



Double superior vena cava. A case report

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ABSTRACT

Double superior vena cava is a rare anomaly of systematic venous return along with 0, 3 % occurrence of the general population. We account for a patient's case whose diagnosis was discovered in an unexpected way on the angioscanner and made in suspected pulmonary embolism. In fact, the agioscanner confirmed the existence of two superior vena cava with anastomosis of the left superior vein in the right auricle (coronary sinus). If isolated, this congenital anomaly has no hemodynamic repercussion.

Keywords : left ; superior ; vena ; cava ; angioscanner.

INTRODUCTION

The upper double vena cava is a rare anomaly present in 0.3% of the general population and 10% of patients with congenital heart disease, mainly inter-atrial communication [1], inter ventricular and tetralogy of Fallopien [3]. It is associated with pulmonary atresia in 6% of cases [1, 2].

MATERIALS AND METHOD

He was a 56-year-old man followed for high blood pressure who was brought to a cardiology consultation for dyspnea. The clinical examination found a dyspneic patient without cyanosis. Faced with the strong suspicion of pulmonary embolism, a CT angiography was carried out in emergency which highlighted the absence of any sign of embolism, however one noted the presence of a right superior vena cava and a 2nd vein upper left cellar each receiving an azygos vein. These two upper cell veins are of the same caliber and symmetrical (Figure 1). The upper left vena cava flows into the dilated coronary sinus (Figure 2). The right atrium is also dilated. It is actually a persistence of the left superior vena cava. The reconstructions, coronal in maximum intensity projection mode (MIP) (Figure 2), allow to visualize these different anomalies.

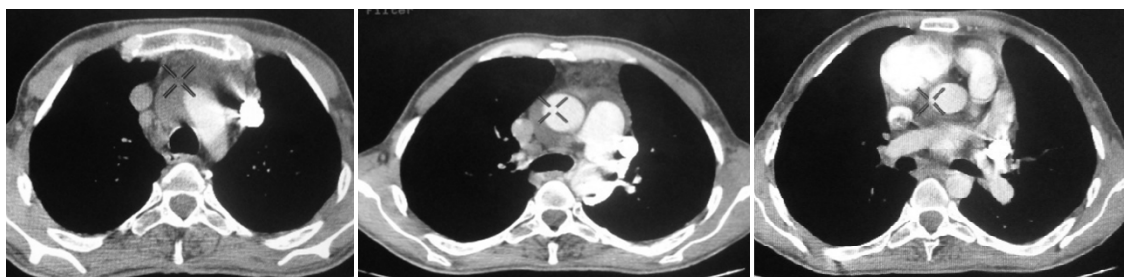


Figure 1: Chest CT scan: axial sections showing the presence of two superior vena cava each receiving an azygos vein.



Figure 2: coronal reconstructions showing the double superior vena cava with dilated aspect of the coronary sinus and the right atrium.

RESULTS AND DISCUSSION

The upper double vena cava results from the coexistence of the upper left vena cava with the upper right vena cava, an innominate vein allowing communication between the two veins in 60% of cases [3]. Sometimes there is no anastomosis between the two superior cava veins. [6] The right superior vena cava may be absent or hypoplastic or, on the contrary, well developed. In general, it has a caliber inversely proportional to that of the left superior vena cava. If it is not associated with the right superior vena cava, the systemic venous return of the upper part of the body drains through the left upper vena cava into a dilated coronary sinus [3,6].

To understand the anomalies of the systemic venous return, it is useful to know the embryological development and to determine the atrial situs.

Embryological reminder:

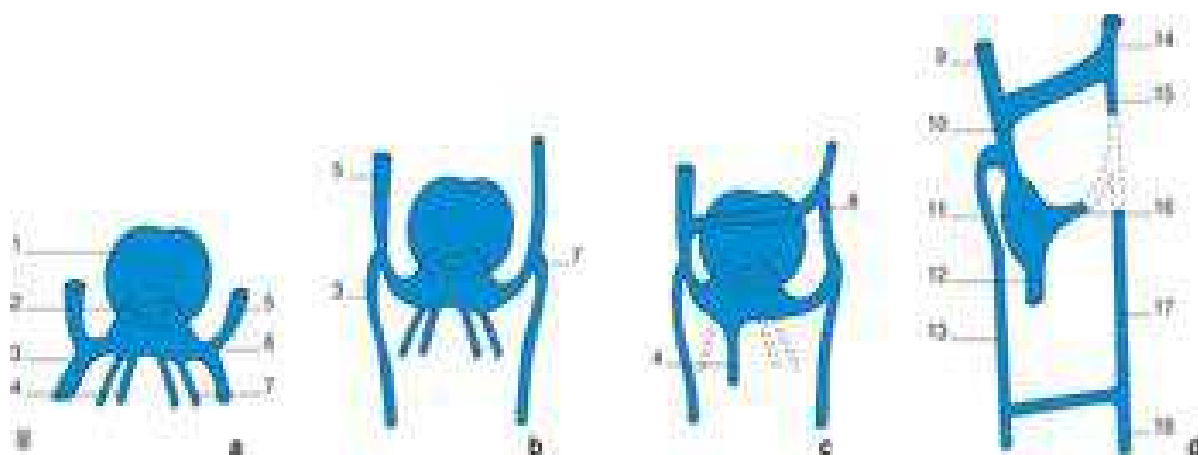


Figure 3: Embryology of the superior vena cava. 1. Primitive atrium; 2. sinus venosus; 3. posterior cardinal vein; 4. yolk vein; 5. anterior cardinal vein; 6. common cardinal vein (from Cuvier); 7. umbilical vein; 8. anastomosis; 9. right brachiocephalic venous trunk; 10. Superior vena cava; 11. right atrium; 12. inferior vena cava; 13. azygos vein; 14. left brachiocephalic venous trunk; 15. upper left intercostal vein; 16. coronary sinus; 17. hemiazygos vein; 18. accessory hemiazygos vein. at. Fourth week. b. Verticalization. vs. Seventh week. d. Eighth week

At the fifth week of fetal life, the embryonic venous circulation is composed of three main right and left veins: a) the cardinal vein giving the future cave system; b) the vitelline vein giving the mesenteric vessels and the hepatic vessels; c) the umbilical vein [6].

During embryonic life, the systemic venous return takes place via a dorsal network at the origin of the cave system. On each side of the embryo, the anterior cardinal vein followed by the Cuvier canal (or common cardinal vein) constitutes the precursor of the superior vena cava system [10]. Cuvier canals flow into the venous sinus of the heart. An anastomosis is established between the anterior cardinal veins. Subsequently, the anterior cardinal vein and the right Cuvier duct give the superior vena cava. The intercardinal anastomosis becomes the left brachiocephalic venous trunk (Figure 3). The left cardinal vein and Cuvier's canal atrophy and do not participate in the formation of the upper cave system [1, 2, 8, 10].

The persistence of the left anterior cardinal vein leads to the formation of the left superior vena cava [1, 2]. In 80 to 90% of cases, it is associated with a right superior vena cava giving a double superior vena cava. The left brachiocephalic venous trunk is absent or hypoplastic in 65% of cases [10]. In 10%, there is an isolated left superior vena cava when the right cardinal vena cava has involuted [8,10]. This vein drains into the right atrium through the coronary sinus in about 92% of cases. For the remaining 8%, it ends in the left atrium: this is the consequence of a defect in the development of the coronary sinus. This creates a right-left shunt [3, 10] causing cyanosis. This anomaly is most often well tolerated and its discovery is fortuitous [8,6]



Figure 4: Diagram of the persistence of the left superior vena cava. 1. Right brachiocephalic venous trunk; 2. Right superior vena cava; 3. Right atrium; 4. Left brachiocephalic venous trunk; 5. Upper left vena cava; 6. Coronary sinus; 7. Left atrium. [9]

The anomalies of the superior vena cava can be classified into four types [3, 5] (Figure 4): the first type is the persistence of the left anterior and common cardinal veins with the presence of intercardinal anastomosis. The second type differs from the first in the absence of intercardinal anastomosis. In the third type, only a left superior vena cava exists. In the 4th type, there is a double superior vena cava with the presence of similar azygos veins on each side. In our observation, it was a combination between the 2nd and the 4th type. [1, 3]

The persistence of the left superior vena cava is usually asymptomatic when the venous return occurs at the level of the right auricle. When this return occurs at the level of the left atrium, it can be at the origin of cyanosis and exposes to a risk of paradoxical embolism [1,7] representing one of the circumstances of discovery in adulthood by short circuit of the pulmonary filter [6,10]

The diagnosis is evoked on the chest radiography of the face which shows a rectilinear left superior paramediastinal opacity projecting on the aortic button and the aortopulmonary window [6,10]

Confirmation of this type of malformation requires trans-thoracic ultrasound and cardiac catheterization. Currently, chest CT and angio-MRI represent non-invasive alternatives [1,2]. Indeed, Echocardiography shows the left superior vena cava immediately to the left of the aorta in suprasternal incidence. The coronary sinus is dilated [12]; contrast echo and color-coded echo confirm the diagnosis by showing the abnormal flow in the coronary sinus [7].

CT and MRI show its path and its termination [10] by visualizing it in the form of a left mediastinal tubular opacity located in front of the arch, draining into the coronary sinus and presenting a vascular-type contrast enhancement kinetics [12].

CONCLUSION:

The superior double vena cava due to the persistence of the left superior vena cava is the most common of intra thoracic venous anomalies, it results from the absence of involution of the left and anterior cardinal veins [8]. Thus, its mode of discovery, usually fortuitous, reflects its good hemodynamic tolerance [3,10] and does not require any treatment.

REFERENCE

[1] H. Nèji a, H. Daoud Ketata a, D. Bouacida c, E. Daoud a, S. Mezghani b, J. Mnif c, Z. Mnif Double veine cave supérieure chez une enfant de 2 ans *Archives de pédiatrie* Volume 17, numéro 9 pages 1346-1348 (septembre 2010)

[2] S. Albay, F.Cankal, N.Cokabiyik, et al. Double superior vena cava *Morphologie* 2006 ; 90 : 39-

42 [inter-ref]

[3] J.Vazquez-Perez, A.Salvador-Sanz Anomalies des veines caves et du retour veineux systémique *Cardiologie* [11-041-G-20]

[4] S.Cosson, J.P Kévorkian, O.Roux, R.Fressonnet, A.Leenhardt, P.Beaufils veine cave supérieure gauche : un diagnostic simple par échocardiographie transthoracique. A propos d'un cas *Annales de cardiologie et d'angéiologie* vol 52, issue 3, june **2003**, 194-197

[5] J.Giebel, J.Fanghanel, S.Hauser, et al. A case of a persistent left vena cava superior with atresia of the right atrial ostium of the coronary sinus *Ann Anat* **2000**, 182 : 1916194 [cross-ref]

[6] C Rey anomalies du retour veineux systémique radiologie et imagerie médicale : cardiovasculaire- thoracique- cervicale [32-016-A-15]

[7] E.Marret, J.F.Meunier, A.M.Dubousset, D.Pariante, K.Samii Diagnostic d'une veine cave supérieure gauche persistante au bloc opératoire lors d'un cathétérisme veineux central *Annales françaises d'anesthésie et de réanimation* Vol 19 ? Num 3 pages 191-194 (mars **2000**)

[8] S.Jaafar, L.Hennequin, A.Bertal, M.Claudon exploration de la veine cave supérieure et sa pathologie radiologie et imagerie médicale : cardiovasculaire-thoracique-cervicale [32-225-F-20]

[9] E.Blondiaux, J.-N.Dacher, P.-H.Vivier, D.Sirinelli, H.Ducou le Pointe Imagerie des retours veineux systémiques et pulmonaires *Radiologie et imagerie médicale : cardiovasculaire-thoracique-cervicale* [58241]

[10] M.El Hajjam, C.Lagrange, J.Desperramons, C.Hardit, S.Binsse, P.Lacombe imagerie diagnostique et thérapeutique de la veine cave supérieure radiologie et imagerie médicale : cardiovasculaire-thoracique-cervicale [32-225-F-20]

[11] M.Ould-Ahmed, B.Mas, E. Hautbois, J.F.Garcia, P.Caroff, M.Guiavarch Pseudo-fausse route d'un cathéter artériel pulmonaire via une veine cave supérieure gauche *Annales françaises d'anesthésie et de réanimation* vol 19, Num 10 pages 745-748 (déc. **2000**)

[12] B.Kastler, C.Clair, E.Delabrousse, Y.Bernard, A.Klinkert Retours veineux systémiques et pulmonaires anormaux : aspects IRM et classification *Radiologie et imagerie médicale : cardiovasculaire- thoracique- cervicale* [32-016-A-20]

[13] Tak T., Crouch E., Drake G.B. Persistent left vena cava: incidence, significance and clinical correlates *Int J Cardiol* **2002** ; 82 : 91-93 [cross-ref]

[14] Miraldi F., Carbone I., Ascarell A., et al. Double superior vena cava : right connected to left atrium and left coronary sinus *Int J Cardiol* **2009** ; 131 : e78-e80