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IMAGES IN CLINICAL RADIOLOGY

Intra-Abdominal Abnormalities Associated with Polysplenia Syndrome

Hadrien Renier

What to look for in case of polysplenia and/or unusual disposition of several intra-abdominal organs.

Keywords: Polysplenia; Syndrome; Isomerism; Abdominopelvic Computed Tomography; Heterotaxy; Ambiguus

Anysmay, polysplenia syndrome is an unusual disposition of intra-abdominal organs and unlike situs inversus it's a spectrum of abnormalities and not a single set.

Report

An asymptomatic 62-year-old woman underwent abdominopelvic computed tomography (CT) for inaugural diabetes.

CT showed dorsal pancreatic (P) agenesis (A) (**Figure 1a**).

Incidental abnormalities were found, including:

- Four spleens (S), one adjacent to the stomach (**Figure 1b**).
- Right renal hypotrophy (**Figure 1c**).
- Midline falciform ligament (**Figure 1d**).
- Duplicated inferior vena cava system (IVC) with dilated azygos (A) and hemiazygos (H) continuation and no communication with hepatic veins (N) (**Figure 2**).
- Intestinal nonrotation: the small bowel was right-sided (S), the colon was left-sided (C), the superior mesenteric artery (A) was to the right of the vena (V), and there was no midline crossing by the duodenum (D) under the aorto-mesenteric junction (P) (**Figure 3**).

The diagnosis of type II diabetes was retained and after four months of metformin and insulin therapy, the rate of HbA1c was almost normalized. The remaining of the follow-up was unremarkable.

Comment

Heterotaxy syndrome (HS) (or situs ambiguus) is the result of an early embryological developmental failure in which there is an abnormal arrangement of thoraco-abdominal organs. In contrast to situs inversus, HS is not characterized by a single set of abnormalities but rather a spectrum.

Polysplenia syndrome (PS) (or left isomerism) is the subtype of HS with features of bilateral left-sidedness. No single anomaly is pathognomonic but the association of a sufficient number allows the diagnosis. The commonest is the presence of multiple spleens, right- or left-sided, with a consistent relationship to the stomach.

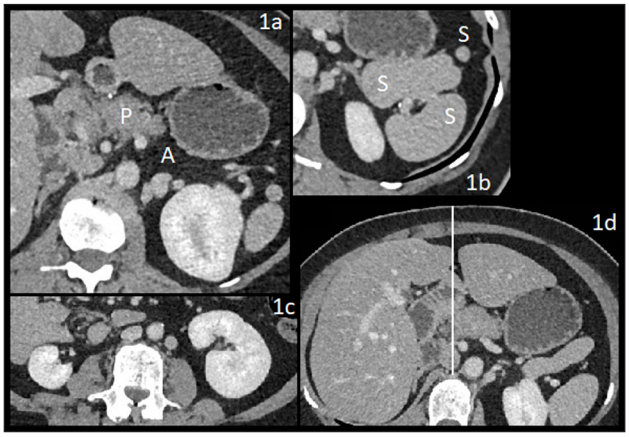

Figure 1.

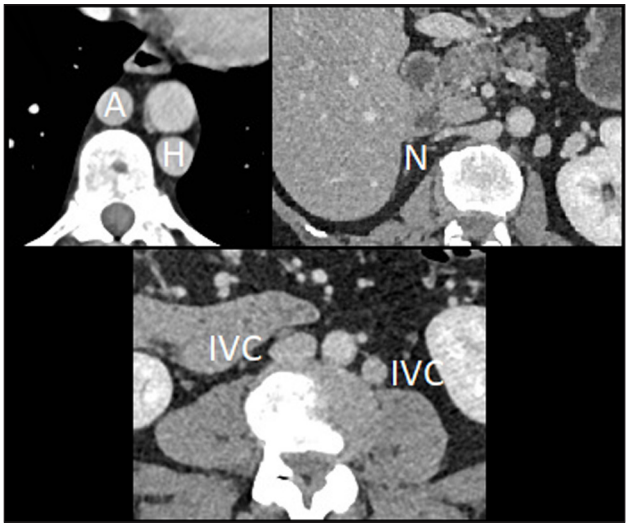

Figure 2.

Université de Liège, BE
hadrienrenier@gmail.com

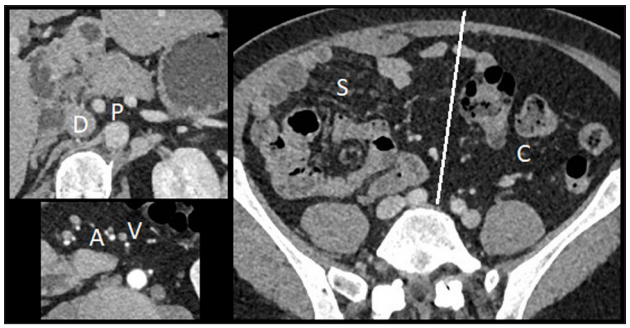

Figure 3.

As in the present case, the other intra-abdominal abnormalities include:

- midline liver with or without biliary abnormality,
- truncated pancreas with presence of the head and a variable portion of the body,
- azygos continuation of the IVC,
- midline or right-sided aorta,
- right-sided stomach and/or abnormalities of the mesentery rotation.

Compared to the other HS (i.e., right isomerism [or asplenia]) PS is often detected incidentally in adults. Indeed, it is associated with less severe or no congenital heart disease and no immune system deficiency [1].

Competing Interests

The author has no competing interests to declare.

Reference

1. **Fulcher, AS** and **Turner, MA.** Abdominal manifestations of situs anomalies in adults. *Radiographics.* 2002; 22(6): 1439–56. DOI: https://doi.org/10.1148/rg.226025016

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