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Fortuitous Diagnosis of Polysplenia Syndrome in an Adult with Mild Abdominal Pain: *Case Report and Review of Literature*

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Abstract **Case Report**

Abnormal arrangement of thoraco-abdominal anatomical structures and their vascular elements in association with dysmorphism, is called heterotaxia syndrome or situs ambiguous syndrome [3]. This syndrome is further dived into two main sets of anomaly bundles, on one hand bilateral left-sidedness or polysplenia syndrome and on the other hand bilateral right-sidedness or asplenia syndrome. It is viewed in literature as a spectrum ranging from situssolitus which is the normal distribution of organs to situsinvertustotalis, the mirror-image of the normal arrangement of organs inside the body and in between the twoentities, a wide range of malformations, as a result of a Disruption of axis determination during early embryonic: situs ambiguous .We are reporting a case of polysplenia syndrome discovered in a patient with only a mild diffuse abdominal pain.

Keywords: Polysplenia, mild abdominal pain, CT scan, adult.

INTRODUCTION

Polysplenia syndrome is a rare congenital subtype of heterotaxia syndrome associated with various visceral and vascular abnormalities. It is essentially characterized by presence of 2 or more spleens and anomalies of other asymmetric organs [6]. However, polysplenia is a controversial and complex entity with no fixed pathognomonic features but rather a broad spectrum of abnormalities [4].

Reports indicatethatmost patients withpolysplenia syndrome die before 5 years of age, due mainly to cardiovascular defects or biliary atresia incompatible with life or unmanageable with modern surgery procedures .Although described in some reports, it is unusual that the clinical aspect, of such radical re-arrangements of inner organs, can be less telling or be diagnosed in adults all together [2].Therefore, polysplenia has been described mainly in childhood [1].

CASE REPORT

A 55-year-old female, with no particular medical and surgical history, consulting for isolated, diffuse, recent, few weeks' long abdominal pain, initial lab work came back normal.

An abdominal ultrasound was performed, showing mainly reduced size pancreas in addition to the existence of multiple spleen which prompt us for further evaluation of the patient by contrasted enhanced computed tomography (CECT).

The scan unraveled a multitude of additional anatomical defects: the presence of a polysplenosis with a common mesentery; a trunk Porte pre duodenal; a portal vein located anterior to the duodenum and pancreas, with a course abnormally distant from the aorta and the Superior Mesenteric Artery; a short Pancreas; an incomplete common mesentery and a mobile Caecum by default of attachment of the appendix fascia.

The second biological assessment following up returned without anomaly. Beside painkillers on demand a wait and see approach has been taken.

DISCUSSION

The polysplenia syndrome is defined by the existence multiple spleens. It can occur in both genders with an identical frequency, mostly at an early age. The precise aetiology of polysplenia is unknown. Embryonic and genetic components have all been implicated as factors in polysplenia [1].

Polysplenia is classically described as left isomerism or bilateral left handedness referring to duplication of left sided structures with bilateral bilobed lungs, bilateral hyparterial bronchi and bilateral pulmonary atria in addition to presence of multiple spleens. On average in 70 % cases, polysplenia is associated with complex cardiac anomalies [3]. Other anatomical defect can be associated to this syndrome but without fixed presentation, therefore the whole range of anomalies is described as a spectrum.

In our case, the patientis in her mid-50s and presented no substantial clinical symptoms besides mild fluctuating in time and intensity abdominal pain, this is very uncommon since only about 10% patients may reach adulthood without any complications [3]. The syndrome is this caseencompasses multiple congenital malformations:

Spleens

The splenic mass is usually divided into fairly equalized masses, varying in number from 2 to 6 and ranging from 1 to 6 cm in diameter. The location of the spleens is in either the left or right upper quadrant, along the greater curvature of the stomach [6]. In our case, 4 spleens slightly varying in sizes are located in the left upper quadrant (figure 1).

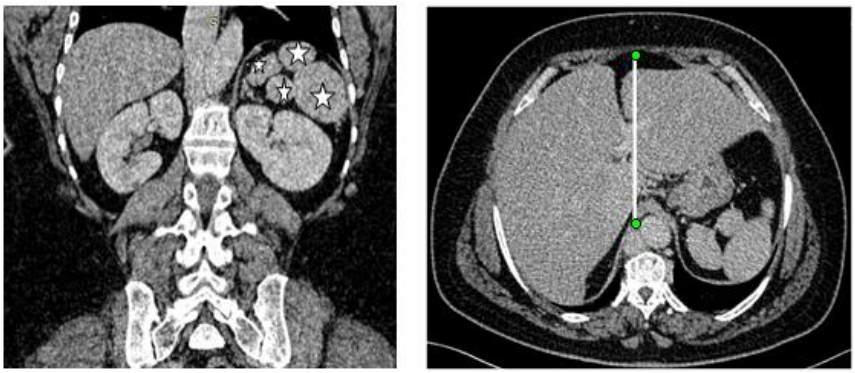

Fig-1: Contrasted enhanced CT in coronal and axial sections showing four spleens different in sizes with an unusually medial falciform ligament

Veinous anomalies

Inferior vena cava interruption with azygous continuation is the second most common abnormality observed in polysplenia patients after multiple spleens [3].To understand this anomaly, it should be noted that the embryologic development of IVC is complex, and the anomaly is mainly a result of failure of anastomis between componments of the four segments of IVC: hepatic, suprarenal, renal, and in frarenal during the embryonic growth [4] (figure 2).

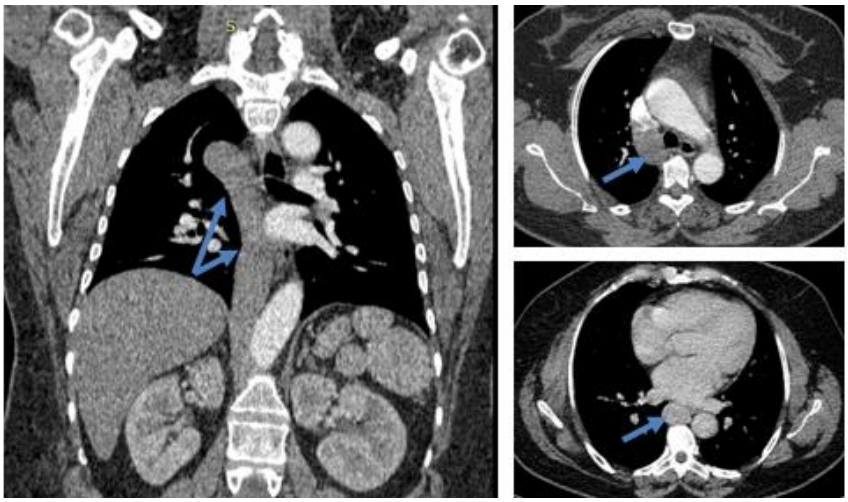

Fig-2: Contrasted enhanced CT in coronal and axial sections displaying the agenesis of the IVC above the renal veins with dilatation of the azygos vein (blue arrow)

Preduodenal portal vein

Cases with only PDPV, first described by Knight HO in 1921, are recognized as very minor anomalous cases [1]. Even though intestinal obstruction is often displayed in such cases, there were no anomalies of the digestion system for our patient [5]. A PDPV can be associated with duodenal atresia, stenosis, web, annular pancreas and malrotation, and surgery may be required for treatment [4].The patient had a PDPV but with no clinical repercussions. Although it

has a been at least a case of polysplenia in an adult presenting with obstructive jaundice, possibly formed by biliary stasis as a result of compression of the common bile duct by the preduodenal portal vein[1].

When surgery is required, especially for procedures involving the upper abdomen, care must be exercised. If PDPV is not detected prior to surgery, it can cause severe complications, such as hemorrhage and vascular ligation. Such accidents can be prevented by performing careful diagnostic imaging in advance, such as CT (figure 3).

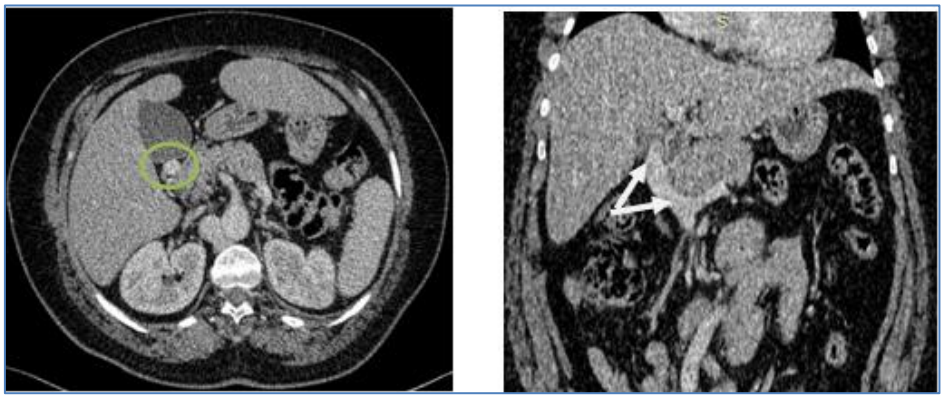

Fig-3: Contrasted enhanced CT in axial and coronal sections showing a pre duodenal PT, located in front of the duodenum and pancreas with a distant trajectory from the aorta and the SMA. (White arrows)

Pancreas anomalies

Anomalies of the pancreas have been described in polysplenia syndrome [1]. Normal pancreas formation occurs from fusion of ventral and dorsal pancreatic buds. The ventral pancreatic bud gives rise to the uncinate process and the head, while the dorsal pancreatic bud gives rise to the body and tail. The development of both dorsal pancreatic bud and spleen occur in the dorsal mesogastrium. Consequently, anomalies in both these organs can be expected in patients with polysplenia syndrome [3].

Clinical significance of dorsal pancreatic agenesis is the development of pancreatitis owing to poor drainage from the remnant ventral duct. CT demonstration of a short pancreas is not synonymous with agenesis and is a pitfall well avoided which is the case in this report where the pancreas seems only shorter (figure 4). An abbreviated or annular pancreas is described in90% of cases [2]. Furthermore pancreatic anomalies differentiations can be achieve with MRCP or endoscopic retrograde cholangiopancreatography.

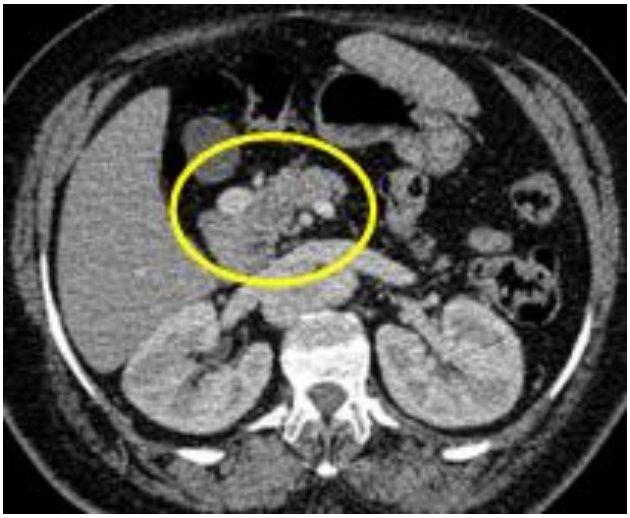

Fig-4: Contrasted enhanced CT in axial section showing a short pancreas and a pre pancreatic PT.

Intestinal malrotation

Common mesentery counts in more than 75% of cases of polysplenia [2], it is commonly associated with anomalies of intestinal rotation. It is the result of a rotation of the midgut during embryologic development, the mesenteric root is broad, extending from the duodeno-jejunal junction in the left upper quadrant to the cecum in the right lower quadrant Intestinal rotation abnormalities are usually collectively termed malrotation and include nonrotation, incomplete rotation and the rare reversed complete or incomplete rotation [4].

Radiologic identification of intestinal malrotation is important even if there are no symptoms because patients with such defects are at risk of midgut volvulus which is a surgical emergency [4, 6]. Surgical correction is advised by many authorities for all surgically fit patients with malrotation, regardless of age and symptoms. As according to our patient, a surgical appoint has been set to tackle prophylactically such eventualities (figure 5).

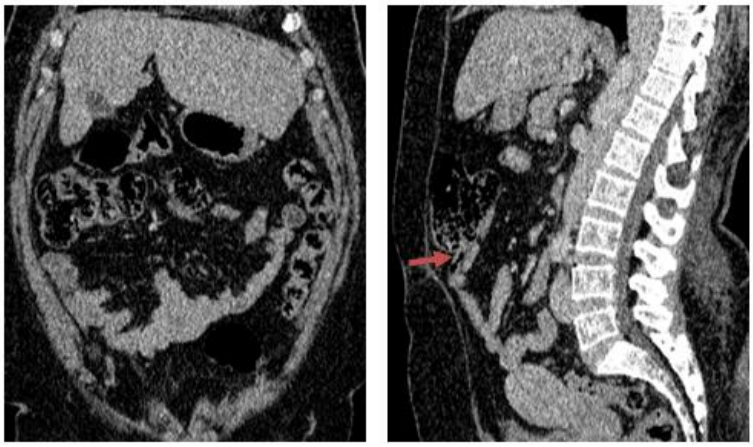

Fig-5: Contrasted enhanced CT in coronal and sagittal sections showing Incomplete common mesentery, mobile cecum due to lack of attachment of Todt's fascia and a an elevated appendix from its usual anatomical placement (red arrow)

Cardiac anomalies

Complex cardiac anomalies arewell known in heterotaxy syndrome and are less common in polysplenia than in asplenia. As far as our case goes, the patient presented thoracic left isomerism in which there is presence of bilobed lungs bilaterally with hyparterial bronchi. Both right and left main bronchi were passing below the corresponding pulmonary artery to reach the lung hilum. Which is only found in 55 % cases of polysplenia reviewed [3]. As for the heart , they were no anomalies noted .backed up by the literature stating that In adult cases of polysplenia cardiac anomalies are fairly rare since their presence would juristically compromise the longevity of the individual making in it unlikely to achieve adulthood in the first place.

The closest differential diagnosis to multiple intra-abdominal spleens is splenosis, an acquired rather than congenital condition that arises in the context of traumatic splenic rupture, can be ruled out by patient history. Splenosis typically consists of multiple small implants of splenic tissue without other visceral anomalies [2].

CONCLUSION

Polysplenia syndrome is a rare disease that occurs in both genders, patients can survive to reach adulthood without any symptoms .The diagnosis is usually made relatively easily with diagnostic imaging including abdominal ultrasound and CT scan. The preeminent anomalies may include, in addition to multiple spleens: a right-sided stomach, a left-sided or large midline liver, right-sided spleen, malrotation of the intestine, a short pancreas, and anomalies of the inferior vena cava.

Conflict of Interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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