ANATOMIC VARIATIONS

Asymptomatic left isomerism with preduodenal portal vein: computed tomography appearance

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Abstract
Left isomerism (polysplenia), one of the two major variants of heterotaxia with right isomerism (asplenia), may be rarely diagnosed in adulthood. Most cases are nevertheless asymptomatic and incidentally detected during imaging or surgery performed for unrelated conditions. We hereby report a case of left isomerism fortuitously diagnosed in a 55-year-old man with unrelated tachy-cardiomyopathy. Thoraco-abdominal computed tomography revealed a typical preduodenal portal vein (PDPV) associated with a large series of other occult anatomic variations comprising: polysplenia, agenesis of both pancreatic body and tail, complete non-rotation of the bowel and finally azygous continuation of the inferior vena cava. Subtle but highly specific thoracic features of left isomerism were also found with a bilobed right lung and bilateral long hyparterial main bronchi. The features of adult left isomerism are remembered with special attention to the PDPV.

Keywords Heterotaxia syndrome · Left isomerism · Polysplenia · Preduodenal portal vein · IVC abnormality · Pancreatic agenesis · Situs ambiguous · Intestinal malrotation

Introduction
During embryology, a series of thoraco-abdominal unpaired organs follow a specific spatial organization. There are typical right-sided structures (liver, superior and inferior vena cava, right atrium and its appendage, trilobed right lung with epi-arterial bronchus, etc.) and typical left-sided structures (stomach, spleen, left atrium and appendage, pulmonary veins and bilobed left lung with hyparterial superior lobar bronchus, etc.).

The normal final positioning refers to *situs solitus* (with thoracic levocardia) and the complete mirror-image arrangement refers to *situs inversus*, a rare but commonly asymptomatic situation [6].

*Situs ambiguous*, more commonly also named *heterotaxia* refers to anatomic arrangements in which the major organs are distributed abnormally within the chest and/or abdomen [2, 5, 6]. This kind of variation may be isolated or associated with an extensive collection of complex thoracic and/or abdominal anomalies [6]. The prevalence of HS is estimated to be 1 in 10,000 people worldwide but the condition appears probably largely underdiagnosed. There are two main variants of *heterotaxia: left isomerism* (LI) also known as polysplenia or bilateral left sidedness and *right isomerism* (RI) also known as asplenia or bilateral right sidedness [2, 5, 6].

Clinical expression of HS variants depends on the gravity of the abnormalities involved. HS may be life threatening in infancy or childhood in a high proportion of cases. This is essentially due to severe cardiac abnormalities particularly in the RI variant [6]. Rare individual adults, especially having an LI, may nevertheless present only mild health disturbances or may be completely asymptomatic. We hereby report a case of LI fortuitously diagnosed in a 55-year-old man with unrelated tachy-cardiomyopathy.

Case report
A 55-year-old patient (144 kg/1.7 m) was admitted with complaints of dyspnea, abdominal ascitic distention, anaasarca and progressive worsening of oedema of the lower limbs. In the health context of COVID-19, thoraco-abdominal computed tomography (CTA) was performed not only to exclude COVID-19 pneumonia and/or pulmonary embolism...
but also to evaluate the etiology of ascitis. Pulmonary embolism was excluded but a large azygous continuation of the inferior vena cava (of which only a short supra-hepatic segment remained visible) was seen (Fig. 1a–c). A large collection of abdominal anatomic variations was also found comprising: an unusual preduodenal portal vein (PDPV) (Fig. 1d, e), two spleens nested one inside the other (poly-splenia), the absence of pancreatic body and tail (Fig. 2a, b), a complete non-rotation of the bowel (Fig. 2c), an undescended epigastric cecum with an epigastric appendix (Fig. 2d). Scrupulous retrospective analysis of the thoracic CT revealed a bilobed right lung (Fig. 2f) with an atypical “z”-shaped scissure and bilateral hyparterial long main bronchi.

The patient was finally diagnosed having a completely asymptomatic LI (polysplenia). It was a fortuitous diagnosis superimposing on symptomatic cardiac decompensation caused by impaired systolic function and atrial fibrillation, both generating secondary tachy-cardiomyopathy. The patient was successfully medically treated.

Discussion

LI and RI are the main variants of HS. Unlike situs abnormalities leading to ordinary or reverse lateralization, isomerism is accompanied by an atypical anatomy with symmetrisation of the organs supposed to be lateralized or even duplication of organs in certain circumstances [2, 5, 6].

RI is the most frequently variant observed at birth. In this situation, there are two atria of the systemic right type and the two lungs of the right type. Both lungs are thus trilobed with bilateral minor fissures and their main bronchi are short and located superior to the ipsilateral main
pulmonary artery (epi-arterial bronchi). Asplenia is associated in 80% of cases. The liver may be centrally located and the stomach in an indeterminate position. Major and very severe cardiac abnormalities are associated in nearly 100% of patients (transposition of the great vessels, tetralogy of Fallot or atrioventricular duct type…) determining a very poor neonatal prognosis. Survival beyond 1 year of life is rare, ranging from less than 5–20%. RI may rarely exist without cardiac anomalies and may exceptionally remain asymptomatic until adulthood [2, 5–8].

In LI as reported in this case, the lungs are of the left type. They are thus bilobed and their main bronchi are long, passing inferiorly to the ipsilateral main pulmonary artery PA (hypo-arterial bronchi). LI is classically associated with both atria of the pulmonary type, a midline liver. A high prevalence of polysplenia is typical and explains the former but still used term of “polysplenia”. Cardiac abnormalities are less common and less complex than in RI. As a consequence, the death rate is lower (60% in the first year) than in RI. Only about 10% of patients may reach adulthood without any complications.

At birth, LI isomerism is more common in females but the exact incidence and sex ratio in adults is more difficult to estimate because, as is the reported case, many adult cases are only incidentally detected during imaging evaluation for unrelated conditions. Minimal cases presenting with only bilobed lungs but a normal abdominal situs and without cardiac abnormality have also been reported [1, 2, 5–9]. LI may also present with other variable symptoms related to a large spectrum of other associated variations. There is no single
pathognomonic abnormality that characterizes LI [1]. In the abdomen, polysplenia, a short pancreas, abnormalities of the portal vein formation, various types of gut malrotations and inferior vena cava anomalies are the most common associated anomalies [1, 2, 7–9] apart from the specific thoracic variations:

- Polysplenia is characterized by the presence of several spleens, usually between two and six. They are about of equal size, associated with the main spleen in the left hypochondrium and vascularized by an artery with normal birth. Polysplenia must be differentiated from supernumerary spleens (which generally measure only 1–2 cm) and from splenosis [3].

- Congenital preduodenal portal vein (PDPV), often present (75%) in LI, is characterized by a portal vein passing in front of the duodenum [3, 7–11]. PDPV is rare as an isolated variation (25%) [4, 11]. In childhood, PDPV may be associated with features of intestinal obstruction caused by coexisting congenital anomalies such as duodenal atresia, duodenal web, annular pancreas, intestinal malformation, biliary atresia, etc. [7]. Surgery may be required for these obstructive conditions. Duodenal obstruction caused by PDPV alone is a very rare situation with only a few reported cases. The reason is probably the fact that PDPV is a thin-walled vessel under low pressure [3, 11]. As reported in our case, patients with PDVP may reach adulthood without any symptoms, the anomaly being often found during examinations or surgery for unrelated conditions. PDPV develops between the 5th and 10th embryonal weeks [11] when the paired primitive vitelline veins loss their left cranial, right distal and middle posterior communicating segments (Fig. 2fb). In normal development, the normal ‘S’-shaped retroduodenal portal vein rather results from the loss of the right caudal, left cranial and anterior caudal communicating segments of the vitelline veins (Fig. 2fc) [8]. An occult PDPV not detected prior to biliary surgery may be responsible of severe complications, such as hemorrhage or accidental vascular ligation. Such accidents can be prevented by performing previous careful diagnostic imaging and especially by searching and noting the possibility of PDPV in cases of polysplenia syndrome.

- In 25% of cases, but not in this case, a median liver is present in LI (but also in RI) with two symmetrical hepatic lobes and a gallbladder interposed between them [3].

- Variations of the digestive tract associated with LI may comprise an intraperitoneal annular pancreas or sometimes, as illustrated in our case, a short pancreas with agenesis of the corpus and tail [3, 8]. Pancreas formation results from fusion of ventral and dorsal pancreatic buds. The ventral bud gives rise to the head and unci-
Declarations

Conflict of interest The author declare that he has no conflicts of interest concerning this article.

Ethical approval The manuscript follows the prescriptions of the institutional ethical board. The author certify that all data and figures have been completely anonymised.

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