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A complete situs inversus viscerum with hepatic, intestinal and renal vascular anomalies: one case report and review of the literatur

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SUMMARY

Situs inversus viscerum (SIV) is a rare congenital anomaly, which is still an intriguing phenomenon to anatomists and physicians alike. A complete SIV is characterized by a left-right transposition and mirror image of all thoraco-abdominal organs and their vasculature. The present report is based on one case with complete SIV, which was observed during the routine educational dissections of cadavers in the authors' Anatomy Department. A transposition of all truncal organs and their vasculature, and several variations of arteries and veins were present. The right branch of the proper hepatic artery was replaced by an artery that emanated from the superior mesenteric artery. The latter also released the inferior mesenteric artery. Additionally, a left accessory renal artery ran anterior to the inferior caval vein and posterior to the ureter to enter the hilum of the left kidney. There was also a variation in the anterior-posterior arrangement of the hilar structures of the left kidney. Additionally, a globally enlarged heart with coronary artery bypasses, a replaced aortic valve and an aortic arch aneurysm was observed. This case report is unique, as it presents a previously unreported co-incidence of SIV and hepatic, intestinal and renal vascular anomalies. It is important for the surgeon to be aware of such variations while planning an abdominal surgery in patients with SIV.

Key words: Anatomy – Dissection – Situs inversus viscerum – Hepatic artery – Mesenteric arteries – Renal artery

Abbreviation list

Situs inversus viscerum: SIV
Coeliac trunk: CT
superior mesenteric artery: SMA
left gastric artery: LGA
common hepatic artery: CHA
proper hepatic artery: (PHA)
left/right hepatic artery: LHA/RHA
replaced right hepatic artery: rRHA
cystic artery: CA
portal vein: PV
renal artery: RA
accessory renal artery: aRA
inferior caval vein: ICV
renal vein: RV
testicular vein: TV

INTRODUCTION

The normal pattern of anatomical positions of thoraco-abdominal organs and vasculature or their

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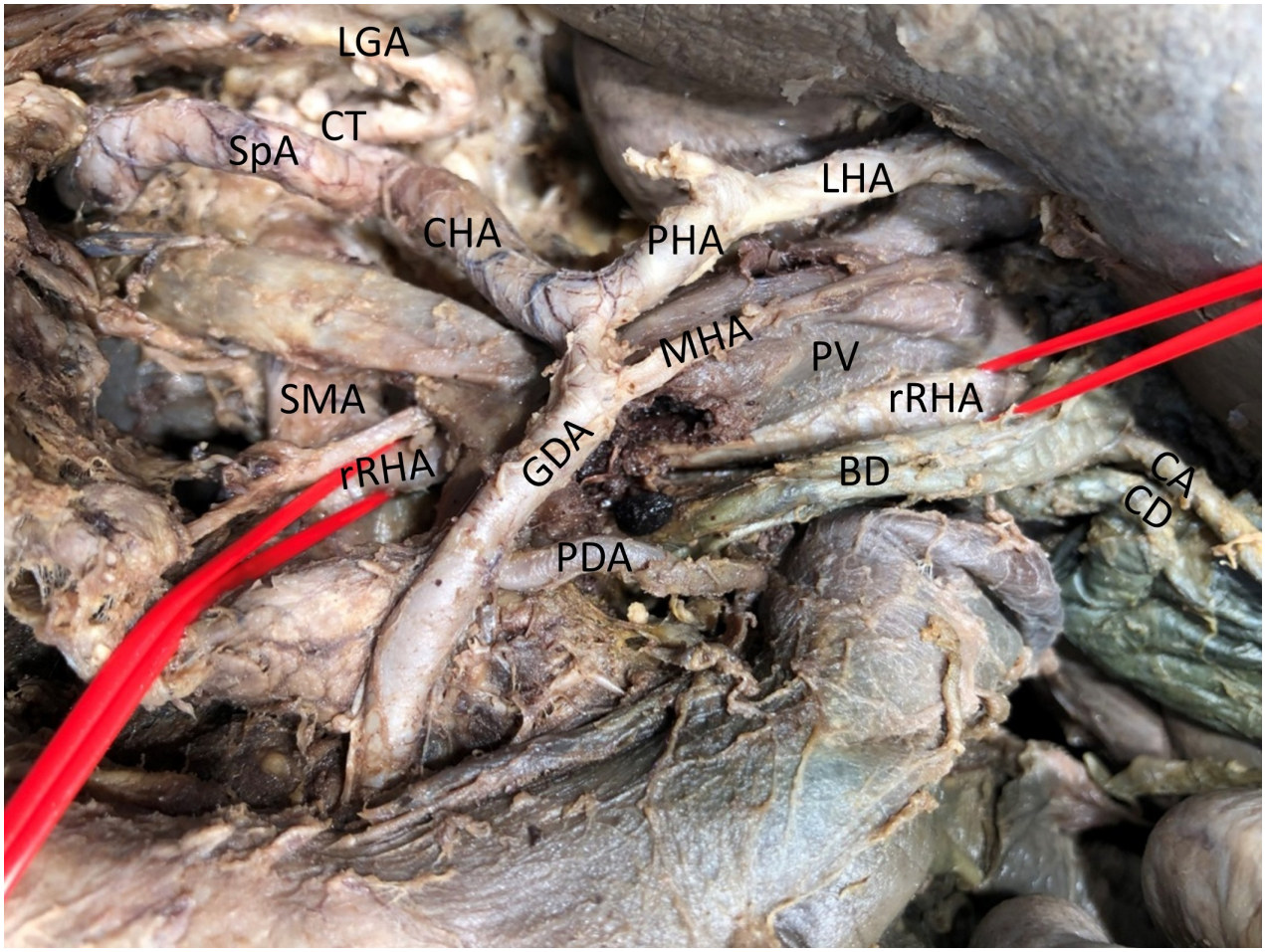

Fig 1. Hepatic arterial variation. Coeliac trunk (*CT*), superior mesenteric artery (*SMA*), splenic artery (*SpA*), left gastric artery (*LGA*), common hepatic artery (*CHA*), proper hepatic artery (*PHA*), left hepatic artery (*LHA*), replaced right hepatic artery (*rRHA*), cystic artery (*CA*), middle hepatic artery (*MHA*), gastroduodenal artery (*GDA*), posterior superior pancreaticoduodenal artery (*PDA*), portal vein (*PV*), bile duct (*BD*), cystic duct (*CD*). The left hepatic artery branched off from the hepatic artery proper that emanated from the common hepatic artery of the coeliac trunk. In addition, there was a replaced right hepatic artery (marked by red ligament), coming from the superior mesenteric artery. An artery branching off from the gastroduodenal artery may constitute a middle hepatic artery (*MHA*).

typical left–right asymmetric arrangement is known as situs solitus (normal position). Laterality disorders like situs inversus viscerum (SIV) and situs ambiguus have a low incidence rate of about 1.1 in 10.000 live births (Lin et al., 2014). SIV is characterized by a right-left transposition and mirror image of all truncal organs and their vasculature. By contrast, the normal asymmetry of the organs is lost in situs ambiguous giving rise to an abnormal symmetrical arrangement of some organs: for instance, the presence of multiple spleens (polysplenia) or absence of the spleen (asplenia) (Brassett and Ellis, 1991). SIV has been known for a long time. An early description of SIV was published by Matthew Baillie (Baillie, 1788). The presence of SIV in primary ciliary dyskinesia is named Kartagener Syndrome (Kartagener, 1933). In the present study, a previously unreported combination of arterial variations is presented in one case with complete SIV.

CASE REPORT

A complete SIV was observed in a 78-year-old male cadaver during the routine educational dissections of body donors. All cadavers in the Department of Anatomy were donated to the Body Donor Program of the University of Frankfurt (Germany). All individuals consented that their remains, after death, be used in whatsoever way shall be deemed most beneficial for the purposes of teaching of anatomy and medical research.

A transposition of all truncal organs including their vasculature showing a mirror image of the normal organ anatomy was present. A globally enlarged heart (cardiomegaly) with aortocoronary venous and thoracic internal artery bypasses and a pacemaker with electrodes in the right atrium and right ventricle was apparent. The aortic valve was replaced and an aortic arch aneurysm was observed.

Table 1. Hepatic arterial variations in SIV. Case reports of surgical procedures or cadaver dissections in SIV with reported hepatic arterial variations

Hepatic arterial variation	Case	Reference
CHA from SMA	cadaver dissection	González-Castillo et al., 2018
	cadaver dissection	Chandraraj, 1976
	LADG for gastric cancer (case 1)	Min et al., 2013
	PD for pancreatic head cancer	Chen et al., 2018
PHA from SMA	hepatic resection for metastatic colon cancer	Uemura et al., 2009
PHA from supracoeliac aorta (recipient)	LDLT for biliary atresia (case 2)	Kamei et al., 2005
RHA and LHA from supracoeliac aorta (recipient)	LDLT for biliary atresia (case 1)	Kamei et al., 2005
LHA from SMA	LADG for gastric cancer	Sumi et al., 2014

HA: hepatic artery, prefix R: right, L: left, C: common, P: proper, LGA: left gastric artery, SMA: superior mesenteric artery, LADG: laparoscopic-assisted distal gastrectomy, LDLT: living-donor liver transplantation, PD: pancreaticoduodenectomy.

Several variations of arteries and veins were present. The coeliac trunk (CT) divided into its regular branches. As a variation, the superior mesenteric artery (SMA) gave off the right hepatic artery (RHA) to supply the right hepatic lobe. This artery is referred to as the replaced right hepatic artery (rRHA), travels dorsal to the pancreas and enters the porta hepatis through the hepatoduodenal ligament, positioned dorsal to the portal vein (PV) and the biliary duct (Fig. 1). The mesenteric arteries were divided into their regular branches, according to the textbook case (Sobotta, 2010). As a variation, the inferior mesenteric artery emanated from the SMA. Furthermore, an accessory renal artery (aRA) of the left kidney travelled ventral to the inferior cava vein, whereas the proper left renal artery (RA) passed posterior to the inferior cava vein (ICV). The anterior-posterior arrangement of the hilar structures of the left kidney varied from normal, insofar as the anterior branch of the RA was followed by the renal pelvis, the renal vein (RV) and the posterior branch of the RA. Finally, two testicular veins (TV) on the left side were also observed (Fig. 2).

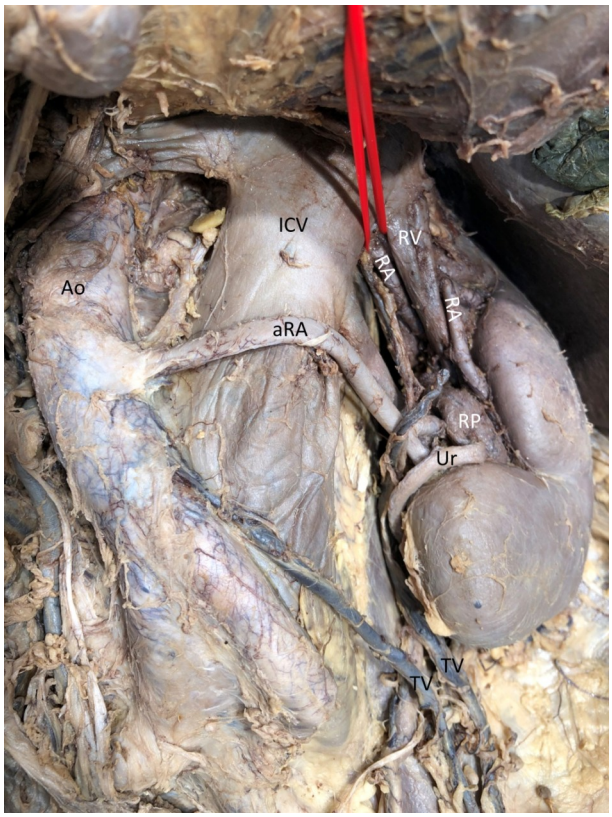

Fig 2. Accessory renal vessel. Aorta (Ao), renal artery branches (RA), accessory renal artery (aRA), inferior caval vein (ICV), renal vein (RV), testicular veins (TV), renal pelvis (RP), ureter (Ur). The accessory renal artery is a branch of the abdominal aorta and runs anterior to the inferior caval vein and posterior to the ureter. Two testicular veins are present, branching off from the inferior caval vein. The renal vein travels between extrahilar anterior and posterior divisions of the renal artery (marked by red ligament).

DISCUSSION

In the development of lateralization, monocilia of the embryonic node generate an extracellular fluid flow directed to the left, which leads to an asymmetrical expression of morphogens and left-right axis specification. In primary ciliary dyskinesia, ciliary components are defective. In this condition, SIV is present in about 50% of affected people (Mclean and Dunwoodie, 2004).

The present study reports on one case of complete SIV with a combination of variations of the hepatic, mesenteric and renal arteries, which is, to the best of the authors' knowledge, not reported yet.

Normally, the CT branches into the splenic, the common hepatic (CHA) and the left gastric arteries (LGA). The CHA releases the proper hepatic artery (PHA), which splits up into a right and left branch, commonly termed the right and left hepatic arteries (RHA, LHA). In a literature review on the main anatomic variations of the hepatic artery, the normal type of hepatic arterial anatomy was present in 81% of cases (Noussios et al., 2017). Variant RHA or LHA stem from the SMA or from the LGA. Accessory vessels run in addition to the regular branch, while a replaced artery substitutes the reg-

ular vessel. The regular RHA and a rRHA usually release the cystic artery (CA) in the cystic Calot's triangle, similar to our case. With an accessory right hepatic artery, two arteries might be found in the cystic Calot's triangle (Reynolds, 2017). Hepatic arterial variations were found in surgical procedures or cadaver dissections in SIV (Table 1).

Usually, the superior and inferior mesenteric arteries branch off from the aorta separately. As a variant, in the present case, the inferior mesenteric artery emanated from the SMA. During development of coeliac and mesenteric arteries, numerous ventral segmental arteries are initially interconnected through a ventral longitudinal anastomosis. Subsequently, most ventral segmental arteries and parts of the longitudinal anastomosis disappear, and, as a result, the coeliac axis is separated from the SMA. When in this remodeling process separation takes place at a higher level, replaced or accessory hepatic artery might result (González-Castillo et al., 2018). A malseparation at a lower level might also explain the irregular offspring of the inferior from the SMA. At least ten cases about a common mesenteric trunk are reported (Yi et al., 2008; Yoo et al., 2011). However, case reports on mesenteric arterial variants in SIV seem to be missing.

Moreover, an aRA was observed, which crossed the ICV anteriorly and the ureter posteriorly to reach the left kidney. In the embryo, the definite kidneys are initially formed in the pelvis. Later, they become abdominal (but remain retroperitoneal) and reach their final position in around the ninth week. During their ascent, the kidneys are supplied by varying branches from the pelvic vessels and later from the dorsal aorta. First, more caudal and then more cranial branches of the dorsal aorta connect to the kidney. Thereby, caudal branches may persist and form accessory renal arteries. In the adult, per kidney, a single RA is present in about 75% of cases. Accessory renal vessels branch from the aorta and follow the proper RA to the renal hilum. Usually, accessory arteries to the right kidney cross the inferior cava vein and the ureter anteriorly (Moore, 2013). In the present case, the aRA ran anterior to the ICV and posterior to the ureter.

Variations of hepatic or mesenteric arteries are important for abdominal surgery. In SIV, anatomical abnormalities including variant vessels may increase the technical difficulty of surgical interventions (Sumi et al., 2014). Accessory renal arteries are of clinical importance because they are end arteries, and damage or ligation will lead to ischemia of renal tissue. A lower polar RA crossing the ureter anteriorly may cause hydronephrosis due to compression of the ureter (Moore, 2013).

Apart from vascular variations, SIV can be associated with other anomalies. For instance, the incidence of cardiac malformations is increased in SIV. Congenitally corrected transposition of great arteries, tetralogy of Fallot and anomalies of the coronary arteries are the most frequently reported congenital cardiac malformations in SIV (Lachhab et al., 2011). Taken together, SIV per se is a non-pathological condition, but can be associated with cardiac defects or intraabdominal anomalies (Lee et al., 2006). In practice, it might require modifications of diagnostic, interventional and surgical procedures.

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