MULTIPLE VASCULAR ANOMALIES WITH DOUBLE GALLBLADDER - A CASE REPORT

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ABSTRACT

While doing the routine dissection in NRS Medical College , Kolkata, we came across a cadaver (a male subject about 70 years old) who had multiple vascular anomalies along with double gallbladder. Among the vascular anomalies there were persistent left superior vena cava (PLSVC), left sided azygos vein (draining into the PLSVC), left vertebral artery arising from the arch of aorta and the right gonadal vein draining into the right renal vein at a right angle. There were two gall bladders on the undersurface of the liver with two separate cystic ducts opening into the common bile duct. One of these two gallbladders was supplied by a cystic artery arising from the gastroduodenal artery and the other one was supplied by the cystic branch of the right hepatic artery. This study may enhance our knowledge in the field of Embryology and may help the clinicians to adopt an investigative (like coronary angiography) or surgical procedure.

Key Words: vascular anomalies, PLSVC (persistent left superior vena cava), azygos vein, origin of the vertebral artery, gonadal vein, renal vein, double gallbladder

INTRODUCTION

We know that the superior vena cava is present on the right side of the thorax in the superior mediastinum and it drains into the right atrium. The azygos vein is present in the posterior mediastinum of thorax on the right side and on the left side the superior (accessory) and the inferior hemiazygos veins are present. The right gonadal vein drains into the inferior vena cava at an acute angle, whereas the left gonadal vein drains into the left renal vein at a right angle. The vertebral artery arises from the posterosuperior aspect of the first part of the subclavian artery on both sides. We also know that there is a single gallbladder on the undersurface of the liver as stated by Standring et al[1]. But multiple anomalies involving these structures may persist in the body of one person concerned in this case.

MATERIALS AND METHODS

While doing the routine dissection for the undergraduate studies in N.R.S. Medical College, Kolkata , we came across a cadaver (a male subject about 70 years old) who had multiple vascular anomalies in thorax and abdomen along with double gallbladder. The structures were dissected properly, coloured and relevant photographs were taken.

OBSERVATION

i) In the thorax there was persistent left superior vena cava (PLSVC).It was crossing the left side of the aortic arch anterior to the left pulmonary hilum before turning to enter the right atrium. It was replacing the oblique vein of the left atrium and coronary sinus, was receiving all the tributaries of the coronary sinus to open into the right atrium through the enlarged orifice of the coronary sinus (common variety).

On the right side there was no superior vena cava. The right sided brachiocephalic vein was joining the left brachiocephalic vein to form the left superior vena cava.

ii) Azygos vein was present on the left side of the thorax (in the posterior mediastinum), anterior to the vertebral column, behind the hilum of the left lung and by the side of the descending thoracic aorta. It was draining into the persistent left superior vena cava (PLSVC) arching on the hilum of the left lung. The left superior istercostal vein was draining into the left brachiocephalic vein passing in front of the arch of the aorta.

Hemiazygos system of veins had been developed on the right side and the superior and the inferior Hemiazygos veins were draining into the

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Azygos vein on the left side. We found the mirror image of the azygos system of veins in this case.

iii) The left vertebral artery was emerging directly from the arch of aorta between the origin of left subclavian and left common carotid arteries. The right sided vertebral artery was arising from the 1st part of the right subclavian artery (a branch of the brachiocephalic trunk) as usual.

iv) The right gonadal vein (here the testicular vein) was draining into the right renal vein at a right angle (like left side) passing in front of the psoas major muscle. The left testicular vein was draining into the left renal vein at a right angle.

v) There were double gallbladders with two separate cystic ducts opening into the common bile duct on the undersurface of the liver. The larger one was present below the right lobe of the liver, in the fossa for the gallbladder and the smaller one was present in front of the second part of duodenum. The blood supply to the larger one was coming from the right branch of the hepatic artery. One cystic artery was arising from the gastroduodenal artery (visible in front of the duodenum) to supply the smaller gallbladder.

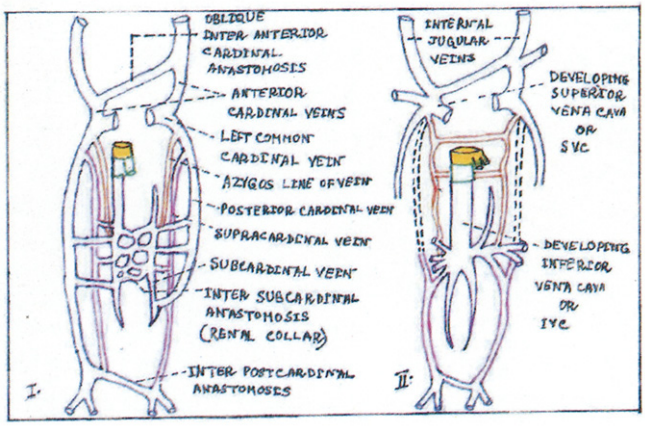

Figure 1. Diagram of somatic venous development (I & II).

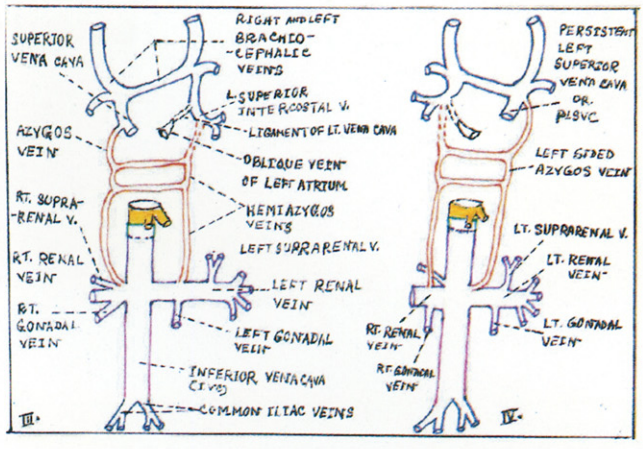

Figure 2. Diagram of somatic venous development (III) with the mirror image of cardinal veins and azygos venous lines (IV).

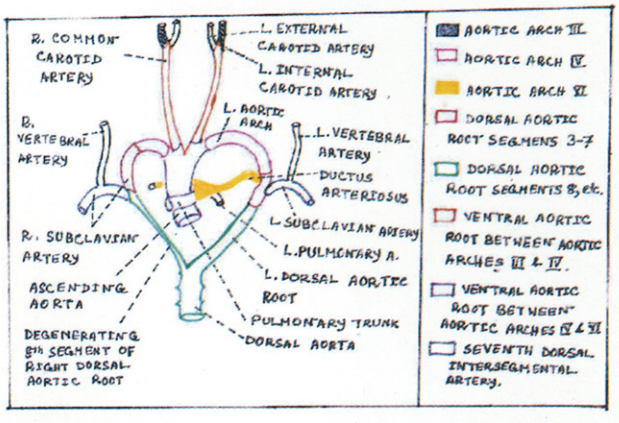

Figure-3. Diagrammatic ventral view of aortic arch complex of a human embryo of 15mm. C.R. length.

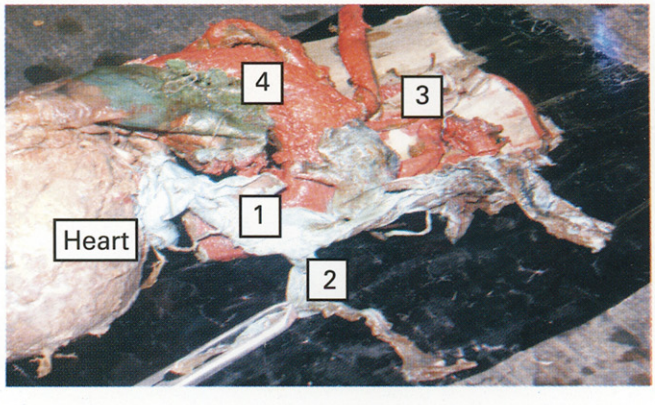

Figure- 4. Persistent left superior vena cava or PLSVC (1), azygos vein (2) on the left side and the left vertebral artery (3) arising from the arch of aorta (4) .

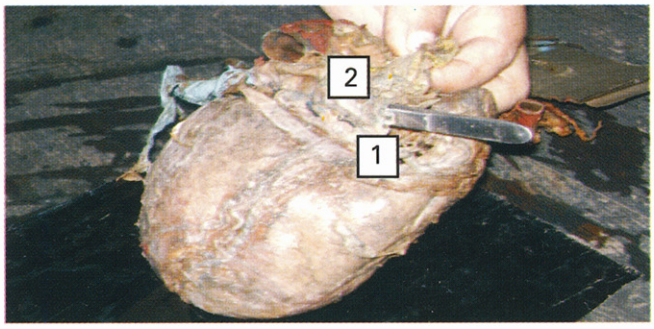

Figure-5. The PLSVC (1) was replacing the coronary sinus to open into the right atrium (2).

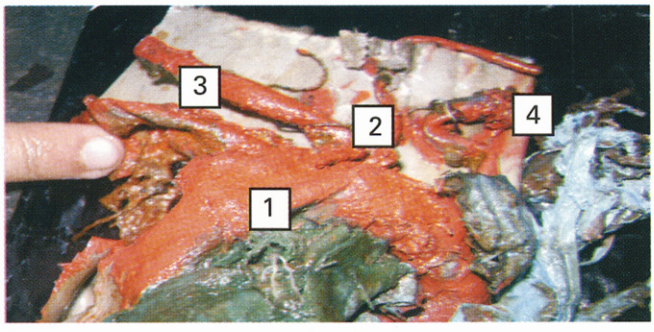

Figure- 6 The left vertebral artery (2) was arising from the arch of the aorta (1) between the left common carotid (3) and the left subclavian arteries (4).

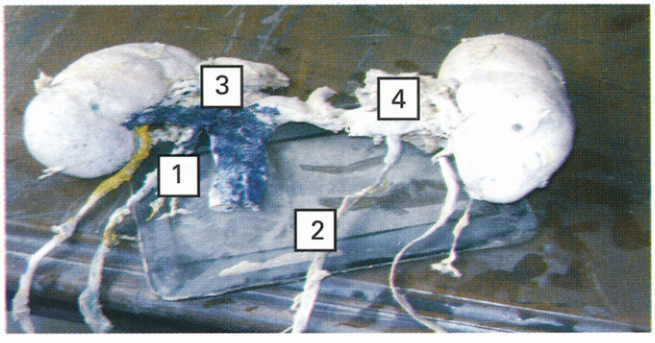

Figure-7. Right and left gonadal veins (1 & 2) draining into the respective renal veins (3 & 4).

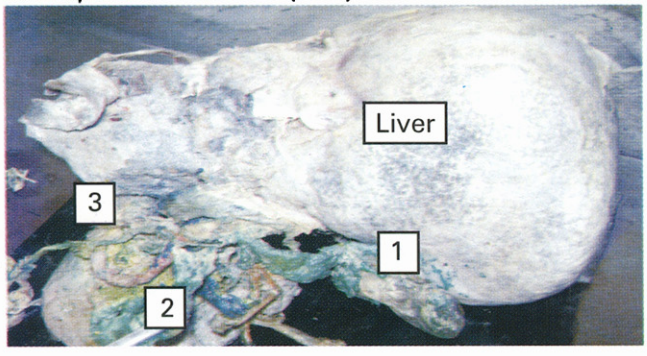

Figure-8. Two gallbladders (1 & 2) on theundersurface of the liver and in front of the duodenum. One cystic artery was arising from the gastroduodenal atery (indicated by an arrow), visible in front of the duodenum (3).

DISCUSSION

EMBRYOLOGICAL CONSIDERATION

1.Initially the two cardinal veins form the main venous drainage system of the embryo - the anterior cardinal veins and the posterior cardinal veins (draining the cephalic and caudal parts of the embryo, respectively). On each side the anterior and posterior cardinal veins join with each other and form the right and left common cardinal veins before entering the respective sinus horn of the developing heart. During the 4th week of intrauterine life the cardinal veins form a symmetrical system. In the 8th week of gestation the two anterior cardinal veins are joined by a large oblique connection which forms the left brachiocephalic vein as described by Sadler[2]. Due to this type of anastomosis the blood from the left side is channeled to the right side due to a right sided dominance and a progressive asymmetry in the vena caval system when some veins enlarge and other veins regress [1].

The upper part of the superior vena cava (SVC) is formed by the right anterior cardinal vein (between the junction of the two brachiocephalic veins and the point of entrance of the azygos vein) and lower part of it is formed by the right common cardinal vein. Caudal to the transverse communication between the two brachiocephalic veins the left anterior cardinal and the left common cardinal veins largely atrophy, the former giving rise to the terminal part of the left superior intercostal vein while the latter forms the ligament of left vena cava and the oblique vein of left atrium. The oblique vein passes downwards across the back of the left atrium to open into the coronary sinus, which represents the left horn of the sinus venosus [1]. Goyal et al[3] also described **the** ligament of vena cava as the 'Ligament of Marshall'.

A persistent left superior vena cava (PLSVC) is caused by the persistence of whole of left anterior cardinal vein, left common cardinal vein and left sinus horn along with the obliteration of the proximal part of right anterior cardinal and the right common cardinal veins. In such a case, blood from the right side is channeled towards the left by way of the brachiocephalic vein [1]. This persistent left superior vena (PLSVC) usually drains into the right atrium by the way of the left sinus horn that forms the coronary

sinus [2]. It is a common variation of the thoracic venous system as stated by Pahwa et al[4] and Gonzalez-Juanatey et al[5].

2. During the 5th to 7th week of the embryo, a number of additional veins are formed [1], [2] -

a) the supracardinal veins which drain the body wall by way of the intercostal veins take over the function of the posterior cardinal veins.

b) The right 4th to 11th intercostals veins drain into a part of the right supracardinal vein which together with a part of the posterior cardinal vein forms the azygos venous line. From this line the azygos vein arises on the right side, whereas, the hemiazygos and accessory hemiazygos veins arise on the left side.

c) The subcardinal veins which mainly drain the kidneys and the caudal parts of these veins are partly incorporated into the testicular or ovarian veins and partly disappear, as stated by McClure et al[6].

3. Usually the vertebral artery on both sides arises from the respective subclavian arteries. The left subclavian artery develops from the left 7th intersegmental artery and the arch of aorta arises from the left aortic sac, left 4th aortic arch and part of the left dorsal aorta [1].

4. The gallbladder including the cystic duct develops from the cystic bud, which arises from the caudal pat of the hepatic diverticulum (endodermal in origin). This diverticulum develops from the junction of the foregut and midgut in the fourth week of intrauterine life The double gallbladder arises due to a septum formation inside the cystic bud. **Rarely the gallbladder may be bifid or duplicated usually with a duplicated cystic duct** [1].

SIMILARITIES WITH THE PRESENT CASE

Keyes & Keyes[7] found a case where the position of the Azygos and Hemiazygos veins was reversed and a left superior vena cava alone was persisting . Present case had similar findings. **The mirror image of superior vena cava and the azygos system of veins was present here.**

Persistent left superior vena cava (PLSVC) is prevalent in 0.3- 0.5% of the general population as noted by Freedom et al[8]; however, it is the most common congenital anomaly of thoracic venous system [3]. According to the statement of Bjerregaard et al[9] it is more frequent in patients with coronary heart disease with the incidence rate of 4.3-11%.

This PLSVC receives the left superior intercostal vein, crosses the left side of the aortic arch to pass anterior to the hilum of the left lung before turning to enter the right atrium. It replaces the oblique left atrial vein, receives the great cardiac vein and other tributaries of the coronary sinus to replace the latter. In 90% cases it drains into the right atrium through the enlarged orifice of the coronary sinus, In the present case PLSVC was following the same course to drain into the right atrium through the enlarged orifice of the coronary sinus. In 10% cases PLSVC drains into the superior aspect of the left atrium, usually with the unroofing of the coronary sinus [1], [4].

Grzybiak et al[10] considered the accessory hemiazygos vein as most variable, draining into the left brachiocephalic, hemiazygos or azygos vein. Accessory hemiazygos and the left superior intercostal vein with the hemiazygos vein may form a continuous channel somewhat similar to the azygos vein as Seib[11] got this finding in almost 40% cases. In the present case there were left sided azygos vein and right sided hemiazygos veins. The left superior intercostal vein may be a direct upward connection of the accessory hemiazygos [12].

The left vertebral artery arises from the arch of aorta in 1-3% cases, found by Dasler and Anson[13]. According to Hollinshead[12], this artery arises from the arch of aorta in 5% cases, most commonly between the origins of the left common carotid and the left subclavian arteries and only rarely about in 0.04% of the cases between the left common carotid and the brachiocephalic arteries. In the present case also the artery was arising from the arch of aorta in between the origins of the left common carotid and the left subclavian arteries. It may also arise from the arch of aorta distal to the left subclavian artery [14]. Possibly this variation is the result of the persistence of the embryonic vascular pattern of 5th to 7th week of the intrauterine life [15].

Occasionally, but not often unless there is an anomalous vena cava, the right ovarian or right testicular vein may enter the right renal vein [11]. The right gonadal vein drains into the right renal vein in 0.66% cases (2 out of 150 cases as studied by Asala et al[16]. In our case also the right gonadal vein was draining into the right renal vein like the left renal vein.

Duplication of the gallbladder with two cystic

ducts is an incidence of about one in every 4000 persons [17], [18]. This occurs in two major varieties. The more common form is that in which each gallbladder has its own cystic duct that empties independently into the same or different parts of the extrahepatic biliary tree. This variety is present in the case concerned. In the other form the two cystic ducts merge before entering the CBD (Common Bile Duct)[12]. Duplication is only clinically significant when some pathological process affects one or both the organs as stayed by Oddsdottir et al [19].

CLINICAL SIGNIFANCE

This study may enhance our knowledge in the field of Embryology and may help the clinicians to adopt an investigative (like coronary angiography) or a surgical procedure. PLSVC, in isolation, is considered to be benign, but is very frequently associated with cardiac abnormalities e.g., atrial septal defect (ASD), ventricular septal defect (VSD), bicuspid aortic valve, coarctation of aorta, coronary sinus ostial atresia and have significant morbidity and mortality [3], [20].

The presence of PLSVC can render access to the right side of heart challenging the left subclavian approach, which is a common site of access for placing pacemakers and Swan-Ganz catheters [3]. Knowledge of the variability of azygos vein system is important in radiological diagnosis (CT and MRI), in the treatment of the thoracic aorta aneurysms and tumours of posterior mediastinum as recommended by Chelic et al[21]. Anomalies of the azygos venous system may become clinically significant in cases of venous obstructions, where they form vital collateral pathways [12]. This study may also help the surgeons to do laparotomy, specially regarding the surgery of kidney.

According to Goiney et al [22] the double gallbladder do not present with specific symptoms and the incidence of disease in this gallbladder is similar to its normal variant. Identification of anatomical details prior to transection along with a high degree of awareness of gallbladder anomalies and meticulous dissection of any additional cystic or tubular structure in the subhepatic region, could possibly prevent catastrophic consequences [23].

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