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CASE REPORT

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An interrupted inferior vena cava in a situs inversus: a case report and review of the literature

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Situs inversus with interrupted inferior vena cava is an uncommon anatomic variant found in the abdominal and thoracic viscera. In this report, we present a 59-year-old woman with this variation, found during gross anatomical dissection. While this type of variation has been variable, in the present case the hepatic veins drained directly into a very short (2.2 cm) inferior vena cava. The infrarenal component of the inferior vena cava was present and drained into the azygos and hemiazygos veins. Clinical considerations of this variant anatomy are of interest, as they may present in patients as pathology on cross sectional imaging. (Folia Morphol 2009; 68, 3: 184–187)

Key words: situs inversus, inferior vena cava, azygos vein, hemiazygos veins

INTRODUCTION

During early human development, totipotent cells migrate to desired locations. Once in their desired location they begin to divide and mature into a terminally differentiate state, they then grow and divide and eventually become tissues and organs. It has been postulated that signalling pathways are critically important in the drive towards normal anatomical positioning of organs. Asymmetric expression of TGF-β, Homeobox genes, Sonic hedgehog and Fibroblast growth factor have all demonstrated their importance in normal left-right development [2]. It is postulated that when improper signalling occurs the result is some level of visceral abnormality, such as situs inversus. The normal positioning of the major organs, i.e. the liver, spleen, stomach, and heart, are referred to as situs solitus. Additionally, the apex of the heart must be defined in which the possible positioning is to the right, left, or centre and is referred to as dextrocardia, levocardia, and mesocardia, respectively. Situs inversus is a congenital abnormality that occurs in approximately 0.01% of the population [5]. It results in abnormal folding of organs and ultimately causes the major visceral organs to be the mirror image of their normal positions, including a right sided spleen, stomach, and the apex of the heart pointed to the right (dextrocardia). Situs inversus with levocardia has also been described; the result is abnormal positioning of all major organs with the exception of the heart, which remains in its normal position with the apex to the left. This abnormality occurs in 1/22,000 of the normal population, although it occurs more frequently in patients with congenital cardiac abnormalities, 0.4––1.2/100 [6]. An additional anomaly is situs ambiguous, or heterotaxy syndrome. In heterotaxy syndrome there is neither a normal arrangement of

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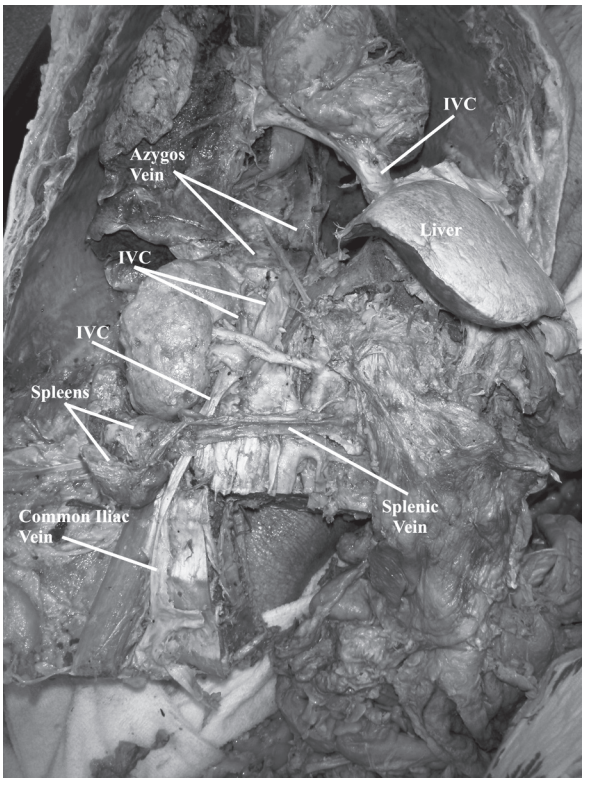

Figure 1. A total interruption of the single left-sided inferior vena cava (IVC) superior to the hepatic veins. Notice the right and left common iliac veins ascending to form a right IVC at the level of the right renal vein. At this level, the IVC split into two vessels and finally drained into dilated azygos and hemiazygos veins. Notice also the multiple spleens which are present as nine isolated smaller spleens characteristic of polysplenia syndrome.

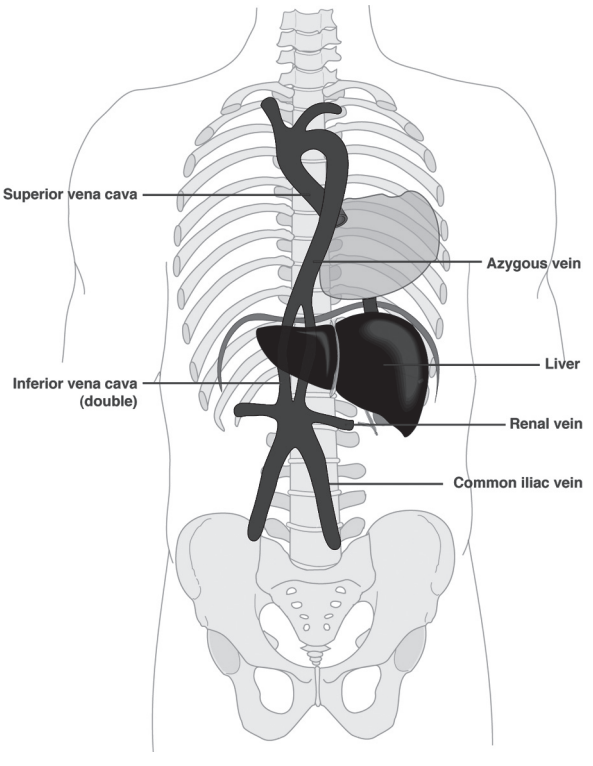

Figure 2. This is a schematic representation of this variation.

the internal organs nor a mirroring of the normal arrangement. It is considered present when the major internal organs have no clear lateralization, and could be considered a combination of both situs inversus and situs solitus [9]. Heterotaxy syndrome is typically linked with cardiac malformations, and often occurs in association with other pathologies, including polysplenia, asplenia, and cardiac malformations [5]. The inferior vena cava (IVC) may be absent with azygos continuation or duplicated in any of the aforementioned abnormalities. With the exception of situs solitus, these terms describe anomalous formation of the visceral organs, which may or may not include dextrocardia and/or vascular abnormalities [10]. The complex development of IVC is a result of the posterior cardinal, subcardinal and supracardinal veins, of which components of each are believed to regress and eventually form IVC [17]. However, if the supracardinal channel on the left and right side persists, a double IVC will remain, which is estimated to occur in ~0.03% of individuals [4, 7]. We report a case of situs inversus with double IVC discovered upon gross dissection.

CASE REPORT

We present a case of situs inversus with interrupted IVC in a 59-year-old female cadaver (Figs. 1, 2). The case was discovered during a routine anatomical dissection of the thorax and abdomen at the University of Alabama in Birmingham during 2008. The cadaver did not show any other gross abnormalities or evidence of procedures involving the thorax and the abdomen.

The dissection revealed a double IVC; one on the left side superior to the hepatic veins and one on the right draining to the azygos vein. The hepatic veins were draining into a short IVC (2.2 cm in length) and then into the right atrium. The right and left common iliac veins ascended and joined to form a right inferior vena cava at the level of the right renal vein. At this level, the IVC split into two vessels and finally drained into dilated azygos and hemiazygos veins. The superior vena cava

and its tributaries were normal. The spleen was present to the left as nine isolated small spleens characteristic of polysplenia syndrome. The splenic vein was formed from tributaries of all nine splenic segments to drain in the portal system. The rest of the organs appeared to be normal during gross examination, and the rest of the cadaver was unremarkable.

DISCUSSION

The literature demonstrates a clear association between situs inversus and other congenital conditions such as tetralogy of Fallot, Kartagener syndrome, duodenal atresia biliary atresia, and gastroschisis [11, 12]. Since this information can be useful for diagnosticians and surgeons, situs inversus is a clinically relevant anomaly, which should be reported as often as it is observed.

A review of the embryogenesis is helpful for an understanding of the etiology of the venous anomalies. The IVC develops between the 6th and 8th weeks of gestation, as a structure comprised of three paired embryonic veins: the posterior cardinal, subcardinal, and the supracardinal veins. There is a cyclical process of development and regression, which eventually culminates into the permanent asymmetric IVC [16]. Briefly, the posterior cardinal veins return all of the blood from the body wall inferior to heart, while blood from the viscera is returned by the vitelline veins. Eventually an intersubcardinal anastomosis forms between the paired subcardinal veins while an anastomosis forms between the subcardinal veins and the posterior cardinal veins. During this step, the right subcardinal vein joins the hepatic segment of the IVC forming the vitelline vein. Eventually the pre-renal division of the IVC is formed by the progressive degeneration of the posterior cardinal veins, therefore shunting blood through the hepatic segment of the IVC [1]. Next, there is formation of the paired supracardinal veins, which eventually separate into the superior and inferior portion of the azygos. The posterior cardinal veins continue to atrophy, and blood from the lower body passes through the suprasubcardinal anastomosis and onto the prerenal IVC, while blood from the left body is sent across the interpostcardinal anastomosis [1]. In general, the normal IVC can be segmented into four main components; suprarenal, renal, infrarenal, and hepatic. Each portion develops from a distinct source. The right subcardinal vein develops into the suprarenal portion while the vitelline vein gives rise to the hepatic portion. The renal segment is derived from two sources, the right suprasubcardinal and postsubcardinal anastomosis, and finally the infrarenal segment is derived from the right supracardinal vein [1].

It has been reported that vascular abnormalities of the IVC in association with situs inversus are extremely rare [14]. IVC duplications occur mainly inferior to the kidneys and have and incidence of 0.3––3% [16]. While it has been reported that with a left-sided IVC, which has an incidence rate of 0.2–0.5%, the statistics are as follows: a retro-aortal renal vein occurs in 1–4% of cases, and a circumaortic venous ring occurs in 1.5–16% of cases [16]. An additional visceral abnormality commonly seen in cases of intestinal malrotation, IVC agenesis, and situs inversus is polysplenia, which is associated with improper lateralization and/or tissue segmentation of the spleen in embryonic development [14]. Furthermore, it has been demonstrated that different visceral abnormalities are associated with asplenia and polysplenia with situs inversus. Cardiac defects are more commonly seen with asplenia while vascular abnormalities are seen in polysplenia [14]. In a study presented by Fulcher and Turner [5], in which 19 adult cases with situs anomalies were examined, 7 cases had multiple spleens and 7 cases had an interruption of the IVC with azygos or hemiazygos continuation. In a different study by Mayo and Rice [13], 7/76 patients with situs inversus had abnormalities of the biliary tract or gallbladder, while in the study of Fulcher et al. [5], 10 cases presented with cholecystitis. The failure of the aforementioned organs and vasculature to attain their normal anatomical location could be the result of aberrant signalling pathways [2]; this should be a point of further work to elucidate the mechanism involved. The findings in our cadaver support the association of vascular abnormalities with situs inversus.

It is clinically important to be able to identify patients with vascular abnormalities such as an interrupted IVC in cases of situs inversus. The precise location of the abnormal vasculature is required in order to prevent any unnecessary injury to the patient or damage to the organs. These variations have been examined in particular for surgical procedures such as aortic bifurcational replacements, inferior caval shunt, and portal caval shunts [15]. In addition, an interrupted IVC can pose problems during retroperitoneal and thoracic surgeries where there is an issue of venous haemorrhage, while also complicating abdominal aortic aneurysm surgeries [3]. Reports have described the requirement of a second

Greenfield filter to be implanted when a previously undetected double IVC was recognized in order to protect against pulmonary embolus [1]. However, in cases in which an IVC filter is required, it has been reported that it is very difficult to identify left IVC and interrupted IVC with azygos continuation on venographic examination, especially with situs inversus. In this case, computed tomography or ultra sound should be used for proper identification and diagnosis [14]. However, caution should still be used when using imaging techniques, it has been reported that a double IVC may be incorrectly identified as a lymphadenopathy [7, 8], which is of particular importance in cancer patients who are being monitored for metastasis. Other potential complications can occur during removal of renal cell carcinoma, lumbar sympathectomy, ureteral surgery, and finally in the dissection of lymph nodes because the lymph drainage flow follows the course of the vasculature [3]. There have been very few reports of double IVC in association with Situs inversus. Kulesza et al. [10] previously described what was believed to be the first case.

CONCLUSIONS

We have demonstrated a patient with situs inversus, polysplenia, and a total interruption of the single left-sided IVC superior to the hepatic veins. The abnormal vasculature is to be expected with polysplenia [14].

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