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Case Report

Polysplenia syndrome in the adult patient. Case report with review of the literature

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Abstract

Aims: **To report a case of polysplenia syndrome (PSS) in an adult patient.** ***Background:*** **The PSS is a form of situs ambiguos with multiple spleen, cardiac anomalies, abdominal heterotaxia, short pancreas, major venous system and bronquial malformations. It is a rare syndrome, more often found in childhood, and only the 10% of the patients that do not have cardiac anomalies can reach adulthood.**

Results: **A 56 y/o male with obstructive jaundice and intestinal obstruction who was submitted to an abdominal laparotomy suspecting cholangiocarcinoma. He had choledocolithiasis, duodenal kinking by a preduodenal portal vein, intestinal levorotation, hypoplasic vena cava with a prominent acigos vein, short pancreas and polysplenia. A cholecistectomy, biliodigestive and gastroyeyunal bypasses were performed without any complications and with a succesful evolution.**

Conclusions: **In conclusion, PSS is a rare hereditary syndrome that often occurs in childhood and its discovery in an adult is frequently fortuitus. Surgical treatment is an excellent therapeutic option, however is reserved just for complications. The outcome is good and the final evolution depends on the degree of the cardiac anomalies.**

Key words: Polysplenia, preduodenal portal vein, and duodenal kinking.

Introduction

Helwig is credited with describing the Heterotaxia (polysplenia) syndrome in 1929. The polysplenia syndrome (PSS) is a type of situs ambiguous characterized by left isomerism,[1,2] conformed by a group of visceral anomalies of unknown etiology, in which the presence of multiple aberrant splenic nodules and wide range of organic malformations exist. The term left isomerism[2] groups all the morphologic variations that acquire the organs, or part of them, located towards the right of the mean line, when trying to adopt the characteristics of those on the left of his counterpart.

The abnormalities that integrate the PSS are wide;[3] the most constant – in addition to the polysplenia – are cardiac malformations, thickening and interruption of the vena cava with direct continuation towards the acigos vein, along with abdominal heterotaxia. Although its presentation is less constant, the presence of a short pancreas has also been described, along with a preduodenal portal vein, pulmonary and genitourinary malformations. It is a frequent alteration that is detected mainly in childhood, 40% of the affected patients reach 2 years of age and the majority dies before the 5th year.[4] 5 to 10% lack cardiac involvement, which allows them to reach adulthood.[5] There are 23 reported PSS cases in an adult patient, in whom the alterations and the symptoms of presentation associated with them are multiple and unclear, which is why in the majority the diagnosis was incidental *(Table I)*.

Case report

56 y/o male, diabetic of 12 years of evolution, taken previously twice to surgical exploration due to extremity ostheochondromas, presented with a 3 month history of general malaise, hiporexia and weight loss. Thirty days before he entered our institute he developed obstructive jaundice which was unsuccessfully handled as an infectious hepatitis. Diagnosis of an unresctable biliary tumor was made and as a consequence the biliary tract obstruction was relieved with the placement of a biliary stent; however, the patient developed progressive abdominal distension and refussed oral intake.

The physical exam was bening except for generalized jaundice, dehydrated mucous membranes and mild abdomi-

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Abbreviations: Polyslpenia syndrome (PSS), Endoscopic retrograde cholangiopancreatography (ERCP).

nal tenderness. Labs: TB 4,4, DB 2,3 Alkaline phosphatase 1828, Alphafetus protein, carcinoembrionic antigen and Ca 19-9 markers were normal. An abdominal ultrasound was performed along with a Endoscopic retrograde cholangiopancreatography (ERCP) which reported an elarged liver with intra and extrahepatic biliary tract dilation, as well as choledocal and gallbladder lithiasis.

Results

The CT scan corroborated the previous findings and demonstrated both hepatic lobes of similiar dimensions extending to both sides of the abdomen, as well as 5 aberrant splenic nodules *(Figures 1, 2)*. The angio MRI reported an unusual preduodenal location of the portal vein, an hypoplastic inferior vena cava which continued to the acigos vein, abdominal heterotaxis was indirectly acigos vein, abdominal heterotaxis was indirectly determined by the anatomical disposition of the superior mesenteric vessels *(Figure 3)*. The patient was taken to the OR for surgical exploration. The intraoperative findings were duodenal kinking secondary to the compression of the preduodenal portal vein *(Figure 4)*, levorotation of the large bowel, hypoplastic inferior vena cava, short pancreas and spleen. Cholecistectomy, exploration of biliary tract and hepatojejunal anastomosis were made prior of making a Billroth II gastrojejunal anastomosis. The postoperative evolution was satisfactory with an adequate permeability of the biliary tract and acceptable tolerance to the oral route.

Discussion

The PSS is a congenital upheaval usually diagnosed in the childhood stage because almost half of the cases (41%) display serious cardiac abnormalities which are frequently fatal, being the most frequent ones interauricular and/or interventricular communication, transposition of great vessels, stenosis or pulmonary atresia and dextrocardia.[4,5]

Since 5 to 10% of the cases lack cardiac damage or only present a small alteration, it allows the patients to reach adulthood.[5] PSS in the adult produces unclear manifestations; the presence of polysplenia, situs ambiguous or situs inversus (21%) is enough to establish the diagnosis;[1-3] which is generally unsuspected and thus fortuitous in nature.[2,3]

The case we present reunites most of the characteristics that define this syndrome. The unique sign of the PSS is the presence of multiple spleens, ranging from 2 to 16 - five in this case -, in their majority located throughout the greater curvature of the stomach, in more of 60% of the cases it is to the right of the mean line, generally comprising or as the only evidence of the intestinal syndrome of

Table I. Heterotaxia (Polysplenia) syndrome in the adult patient. Characteristics and clinical findings.[2]

Age/Sex	Presentation	Polysplenia/Intestinal Malrotation	Cava Vein/Acigos Vein	Preduodenal portal vein	Liver Pancreas
18/M	Anemia	Right/NR	Left/Normal	NR	ML/NR
20/M	Abdominal pain	Right/NR	Left/Continous	Yes	ML/Short
20/F	Abdominal pain	Right/Yes	Right/Continous	NR	ML/Short
26/F	RHP	Right/NR	NR/NR	NR	ML/NR
27/M	Abdominal pain	Right/NR	NR/NR	Yes	CL/NR
30/F	Abdominal pain	Right/Yes	Left/Normal	Yes	MLSIM/NR
32/F	Abdominal tumor	Right/NR	Left/Continous	NR	ML con MO/NR
40/M	Fever+ AT.	Right/NR	Right/Continous	NR	ML/NR
41/F	CXR + AT.	Right/NR	Left/Continous	Yes	ML/Short
42/F	RUQ + AP.	Right/Yes	Left/Continous	Yes	LM/Short
43/F	Abdominal pain	Right/Yes	Left/Normal	Yes	ML/NR
44/M	CXR	Left/No	Right/Continous	Yes	LD/N
44/M	Abdominal pain	Right/NR	Left/Continous	Yes	LCH /N
45/M	Epigastric pain	Right/Yes	NR/NR	NR	LM/NR
46/F	CXR	Left/Yes	Left/Continous	Yes	LD/N
48/F	Dysnea+bleeding	Left/NR	Right/Continous	NR	RL/Short
57/M	Suspition of tumor	Left/No	Right/Continous	Yes	RL/Short
57/F	Abdominal tumor	Left/NR	Left/Continous	Yes	RL/Short
62/M	Glioblastoma	Left/Yes	Right/Normal	Yes	RL/Short
68/F	CN asociated	Left/NR	Right/Continous	NR	RL/Short
70/F	Abdominal tumor	Left/Yes	Left/Continous	Yes	LL/N
73/M	Polysplenia	Right/Yes	Right/Continous	Yes	ML-LL/Short
78/M	Anemia	Left/Yes	Right/Continous	NR	RL/N
56/M*	Jaundice, Intestinal obstruction	Left/Yes	Left/Continous	Yes	RL-LL/Short

N: Normal, NR:Not reported, RL: Right lobe, LL: Left lobe, ML: Middle Lobe,
CL: Central Lobe, LCH: Left change:
*Our case

intestinal disrotation. The abdominal CT scan and the presurgical MRI allowed us to identify in the patient the second more frequent alteration of the PSS; hypoplasia of the inferior vena cava with absence of its intrahepatic segment and direct continuation towards the acigos or hemiacigos vein.[6] As it happened in this case, the liver occupied the center of the abdomen (50% of the cases) and their lobes were of similar dimensions (25%).

The clinical picture of our patient correlates with the kinking produced by the presence of a portal vein that carried to the second portion of the duodenum, this finding is documented in 50% of the PSS cases, although the development of intestinal occlusion has not been reported previously. Preduodenal portal vein is a common venous anomaly in this syndrome. It passes ventral to the duodenum and the head of the pancreas, and appears as a round structure ante-

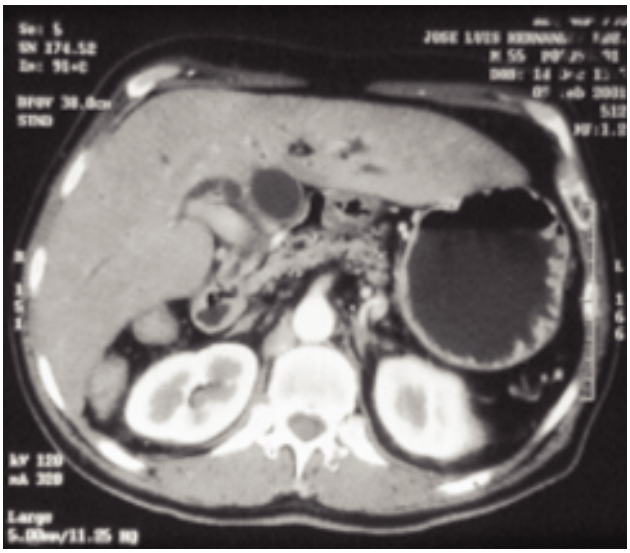

Figure 1. Abdominal CT scan: Hepatomegaly. The left lobe extends up to the hypochondrium. Preduodenal portal vein location, inverted superior mesenteric vessels, continuity of the vena cava with the acigos vein and absence of the intrahepatic vena cava.

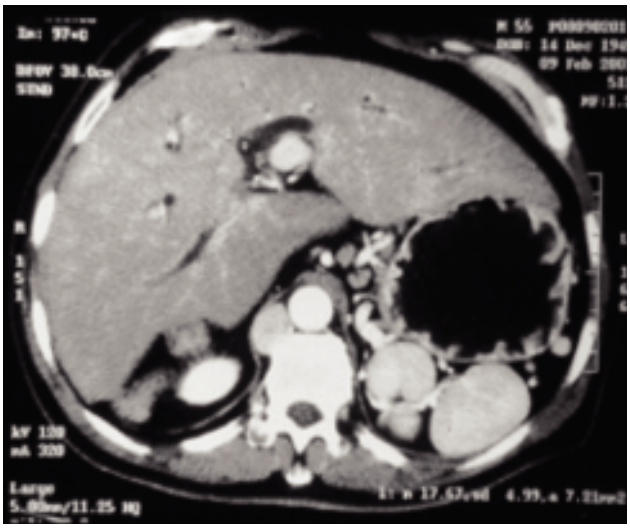

Figure 2. Abdominal CT scan: Hepatomegaly. The left lobe extends up to the hypochondrium. Continuity of the vena cava with the acigos vein and absence of the intrahepatic vena cava. Polysplenia.

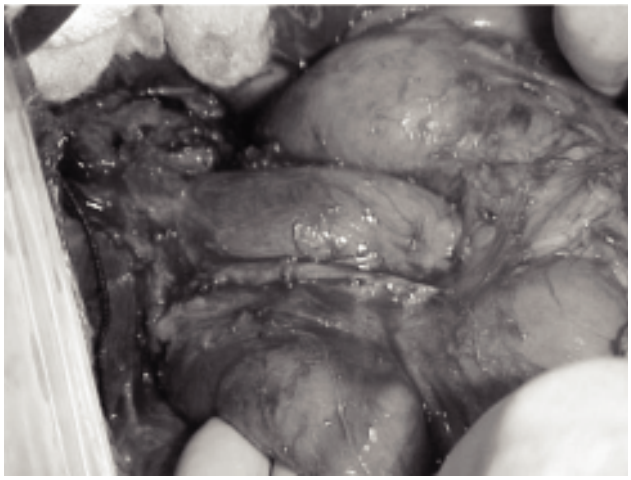

Figure 3. Duodenal kinking secondary to a preduodenal portal vein.

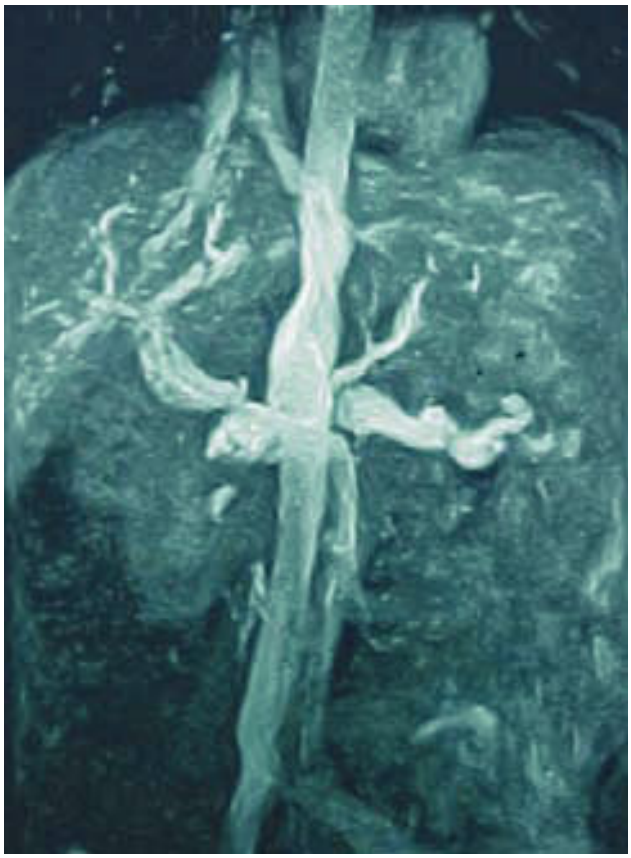

Figure 4. MRI: Polysplenia with enlargement of the acigos vein. Angio MRI: direct drainage of the suprehepatic veins into the right auricle, the extra hepatic vena cava and its continuity to the acigos vein.

rior to the pancreatic head on CT and MRI. Preduodenal portal vein might interfere mechanically with pancreatic development, thereby increasing the risk of pancreatic anomalies such as annular pancreas.

Potential hazard to the preduodenal portal vein in some surgical procedures is obvious. Accidental injury to the vein itself was reported in a case of polysplenia syndrome undergoing biliary surgery.[7] When the alteration involves the pancreas, a diminution in its dimensions takes place (short pancreas).[8-10] All of these anatomical alterations are representative of the PSS, but none of them are pathognomonic. The extra abdominal alterations include the cardiac defects already mentioned, the development of bilateral lobulated lungs (60%) and genitourinary malformations like renal agenesis, hypoplastic kidneys, and duplication of the collector systems, which were luckily absent in this case. The malformations of our patient were limited to the abdomen, which is why the surgical corrective measures we could offer him, correcting the biliary flow and duodenal transit, proved to be a good result for the patient in the medium to long term with a favorable prognosis.

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