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Bilateral Thrombophlebitis Revealing an Agenesis of the Inferior Vena Cava as Part of Polysplenum Syndrome

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Abstract **Case Report**

Polysplenum syndrome is a rare malformation characterized by the association of multiple spleen with other abnormalities dominated by cardiovascular and visceral malformations. We report the observation of a young patient admitted to the emergency room for a clinical presentation of acute appendicitis. The abdominal ultrasound scan had invalidated the diagnosis and pushed to perform an abdominopelvian computed tomography detecting a bilateral iliac acute thrombophlebitis with agenesis of the inferior vena cava supplemented by a large azygos vein, short pancreas and multiple splenic lobules.

Keywords: Polysplenum syndrome, Thrombophlebitis, Agenesis of the inferior vena.

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INTRODUCTION

Polysplenum syndrome refers to a set of congenital abnormalities classically associating a number of spleen greater than or equal to two to other malformations the most common are cardiovascular and visceral [1, 2]. It is a rare condition with a variable frequency [1]. The association with a situs inversus anomaly is noted in half of cases and in 30 to 40% of cases associated with cardiac abnormalities [2-4].

CASE REPORT

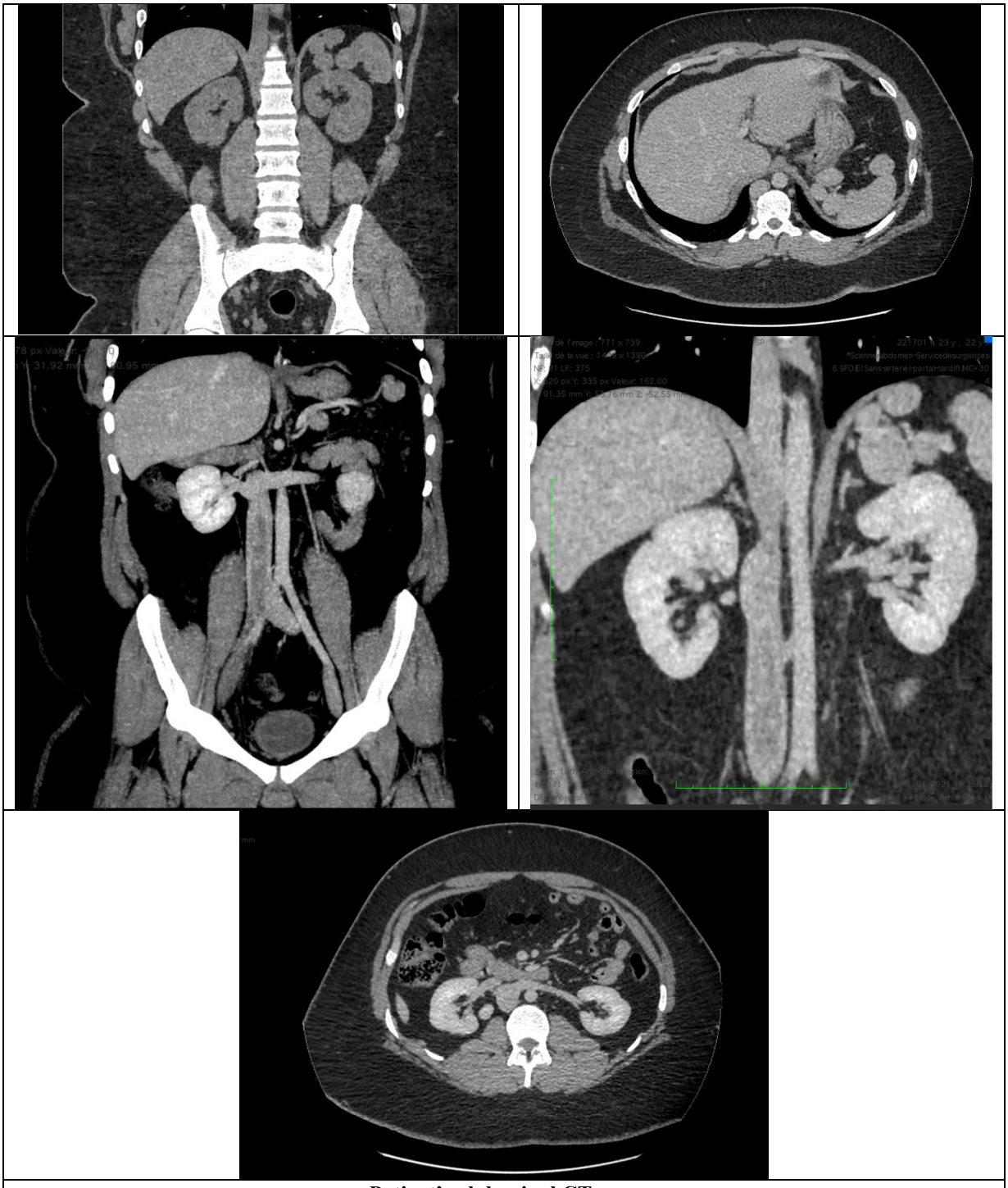
A 37-year-old man with chronic active tobacco use, sedentary and followed for morbid obesity, admitted to the emergency department for acute abdominal pain localized to the right iliac fossa going back 48 hours and evolving in a febrile context. The clinical examination regains a sensitivity of the right iliac fossa. The rest of the physical examination is without abnormality.

An abdominal ultrasound was performed retrieving an appendix with normal size, not infiltrated, without effusion or underlying abscess.

In front of the noisy clinical picture, a complement by abdominal computed tomography was performed detecting: A visible sub-hepatic appendix without infiltration of peri-appendicular fat. Ascent of the caecum which is in the sub-hepatic position.

Multiple peri-splenic nodules having the same enhancement kinetics as the spleen in relation to a splenic pulley.

A complete agenesis of the inferior vena cava supplemented by a voluminous azygos vein. Infiltration of the inferior vena cava extended to the iliac veins and the superficial right femoral vein. A short pancreas. An incomplete common mesentery.



Patient's abdominal CT scan

All these abnormalities constitute the polysplenum syndrome associating a splenic pulley, a short pancreas, a malposition of the caecum, an incomplete common mesentery with agenesis of the inferior vena cava substituted by a voluminous azygos vein.

A vascular doppler ultrasound of the right lower limb detected iliac thrombophlebitis extended to the superficial femoral vein.

In total, this is a young patient with extensive vena cava thrombophlebitis with atypical clinical presentation; abdominal pain with a normal right lower limb in which polysplenum syndrome was accidentally discovered at abdominal computed tomography.

The patient was hospitalized in the department of cardiology and vascular diseases for surveillance, thrombophilia check-up, investigation of associated cardiac abnormalities and introduction of anticoagulation.

DISCUSSION

Polysplenum syndrome is a rare heterotaxy. It is characterized by an abnormal disposition of one or more viscera responsible for clinical polymorphism. Heart disease is quite common. It is more complex and early in the group of heterotaxies with asplenia, often minimal and asymptomatic in heterotaxies with polysplena, hence its discovery often fortuitous in adults, during an imaging assessment for example as in our patient [5-7].

The diagnosis of polysplenary syndrome is often made during the first months of life, this depends on the type of malformations associated. Antenatal diagnosis is possible and is based on the contribution of obstetric ultrasound. This exam allows as to visualize elements of polysplenum syndrome [8-10]. All the abnormalities encountered in this condition include:

Cardiovascular involvement in the form of left atrial isomerism, an anomaly of systemic venous return to the retrohepatic inferior vena cava with continuation by azygos which flows into the superior vena cava. The hepatic veins flow either directly into the right atrium or through an intrathoracic vena cava stump. The embryological development of this anomaly is explained by the absence of fusion of the pre-renal and hepatic segments of the inferior vena cava with segmental hypoplasia of its suprarenal portion. Its sub-renal portion then drains through the azygos system.

Abdominal organ damage in the form of multiple spleen located in the left or right hypochondria depending on whether or not there is an associated gastric rotation abnormality.

The liver may be normal in the right hypochondria or abnormal in the left or middle hypochondria with an interposed gallbladder between the two hepatic lobes or sometimes absent [5, 6, 12-15]

The pancreas can be either normal morphology, short with presence only of its cephalic or annular portion. This pancreatic malformation is due to agenesis of the dorsal pancreas linked to a lack of blood supply during fetal life by secondary diversion to the presence of multiple rats [7, 12, 16].

Finally, a few rare cases of malformations of the central nervous system have been reported in the literature with the type of agenesis of the corpus callosum, holoprosencephalia, and myelogengocele [16, 17]. Imaging easily diagnoses all malformations of this congenital condition. It is based on cross-sectional imaging techniques. Chest radiography may suspect the diagnosis by showing dilatation of the azygos vein in the form of a displacement of the right paravertebral line and a protrusion of the azygos and locating the gastric air pocket. Abdominal ultrasound can show morphological and rotation abnormalities. Doppler can show vascular abnormalities. The CT remains an excellent tool of diagnosis and above all a more complete assessment of all thoracic-abdominal abnormalities, especially the vascular and digestive ones [5, 7, 13-15].

CONCLUSION

Polysplenum syndrome is a rare condition that we must think about it in front of any bilateral thrombophlebitis of the lower limbs, its morbidity remains important compared to the associated malformations especially cardiac, vascular and neurologic which can sometimes engage the vital prognosis hence the importance of a systematic assessment.

Conflicts of Interest: The authors declare no financial interest nor any other conflict of interest

Informed Conscent: The patient conscented

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