination was performed to evaluate the patient for presence of cancer or inflammatory disease.

Abdominal CT scan showed prominence of azygos vein with absence of the suprarenal inferior vena cava (IVC) (Fig 1). The hepatic segment of the IVC was not seen and hepatic veins drained directly into the right atrium. A large hemiazygos vein was also seen (Figure 1). There was a mirrorimage location of digestive organs including Colonic segments were totally located on the left, and small intestinal segments were totally placed in the right abdomen (Fig 2).

The spleen was segmented by a fissure and multilobed consistent with polysplenia (Fig 3). Pancreatic head was extending in anterolateral direction and partially encircled part of duodenum consistent with semiannular pancreas (Fig 4). The portal vein passed ventral to the duodenum and pancreatic head, consistent with preduodenal portal vein.

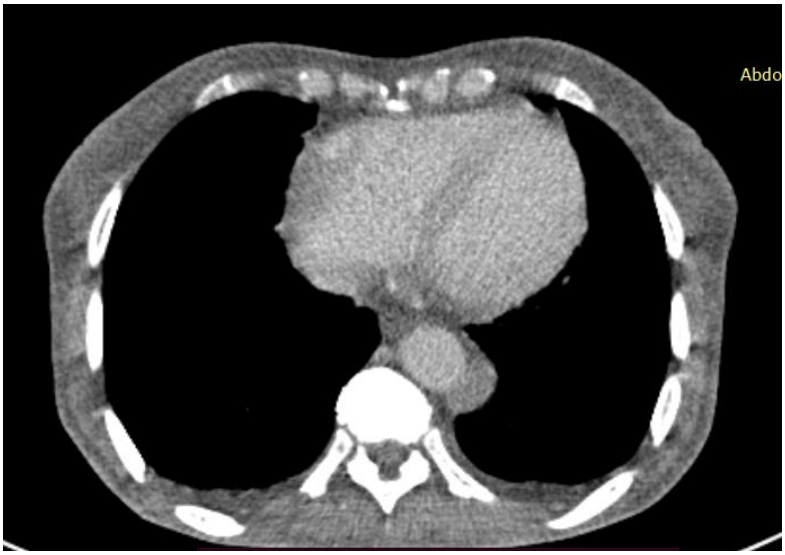

Figure 1: Absence of IVC with large hemiazygos vein

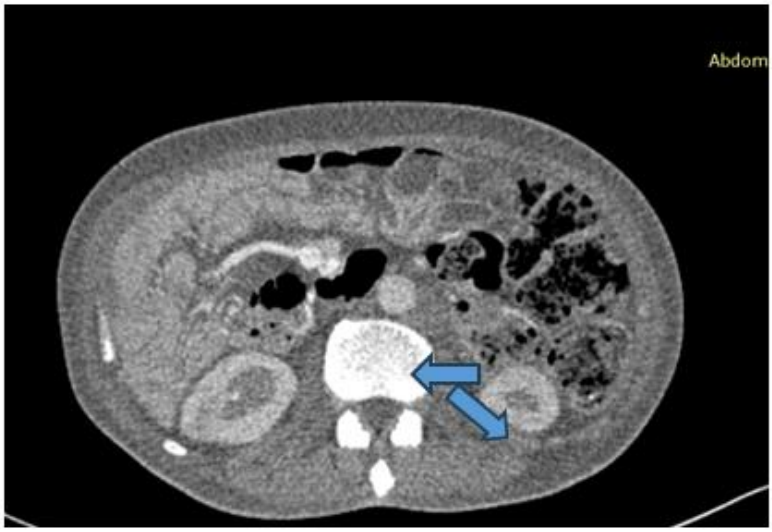

Figure 2: Mirror image of digestive organs

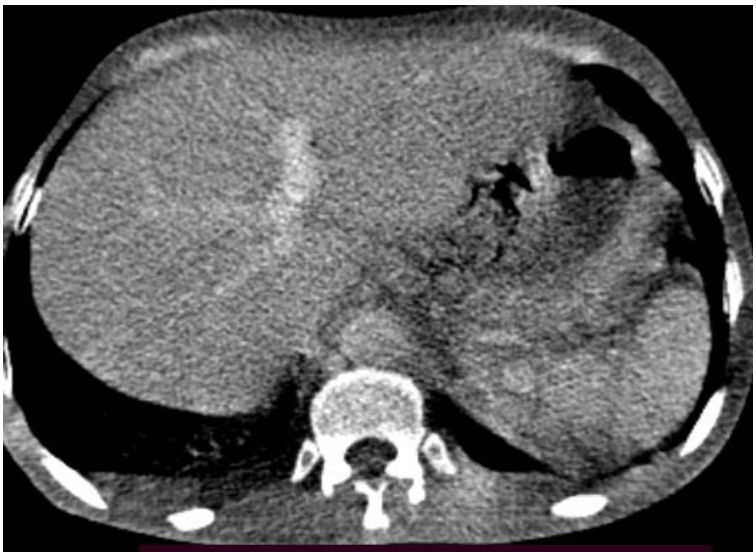

Figure 3: Multilobed left spleen

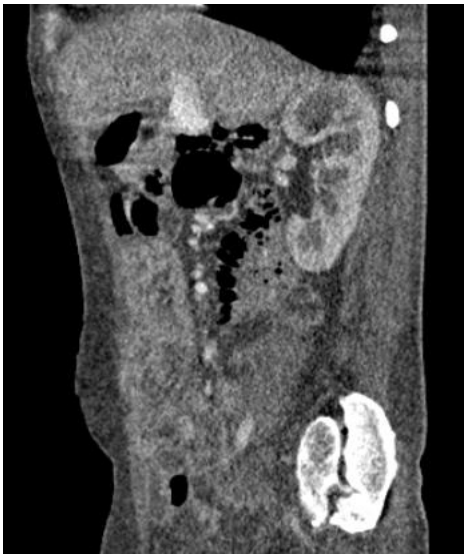

Figure 4: Semi annular pancreas

DISCUSSION

PS is a rare (about 2.5:100,000 live births) congenital disorder generally diagnosed in early childhood due to various and severe cardiac abnormalities [5]. Fifty percent of patients with PS die by 4 months of age and 75% before 5 years of age due to severe cardiovascular anomalies [1]. Patients with PS often present with atrioventricular septaldefects. Approximately 5–10% of patients with PS have normal hearts or only minor cardiac defects, and it may firstbe seen in adulthood [1].

PS is rarely encountered in theelderly [5, 6]. Adult PS cases are usually incidentally recognized on CT or magnetic resonance imaging performed while other conditions are being searched. PS has no pathognomonic anomaly; it is rather a condition of complex abnormalities. The syndrome is presented with multiple spleens associated with various anomalies such as visceral heterotaxia with a right-sided stomach, a left-sided or large midline liver, right-sided spleens, malrotation of the intestine, a short pancreas, and azygos or hemiazygos continuation of IVC with absence of the hepatic segments.

In their series, Gayer et al reviewed mainly the CT findings of eight adult cases with PS, and they reported that the most common findings in PS were multiple spleens along the greater curvature of the stomach, a left-sided IVC with azygos/hemiazygos continuation. Preduodenal portal vein, short pancreas, intestinal nonrotation, and dextrocardia were also other findings. The researchers also reviewed the CT findings of 15 adult patients described in the literature and they found similar prevalence of these anomalies [2].

Classic PS is usually accompanied with situs ambiguous likely in our case, SIT is the mirror-image location of the viscera. It is a rare congenital anomaly, inherited in an autosomal recessive manner with an incidence of 1:1400 to 1:35,000 [3, 4]. It does not usually affect life expectancy and it is not considered to be premalignant. Congenital heart disease occurs in 3–5% of the cases [7]. The recognition of this anomaly is important to avoid surgical or interventional incidents. There has been much information about the imaging features of situs anomalies in the pediatric age group because most children are evaluated clinically or radiologically due to severe congenital heart disease, immune deficiency, or bowel obstruction related to malrotation [8].

In contrast, as the conditions primarily affecting the pediatric population are not present in adults, they do not undergo imaging and radiologic information; particularly, abdominal manifestations have not been sufficiently documented [5, 9]. Situs anomalies are detected incidentally in adults investigated for conditions such as cholecystitis and appendicitis.

Moreover, the technical advances in ultrasonography, CT, and magnetic resonance imaging techniques have enabled radiologists to delineate such abnormalities.

In our case, an annular pancreas was also another accompanying finding. Annular pancreas is an uncommon congenital anomaly with a prevalence of 5–15 cases per 100,000 adults [10]. Anomalies of the pancreas have also been reported in PS. Usually, a short pancreas with agenesis of the dorsal portions is stated [2]. Maier *et al*., [11] reported an annular pancreas and agenesis of dorsal pancreas in a patient with PS. In addition, Kobayashi *et al*., [12] described a semi annular pancreas.

CONCLUSION

PS is a rare congenital anomaly that may be detected incidentally in adults, and CT can accurately detect the associated anomalies. As PS is rather a

combination of various organ abnormalities, it is important to be aware of these accompanying findings to avoid misdiagnosis of these pathological processes.

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