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Polysplenia Syndrome: A Rare Condition Discovered Incidentally

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Abstract

The Polysplenia Syndrome associates, in addition to the presence of several spleens, abnormalities falling within the framework of heterotaxy (situs inverses, pre-duodenal portal vein, absence of retro-hepatic IVC, Abnormal Hepatic Vascularization and intestinal malrotation). Case of a 32-year-old asthmatic patient who seeks therapy for abdominal pain and diarrhoea. Upon examination, it was discovered that the right iliac fossa and hypogastrium of the afebrile patient were tender. Acute appendicitis was thought to be the cause. The range of aberrant rotation of the thoraco-abdominal organs in Polysplenia Syndrome, a rare congenital disorder, differs from patient to patient. This syndrome's origin has not yet been determined.

Keywords: Polysplenia syndrome • Heterotaxy • Common mesentery • Splenic cysts • IVC interruption

Background

The Polysplenia syndrome associates, in addition to the presence of several spleens, abnormalities falling within the framework of heterotaxy (situs inverses, pre-duodenal portal vein, absence of retro-hepatic IVC, Abnormal Hepatic Vascularization and intestinal malrotation) [1]. The diagnosis is often made during the neonatal period. Except that the diagnosis can linger and can be made in adulthood.

Case Presentation

We report the case of a 32-year-old patient, asthmatic under treatment, who consults for abdominal pain with diarrhea. On examination, an afebrile patient was found with tenderness in the right iliac fossa and hypogastrium. The diagnosis of acute appendicitis was suspected. An abdominopelvic CT scan showed an internal latero-caecal appendage with a non-swollen, ventilated descending course, multiple splenic nodules located in the splenic compartment (approximately 7) including the largest measuring

56 x 41 mm. The pancreas is short (atrophic caudal part) with homogeneous enhancement. The small loops are lateralized on the right and the colonic frame is on the left. The superior mesenteric artery is located posterior to the superior mesenteric vein. Atresia of the retro-hepatic inferior vena cava with azygous continuation. The adrenals and kidneys appear normal. A syndrome of Polysplenia was evoked and no therapeutic management was indicated in this patient for these malformations' abnormalities discovered fortuitously. She was put on symptomatic treatment.

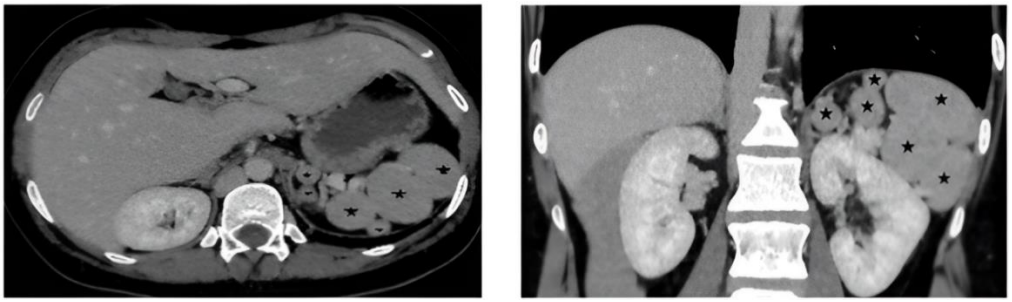

Figure 1a, 1b. Axial and coronal slice showing multiple splenic nodules (stars).

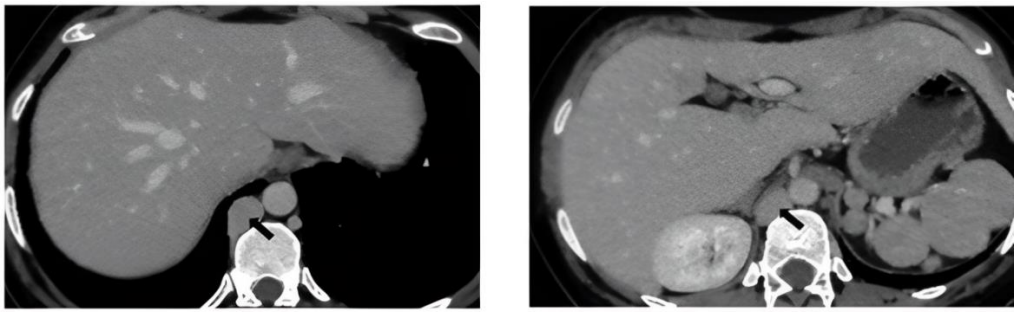

Figure 2. Axial sections showing atresia of the retro hepatic inferior vena cava with azygous continuation (arrows).

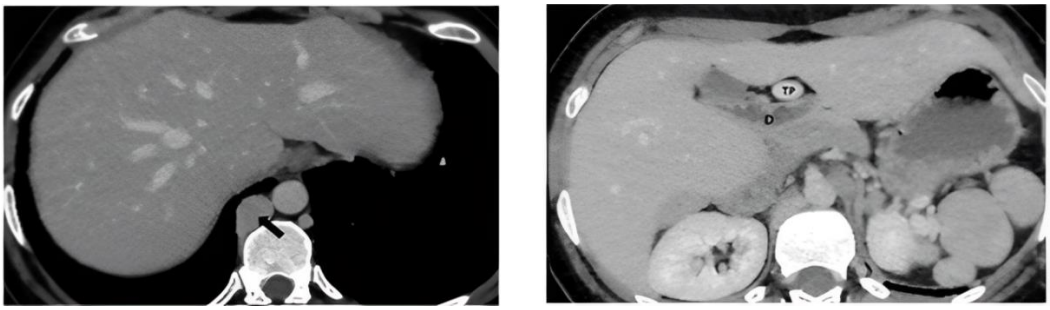

Figure 3a. Axial slice showing atrophy of the caudal portion of the pancreas: "Short pancreas", 3b.Pre-duodenal portal vein
TP: Door Trunk; D: Duodenum

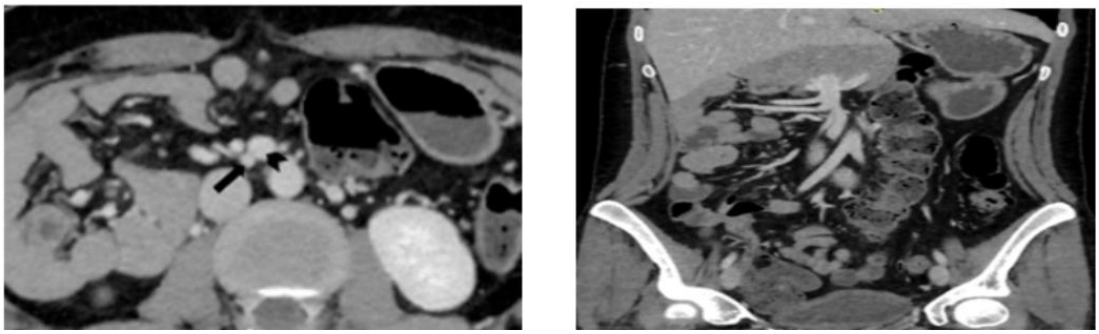

Figure 4a. Complete common mesentery: Superior mesenteric vein (arrowhead) located to the left of the superior mesenteric artery (arrow), 4b. Complete common mesentery: colonic framework located on the left and the small loops on the right.

Discussion

Heterotaxis syndrome is a set of globally symmetrical hereditary malposition with respect to the Z axis, of the abdominal, and thoracic viscera. Its prevalence is 1 to 9/100,000 and transmission is autosomal dominant or autosomal recessive or X-linked recessive. It most often appears in early childhood (manifestation of heart damage).Its incidence is of the order of 1/250,000 new-borns [2]. Abdominal anomalies are as a rule asymptomatic, unlike cardiac anomalies which are frequent. [3,4] These cardiac abnormalities are the cause of increased mortality at young age. Thanks to the advent and development of means of cross-sectional imaging, a few rare cases of fortuitously discovered Polysplenia have been described. Splenic malformations: can present in the form of asplenia which corresponds to a defect of lateralization with small spleen or absent spleen, or in the form of Polysplenia which corresponds to a lateralization defect with the presence of multiple small spleens whose size varies between 1 and 6cm.[4,7,8] Hepatobiliary malformations: which correspond to a median liver with a median gallbladder Pancreatic malformations: Embryologically, the pancreas is the result of the fusion of the ventral and dorsal pancreatic buds. The ventral bud produces the uncus and head, while the dorsal bud produces the body and tail. In addition, the development of the dorsal pancreatic bud and the spleen occur in the dorsal midgut hence the coexistence of abnormalities of the spleen and pancreas in patients with Polysplenia syndrome [12].Generally, the pancreas is short rounded or oval in shape [4.5]Vascular anomalies: which include a pre-duodenal portal vein, the azygous continuation with atresia of the retro-hepatic inferior vena cava, and arterial variations such as several accessory splenic arteries which arise directly from the aorta and a trunk Coelio-mesenteric joint [6.9]. To understand the anomalies of the VCI, it is necessary to know that this vena cava develops as being the fusion of 4 parts: a hepatic part which forms from the vitelline vein, a renal part which develops from the fusion supra - right sub-cardinals and post-sub-cardinals, an adrenal part which is formed by the right sub-cardinal vein and the hepatic sub-cardinal anastomosis and finally a sub-renal part which is formed from the right supra-cardinal vein [12]. Anomalies of the rotation of the digestive tract: which include dextrogastria, the complete and incomplete common mesentery with anomalies of rotation of the mesenteric vessels [9] Extra-digestive abnormalities: pulmonary (left isomerism of the bronchial tree, etc.) [10], renal (renal agenesis, etc.), nervous (cervical myelomeningocele, etc.) [11]. Within the same family, the severity of the malformations is very variable. A complete situsin versus is not in itself a source of problem. In contrast, incomplete situsin versus can be associated with any type of heart defects, as well as renal, biliary, midline, etc. abnormalities. Cardiac involvement is most often early, especially in the group of Heterotaxis with asplenia, and minimal and asymptomatic in Heterotaxis with Polysplenia, hence its often-fortuitous discovery in adults, during an imaging assessment [3] Only 5 to 10% of carriers of this syndrome reach adulthood. The variable involvement of these different organs results in a large clinical polymorphism. In the case of acquired pathologies, the symptomatology may be modified, for example appendicitis, cholecystitis and sigmoid diverticulitis may be more difficult to diagnose due to the unusual localization of the symptoms.

Conclusion

Polysplenia syndrome is a rare congenital condition with a spectrum of abnormal rotation of thoraco-abdominal organs that varies from one patient to another. The cause of this syndrome is not yet established. Indeed, a teratogenic, embryological and genetic component have been incriminated. The diagnosis is often made during the first months of life. Its treatment is based on the prevention of the evolution of certain abnormalities, especially biliary atresia which evolves towards cirrhosis.

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