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Polysplenia with agenesis of the dorsal pancreas and preduodenal portal vein, about a case

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ABSTRACT

PURPOSE OF THE STUDY: Polysplenia is a complex polymalformative syndrome that is frequently accompanied by an inconsistent spectrum of visceral and vascular anatomical abnormalities and is extremely undiscovered in adulthood.

The objective of this article is to limit the intraoperative risks generated by the lack of knowledge of these anatomical variations by insisting on the inconstancy of all these variations and the perfect knowledge of the reference anatomy and the exploration of the anatomical variations in imaging before the surgery.

PATIENT AND METHODS: The patient was 50-year-old who was hospitalized in our department for gastric adenocarcinoma. During her extension assessment, a polysplenia syndrome was accidentally discovered on the scan images, which showed an exceptional association between preduodenal portal vein, agenesis of the dorsal pancreas, and polysplenia.

CONCLUSION: There are neither specific clinical symptoms of the polysplenia syndrome, nor any biological sign; hence, the interest of recognizing this pathology is to avoid diagnostic errors, but also to guide the surgeon during the surgical act performed.

1. Introduction

Polysplenia is a rare polymalformative syndrome characterized by the presence of multiple spleens variably associated with one of the following abnormalities:

Cardiopulmonary, Vascular, Digestive, Hepato-biliary, Pancreatic, Genitourinary Central Nervous System [1].

However, some cases of polysplenia syndrome (PS) have been described with a single polylobed spleen or a single normal splenic gland [2].

The main vascular abnormalities include agenesis of the suprarenal inferior vena cava (IVC) with azygos continuity and direct drainage of hepatic veins in the right atrium and a position of the portal vein (PV) anterior to the duodenum. Biliary abnormalities are represented by biliary atresia (BA) present in 50% of cases. Visceral abnormalities may include an ambiguous situs or heterotactic syndrome with a centrally located liver (straddling the midline), a common mesentery, and partial or complete agenesis of the dorsal pancreas. The main cardiac abnormalities are the transposition of the large vessels and atrial and/or interventricular communication.

We will illustrate the diversity of morphotypes observed by presenting a case of PS diagnosed in adulthood incidentally during the extension assessment for a patient operated on in our department for gastric adenocarcinoma. This case has been reported in line with the SCARE criteria [3].

2. Presentation of case

2.1. Case history

The 50-year-old patient had no medication history or surgical history, and no relevant family history, she had been presented for epigastric pain with postprandial vomiting that has been progressing for 3 months prior to her hospitalization, without any of digestive hemorrhages externalized and transit disorder, the whole evolving in a context of alteration of the general state and weight

Abbreviations: PS, polysplenia syndrome; IVC, inferior vena cava; BA, biliary atresia; PV, portal vein.

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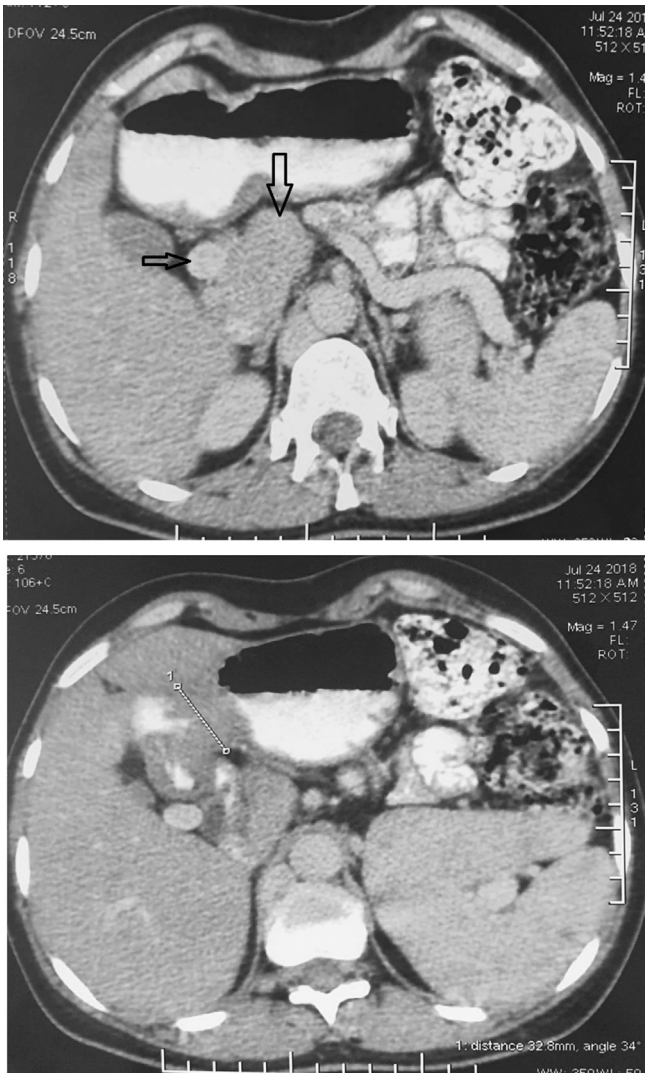

Fig. 1. (a) Axial scan section after injection of the iodinated contrast medium showing a short pancreas with a preduodenal portal vein situation.
(b) Axial scan section after injection of iodinated contrast material objectivizing the antropyloric tumor with the presence of a bilobed spleen.

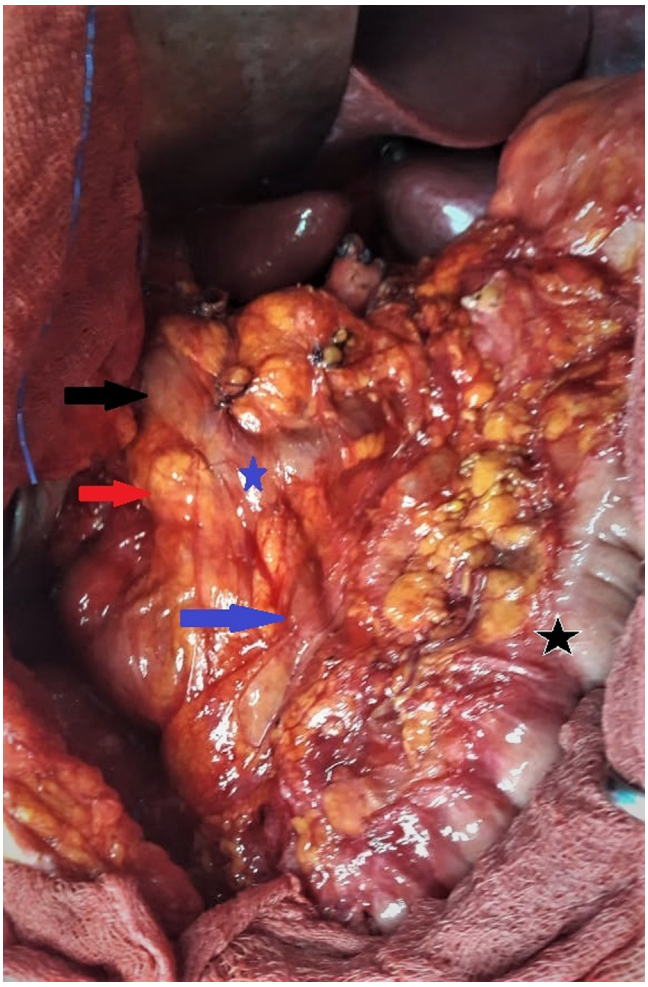

Fig. 2. Peroperative image after total gastrectomy showing a portal vein (black arrow) with a pre-duodenopancreatic trajectory with a short pancreas (red arrow) and an agenesis of its dorsal portion.

★ : transverse colon ★ : the portovenous confluence

➡ : Inferior mesenteric vein

loss ; the clinical examination found a conscious patient, with slightly discolored conjunctiva her BMI is 24. 84 kg/m^2; abdominal examination revealed a soft abdomen without any palpable mass; examination of the lymph node areas was free.

Esogastroduodenal fibroscopy revealed an antro-pyloric stenosis thickening; histological examination showed a well-differentiated adenocarcinoma.

A Thoraco-abdominopelvic CT scan showed irregular pyloric tissue thickening, measuring 22 mm thick extended over 80 mm to the cardia.

Also, a poly malformative syndrome with a bilobed spleen was found to be associated with a portal vein located in the preduodenal; its course is abnormally distant from the aorta and the AMS and a dysmorphic pancreas with agenesis of its corporal-caudal portion. Fig. 1. (a; b).

The thoracic stage did not object to any detectable malformations and the cardiac ultrasound did not reveal any deformities.

2.2. Operative techniques

A total gastrectomy with Roux -en- Y- oesophago-jejunostomy reconstruction was performed. Surgical exploration confirmed the CT scan data for poly malformative syndrome by the presence of a bilobed spleen associated with a short pancreas with corporo-caudal agenesis with a portal vein in a pre duodenopancreatic situation (Fig. 2).

Histopathological examination of the specimen showed an invasive adenocarcinoma with independent cells invading the subserosa, with the presence of a single metastatic regional lymph node. The tumor was classified as pT3N1M0.

The postoperative sequelae were simple and the patient was discharged on the sixth post-operative day, then she was referred to oncology where she received 6 courses of chemotherapy. Follow-up at two years did not reveal any tumour recurrence.

3. Discussion

Polysplenia syndrome (PS) is defined by the presence of several spleens, usually between two and six. This is a major difference between polysplenia and supernumerary spleens. The spleens in the polysplenia syndrome are always of equal size and are associated with the main spleen in the left hypochondrium, which is vascularized by an artery with normal birth. On the other hand,

accessory spleens generally measuring 1–2 cm are not part of this syndrome. Another diagnosis of elimination is splenosis, which belongs to neonatal acquired abnormalities.

In the context of malformations of the digestive tract associated with the polysplenia syndrome, we cite the annular pancreas with intraperitoneal localization or sometimes a short pancreas characterized by agenesis of the tail of the pancreas [4] which is the case of our patient; For Sener : It would be agenesis of the dorsal bud related to a defect of blood supply during intrauterine life by hemodiversion secondary to the presence of multiple spleens [5]

A pre-duodenal portal vein is frequently found; it was first described by Knight in 1921 [6] Polysplenia associated with this anomaly has been reported about ten times since then [7]; it is important in practice to inform the surgeon of this pre-duodenal portal vein. This complicates the operating procedure in visceral abdominal surgery and lack of knowledge about it can lead to hemorrhagic or thrombotic complications [8,9].

During the surgical management of our patient, it was, therefore, important to have a perfect knowledge of the reference anatomy and to have explored the anatomical variations in imaging, to limit the perioperative risks that could be encountered during the gastrectomy, particularly during the adapted lymph node dissection, which implies a good knowledge of the anatomy of the abdominal vessels, which form our landmarks guiding the carcinological excision.

The first case of PS was described in 1781, Peoples et al. [10]. performed a series of autopsies on 146 cases of PS to evaluate the frequency of the most frequently found abnormalities. In 58% of the patients had bilateral bilobed lungs with left-sided bronchial segmentation, 56% had gastrointestinal positional abnormalities, 47% had bilateral superior vena cava and more than 60% had cardiac abnormalities [2].

The hepatobiliary abnormalities are characterized by the discovery in 25% of a liver of unusual morphology composed of two symmetrical lobes concerning the median line called a median liver, with gallbladder interposed between the two lobes or agenesis in rare cases.

The digestive abnormalities are represented by an ambiguous situs, by a common mesentery on the right more or less complete with the intestinal inlets and the colonic loop on the left imposing an outlet of the last ileal loop on the right edge of the cecum [11]; by dextrogastria; gastric duplication; esophageal atresia; by jejunal digestive stenoses; ileal or duodenal stenoses, by a micro colon [4].

The most frequent cardiac abnormalities are represented by a defect of the interventricular or interauricular septum, by a common atrioventricular canal, by a transposition of the large vessels, more rarely by the persistence of the arterial trunk, by a partial connection of the pulmonary veins, by pulmonary arteriovenous malformations, by a double exit of the left ventricle and by a common atrium [12,13].

Vascular malformations are represented primarily by abnormalities of the inferior vena cava return reported by Peoples in 65% [10]; it is the absence of hepatic segment of VCI, which causes a continuation azygos [14,15]. The first abnormality was reported in the mid-19th century and in 1951; Effler et al. diagnosed an interruption of IVC with azygos continuation when a patient died after azygos ligation during thoracic surgery [16].

Variations in the arterial branches of the abdominal aorta are also described [17] essentially consisting of a common celiomesenteric trunk, multiple accessory splenic arteries originating directly from the aorta. Finally, heart disease is present in 85–90% of cases [18,19].

Only 5%–10% of patients with this syndrome reach adulthood and are discovered by chance.

Since the advent and development of modern imaging methods, a few rare cases of multiple sclerosis of accidental discovery have been described. CT scans accurately determine the topography, nature, and morphology of the abnormalities encountered [20,21].

4. Conclusion

Polysplenia is a complex poly malformative syndrome, most often involving abdominal cardio-pulmonary and visceral abnormalities. It occurs in the neonatal period or during childhood due to symptomatic cardiac malformations, whereas malformations without associated cardiac involvement reach adulthood.

Visceral and vascular malformations can simulate tumour masses or adenomegalia.

It is important to stress the inconsistency of all these variations: there is no clinical picture characteristic of polysplenic syndrome, nor any biological signs.

Declaration of Competing Interest

The authors report no declarations of interest.

Sources of funding

No sources of funding.

Ethical approval

We have reported a single case, not a clinical study, with no requirement for ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Khalid Elhattabi (KH) came up with the study concept. Othmane Elyamine, Mounir Bouali, Abdelilah el bakouri, Fatimazahra Bensardi and Fadil abdelaziz collaborated in the patient's medical care. Othmane el yamine examined the pathology. Khalid elhattabi, Othmane el yamine reviewed the manuscript. All authors have approved the final article should be true and included in the disclosure.

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