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ORIGINAL ARTICLE

DOUBLE SUPERIOR VENA CAVA

Two cases

S. ALBAY, F. CANKAL, N. KOCABIYIK, B. YALCIN, H. OZAN

Department of Anatomy, Gulhane Military Medical Academy, Ankara, Turkıye.

SUMMARY

Duplication of superior vena cava (SVC) is a rare anomaly. The incidence of double SVC in general population is 0.3% whereas in patients with congenital heart disease it varies between 10-11%. Double SVC cases have clinical importance if especially the one on the left side drains into the left atrium. Moreover, double SVC is surgically important in the presence of congenital heart disease. In this article, we presented adult patients who incidentally diagnosed with double SVC, one drained into the right atrium while the other into the left atrium and discussed the embryologic basis of these patent vessels.

Key words: vascular anomaly. venous abnormalities. embryology.

RÉSUMÉ

Double veine cave supérieure. À propos de deux cas

La duplication de la veine cave supérieure (SVC) est une anomalie rare. L'incidence de la double SVC est de 0,3 % dans la population générale alors qu'elle atteint 10 à 11 % chez des patients porteurs d'une cardiopathie congénitale. Les doubles cas de SVC ont une importance clinique particulièrement si celle du côté gauche s'écoule dans l'atrium gauche. De plus, la présence d'une double SVC est importante à connaître en cas de chirurgie d'une cardiopathie congénitale. Dans cet article, nous avons présenté des patients adultes présentant une double SVC, une qui se draine dans l'atrium droit tandis que l'autre se draine dans l'atrium gauche. La base embryologique de ces vaisseaux persistants est discutée.

Mots-clés : *anomalie vasculaire. pathologie veineuse. embryologie.*

INTRODUCTION

Cardinal veins comprise the major venous drainage system of the embryo. The oblique anastomotic branch connecting the anterior cardinal veins becomes left brachiocephalic vein. Right anterior cardinal vein and right common cardinal vein unite to form the SVC [9]. SVC primarily originates from the right anterior cardinal vein [6]. Left anterior and left common cardinal veins on the caudal side of the left brachiocephalic vein usually undergo atrophy [13]. During embryonic period, atrophy or patency of certain veins related to this system can result in variations such as double SVC, double inferior vena cava (IVC) or other venous connections that do not normally exist. Double SVC is a rare condition that is characterized with persistence of left SVC [4]. Here we present two patients with double SVC to discuss possible embryologic mechanisms underlying the condition.

CASE 1

Physical examination of a 39-year old female patient with deterioration in hepatic and renal functions revealed signs of heart, liver and kidney failure. Computed tomography showed marked dilatation of heart chambers, pulmonary artery and veins. Left brachiocephalic vein did not join the right brachiocephalic vein, proceeded downwards and bifurcated at the level of the fifth thoracic vertebra. One of its branches traveled posteriorly to join the hemiazygos vein *(figures 1 to 4)* while the other branch drained into the right atrium at the level of the tenth thoracic vertebra *(figure 3)*. Hemiazygos vein, on the other hand, joined the left renal vein at subdiaphragmatic level *(figure 4)*. Disseminated regeneration nodules and perihepatic collaterals in liver were also noted.

CASE 2

A 34-year old male patient presented to the hospital with shortness of breath. Postero-anterior chest radiograph revealed that the mediastinum was wider than normal. Computed tomography (CT) of the thorax depicted a widened ascending aorta (5 cm at the widest region). Left brachiocephalic vein did not unite with the right brachiocephalic vein, only traveled caudally. At the level of sixth thoracic vertebra, it divided into two branches; the posterior branch joined the hemiazygos vein, and the anterior branch joined the left atrium *(figures 5 to 6)*. Hemiazygos vein, in turn, joined the left renal vein at subdiaphragmatic level *(figure 6)*.

Correspondence: S. ALBAY, Gulhane Askeri Tip Akademisi, Anatomi Anabilim Dali, 06018 Etlik, Ankara, Turkıye.
E-mail: soneralbay@yahoo.com

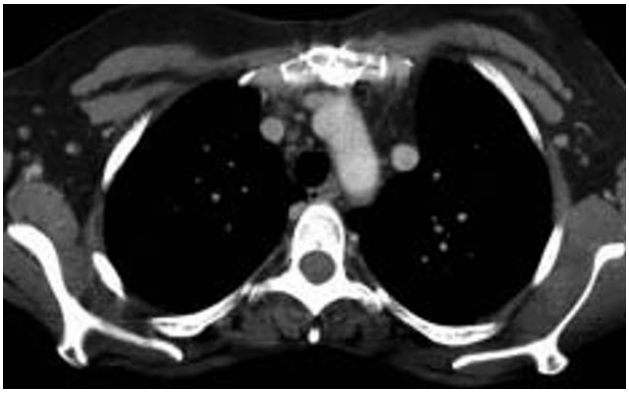

FIG. 1. — Brachiocephalic veins of both sides are shown at the level of aortic arc.

FIG. 1. — *Les veines brachiocéphaliques sont visibles des deux côtés au niveau de l'arche aortique.*

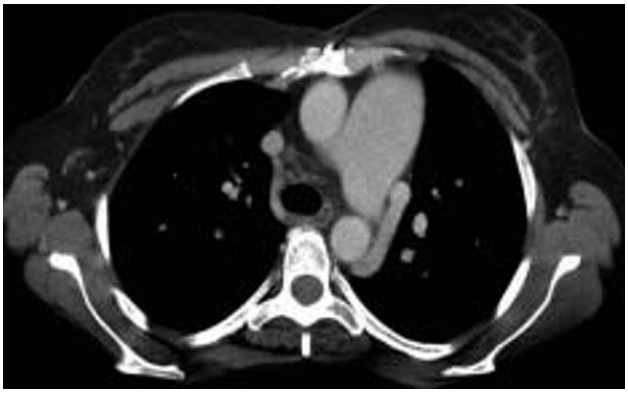

FIG. 2. — Bifurcation of left vena cava and neighboring of its posterior branch and thoracic aorta at the level of enlarged pulmonary trunk.

FIG. 2. — *Bifurcation de la veine cave gauche et environnement de sa branche postérieur et de l'aorte thoracique au niveau du tronc pulmonaire qui est élargi.*

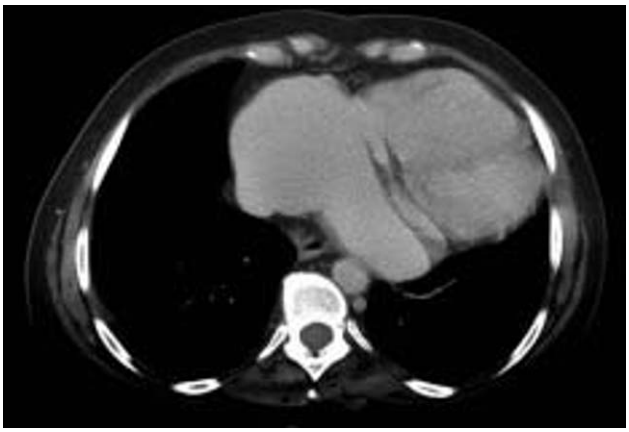

FIG. 3. — Joining of anterior branch of left vena cava to the right atrium.

FIG. 3. — *Jonction de la branche antérieure de la veine cave inférieure à l'atrium droit.*

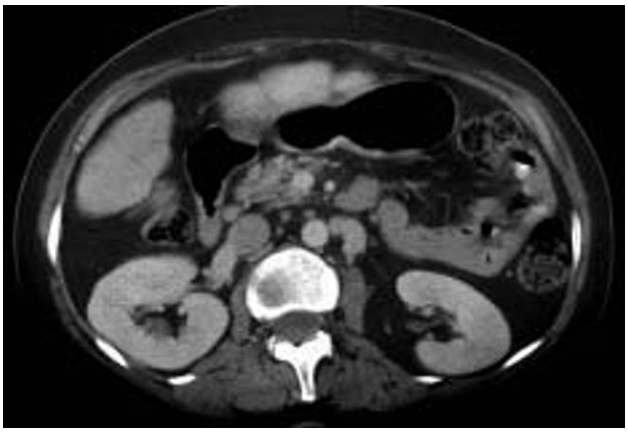

FIG. 4. — Joining of descending branch of left renal vein.

FIG. 4. — *Jonction de la branche descendante de la veine rénale gauche.*

DISCUSSION

During the eighth week of the embryological period, an oblique anastomosis appears between two anterior cardinal veins. This anastomosis becomes the left brachiocephalic vein when the caudal part of the left anterior cardinal vein degenerates. Right anterior cardinal vein and right common cardinal vein gather to form the SVC [7]. Left anterior and left common cardinal veins caudad to the left brachiocephalic vein atrophy to a greater extent [9]. SVC originates primarily from the right anterior cardinal vein [6]. Remnants of the left common cardinal vein continues with the oblique vein of the left atrium (v. obliqua atrii sinistri, Marshall's vein). A fibrous ligament which extends beyond this vein behind the left atrium is the remnant of the left common cardinal vein (left duct of Cuvier). This ligament creates a fold of the serous pericardium called fold of left vena cava [9]. A left SVC is the result of persistence of left anterior and left common cardinal veins [2, 8, 12]. Posterior cardinal veins disappear together with temporary kidneys. Subcardinal and supracardinal veins replace posterior cardinal veins stage by stage. The root of the azygos vein and common iliac veins are the two posterior cardinal veins in adults. Subcardinal vein constitutes the origin of the left renal vein [7]. In both of our cases, the connections between the hemiazygos and left renal veins occurred due to the patency of the portion of subcardinal vein between the points where it formed the renal and the hemiazygos veins.

Double SVC is a rare anomaly [4]. The incidence of double SVC in general population is 0.3% [4]. On the other hand, in patients with a congenital heart disease, this incidence rates are reported as 10% [4] and 11% [2]. Double SVC cases are especially important if the one on the left side joins the left atrium [11]. Double SVC is also important in surgery of the congenital heart diseases [8].