

Heterotaxy Polysplenia Syndrome in an Old Female

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A 74-year-old woman, with no medical history, presented to the emergency room with epigastric pain for three days. On physical examination, vital signs were all within normal limit, and abdominal palpation revealed epigastric tenderness. Chest and abdominal CT scan was performed and showed typical findings of COVID-19 pneumonia, affecting 20% of lung parenchyma (Fig. 1). CT scan also showed multiple splenic nodules (about eight) in different sizes ranging from

3 to 6,5 cm (Fig. 2), the superior mesenteric artery (SMA) was to the right of the superior mesenteric vena (SMV) (Fig. 3), the small bowel was right-sided and the colon was left-sided, dilated azygos vein with agenesis of the inferior vena cava (IVC) and agenesis of the dorsal pancreas (Fig. 3). The patient was hospitalized in a Covid-19 unit. She was discharged after six days of oxygen therapy and medical treatment.



Fig. 1: Chest computed tomography showing COVID-19 pneumonia, affecting 20% of lung parenchyma

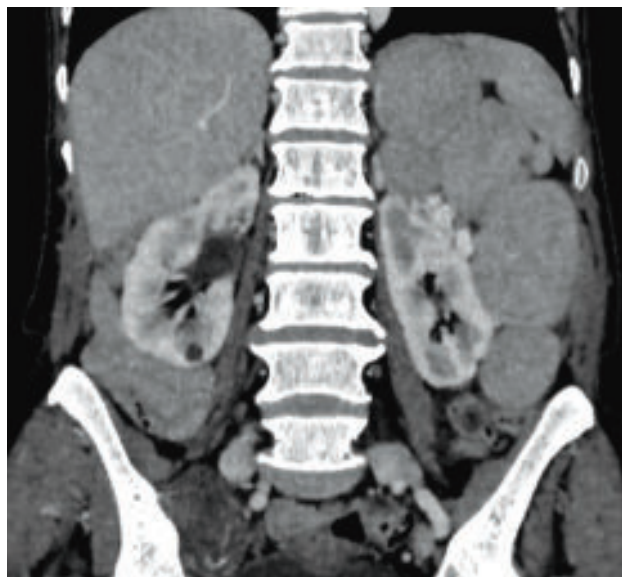


Fig. 2: Abdominal computed tomography showing presence of multiple spleens in different sizes

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Heterotaxy syndrome (HS) (or situs ambiguus) is a rare condition occurring in approximately 1/250,000 births, in which there is an abnormal arrangement of thoracoabdominal organs.¹ It's the result of an early embryological developmental failure.² Polysplenia syndrome (PS) (or left isomerism) is the subtype of HS with features of bilateral left-sidedness. As in our case, typical findings are: presence of multiple spleens, truncated pancreas with agenesis of the dorsal pancreas, interrupted IVC with azygous continuation and intestinal non rotation with common mesentery. It can

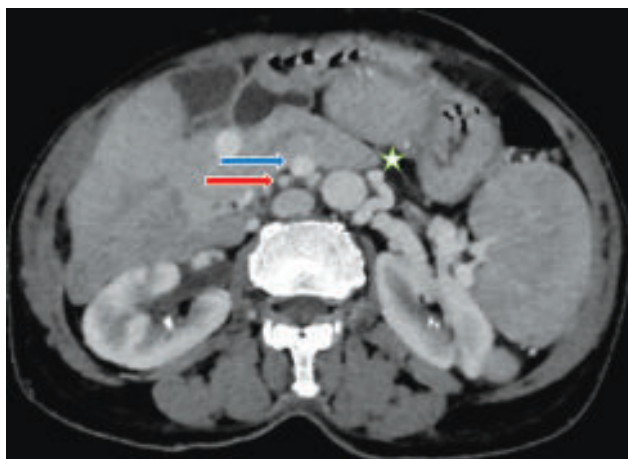


Fig. 3: Abdominal computed tomography showing agenesis of the dorsal pancreas (Asterisk) with inversion of superior mesenteric vessels (SMA (red arrow) to the right of the VMA (blue arrow))

also include a midline liver with or without biliary abnormality and a midline or right-sided aorta.³

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