J Surg Med. 2019;3(10):774-776.
DOI: 10.28982/josam.630593

Journal of Surgery and Medicine
e-ISSN: 2602-2079

Situs inversus totalis with double superior vena cava: An unusual case report

Sitüs inversüs totalis ve çift superior vena kava: Nadir bir olgu

İsmail Taşkent [1], Gürkan Danışan [1], Ayşe Murat Aydın [2]

[1] Mus State Hospital, Department of Radiology, Mus, Turkey
[2] Firat University, School of Medicine, Department of Radiology, Elazig, Turkey

ORCID ID of the author(s)
İT: 0000-0001-6278-7863
GD: 0000-0003-2052-0006
AMA: 0000-0001-9619-4142

Corresponding author / Sorumlu yazar:
İsmail Taşkent
Address / Adres: Muş Devlet Hastanesi, Muş, Türkiye
e-Mail: dr.taskent@gmail.com

Informed Consent: The authors stated that the written consent was obtained from the patient presented with images in the study.
Hasta Onamı: Yazar çalışmada görüntüleri sunulan hastadan yazılı onam alındığını ifade etmiştir.

Conflict of Interest: No conflict of interest was declared by the authors.
Çıkar Çatışması: Yazarlar çıkar çatışması bildirmemişlerdir.

Financial Disclosure: The authors declared that this study has received no financial support.
Finansal Destek: Yazarlar bu çalışma için finansal destek almadıklarını beyan etmişlerdir.

Published: 10/23/2019
Yayın Tarihi: 23.10.2019

Abstract
Situs inversus totalis (SIT) with double superior vena cava (SVC) is a rare congenital anomaly. Most cases are diagnosed incidentally after imaging for other reasons. Double SVC is usually asymptomatic, unless associated with other cardiac anomalies. A 22-year-old female patient with the complaints of cough, headache, weakness, and shortness of breath was admitted to the cardiology department. The patient, who was hospitalized with a diagnosis of pulmonary embolism and pulmonary hypertension, had a history of surgical repair of atrial septal defect and ventricular septal defect 7 years ago. Contrast-enhanced multislice computed tomography (CT) of the chest was obtained in our department. CT demonstrated SIT with double SVC, with the right SVC draining into the left atrium. The variations of anomalous venous connections accompanying cardiac anomalies should be fully defined before surgery with a combined imaging approach with echocardiography and CT.
Keywords: Situs Inversus, Superior Vena Cava, Imaging

Öz
Situs inversus totalis (SİT) ve çift superior vena kava (SVK) nadir görülen Konjenital anomalilerdir. Olguların çoğunluğu başka nedenlerle görüntülemede tesadüfen teşhis edilir. Diğer kardiyak anomalilerle birlikte olmadıkça çift SVK genellikle asemptomatiktir. 22 yaşındaki kadın hasta öksürük, baş ağrısı, halsizlik ve nefes darlığı şikayetleri ile kardiyoloji kliniğine başvurdu. 7 yıl önce atriyal septal defekt ve ventriküler septal defekt cerrahi onarım öyküsü olan hasta pulmoner emboli ve pulmoner hipertansiyon tanısı ile hastaneye yatırıldı. Göğüsün kontrastlı çok kesitli bilgisayarlı tomografisi (BT) kliniğimizde çekildi. BT, SİT ile çift SVK'ı gösterdi. Çift SVK'nın sağ SVK'sı sol atriuma drene idi. Kalp anomalilerinin eşlik ettiği anormal venöz bağlantı varyasyonları ekokardiyografi ve BT ile kombine görüntüleme yaklaşımıyla ameliyattan önce tam olarak tanımlanmalıdır.
Anahtar kelimeler: Situs İnversus, Superior Vena Kava, Görüntüleme

Introduction

Situs inversus is a rare congenital anomaly that occurs due to inhibition of visceral rotation during embryonic development, with an incidence of approximately 0.01% worldwide. In situs inversus, all thoracoabdominal and retroperitoneal organs (i.e., kidneys and adrenal glands) are positioned symmetrical to their normal localizations, which is often referred to as a mirror image [1]. When both abdominal and thoracic viscera are affected, it is called situs inversus totalis (SIT) [2]. It can be associated with Kartegener triad (bronchietasis, sinusitis, and situs inversus), cardiac anomalies, genitourinary anomalies, polysplenia syndrome and intestinal rotational disorders [1,3,4].

The most common congenital abnormality of the superior vena cava (SVC) is a double SVC [2,5]. Double SVC with persistence of a left SVC [PLSVC) is rarely encountered. The prevalence in the general population is 0.3% but may reach 1.3-11% in patients with congenital heart diseases [5-8].

In 90% of cases, PLSVC connects to the right atrium via the coronary sinus, but in 8-10% of cases, PLSVC connects to the left atrium causing a right-to-left shunt, and it is in this variant that there is a potential hazard of systemic embolization of thrombus or air. Cyanosis, sepsis, and cerebral abscess may occur [1,5,9,10]. PLSVC draining to the left atrium is associated with many types of congenital heart disease but is rare if the heart is normal [7].

How to cite / Atıf için: Taşkent İ, Danışan G, Aydın AM. Situs inversus totalis with double superior vena cava: An unusual case report. J Surg Med. 2019;3(10):774-776.

Case presentation

A 22-year-old female patient with complaints of cough, headache, weakness, and shortness of breath was admitted to the cardiology department of our hospital. The patient, who was hospitalized with a diagnosis of pulmonary embolism and pulmonary hypertension, had a history of surgical repair of atrial septal defect and ventricular septal defect 7 years ago.

Her heart rate was 80 beats/min and blood pressure was 110/90 mmHg. Body temperature was 37^{o}C and hemoglobin level was 12.4 g/dL. Peripheral capillary oxygen saturation was 91%. The patient had no cyanosis.

Transesophageal echocardiography (ECHO) was performed with a Sonos 5500 using (Hewlett Packard, Inc., Andover, MA, USA) a 5-MHz omniplane probe. ECHO revealed dextrocardia. Residual ventricular septal defect was detected. Systolic pulmonary artery pressure was 83 mm Hg, consistent with pulmonary arterial hypertension.

Contrast-enhanced multislice computed tomography (CT) of the chest was obtained in our department. 64-detector CT (Aquilion, Toshiba Medical Systems, Tokyo, Japan) was used for imaging. 60 ml non-ionic contrast medium (Omnipaque, Amersham Health, Cork, Ireland) was administered via the antecubital vein with an autoinjector. CT demonstrated SIT with double SVC. The Left SVC was draining directly into right atrium, while the right SVC was draining into left atrium (right-to-left shunt) (Figure 1). There was a horizontal communicating vein between upper parts of the SVCs (Figure 2). Pulmonary trunk diameter was measured as 32 mm. Pulmonary embolism wasn't detected.

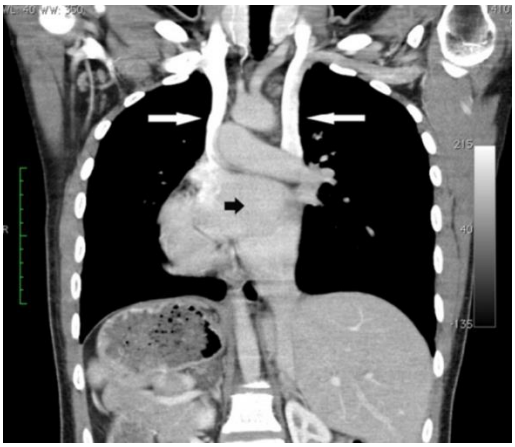

Figure 1: Contrast-enhanced chest computed tomography (CT); mediastinal window (coronal slice). Situs inversus totalis is shown. Apex of the heart is the right hemithorax. The stomach is on the right while the liver of the abdominal organs is seen on the left. Double superior vena cava is observed (white arrows). On the right superior vena cava is opened to the left atrium (black arrow).

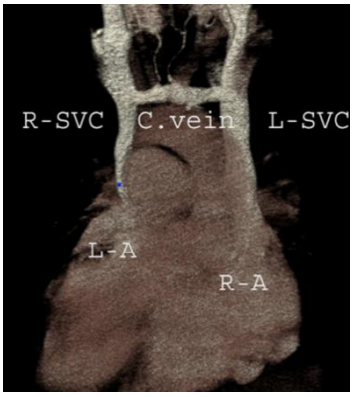

Figure 2: On three-dimensional volume rendering CT image, double superior vena cava is seen. On the right superior vena cava is opened to the left atrium. The horizontal communicating vein between upper parts of the superior vena cava is seen.

The patient was followed up 9 days in the cardiology clinic. Bosentan, sildenafil and iloprost triple combination therapy was administered to the patient for pulmonary hypertension secondary to congenital heart disease. Acetyl-salicylic acid and furosemide were also included in the treatment. The cardiovascular surgeon suggested surgery to treat the congenital abnormality of right SVC, which the patient refused. The patient was discharged with medical therapy and recommendations. The patient was later lost to follow-up.

Discussion

SIT is an unusual entity, first reported by Fabricius in 1600. SIT is usually incidentally detected on radiological images obtained due to another medical condition and does not threaten life [1,3,4].

The double superior vena cava with PLSVC is a disorder in the regression of the left anterior cardinal vein during the first weeks of embryological development [5-7]. In adults, the absence of right superior vena cava with PLSVC is rare and most patients with PLSVC have both right and left-sided SVC [10].

The presence of double SVC may be alone or concurrent with anomalies like double coronary sinus, absence of left brachiocephalic vein, atrial septal defect, common atrium, univentricular heart, transposition of large vessels, dextrocardia, conductive tissue abnormalities and horseshoe kidney [9]. Although it is a benign condition, PLSVC has important clinical implications. It may be associated with a variety of congenital malformations of the heart and great vessels, and it may technically complicate some endovascular and surgical procedures [11]. CT is a useful technique to investigate additional anomalies of SVC. Magnetic resonance imaging and venographic techniques can complement CT [5,6]. ECHO may be useful [2,5].

PLSVC rarely connects to the left atrium causing a right-to-left shunt responsible for cyanosis and heart failure. There was no evidence of cyanosis or systemic arterial desaturation in our patient.

PLSVC draining to the left atrium can result in a significant intracardiac shunt. Various surgical procedures have been reported to correct this anomaly. Some of these may include intra-atrial redirection of flow from the left SVC to the right atrium, and re-implantation of the left SVC into the right atrium, pulmonary artery, or SVC. If the LSVC is connected to the right SVC by a left innominate vein of adequate size, simple ligation is feasible. Polytetrafluoroethylene graft is a successfully used method for connecting SVCs [10,12].

The variations of anomalous venous connections accompanying cardiac anomalies such as ASD should be fully defined before surgery, preferably with a combined imaging approach with ECHO and CT.

Double SVC is usually asymptomatic unless it is associated with other cardiac anomalies. Therefore, it is diagnosed incidentally in surgery or autopsy [2]. The most commonly associated cardiac abnormality is ASD. Others include tetralogy of Fallot, coarctation of the aorta, pulmonary stenosis, interventricular septal defect [5].

Double SVC may cause enlarged mediastinum on chest radiography and the definitive diagnosis is made by CT. However, if chest CT scans are evaluated without proper attention, double SVC can be missed [2,5]. Unexpected double SVC may give rise to difficulties in venous catheterization, pacemaker insertion, radiofrequency ablation, coronary artery bypass graft, during cardiopulmonary bypass or surgery of the congenital heart diseases [2,6,9].

Although our case is a good example for showing these two anomalies together, the patient's refusal to accept the operation is a limiting factor in presenting the post-operative findings and clinical progress.

Conclusion

Situs inversus totalis with a double SVC is a rare congenital anomaly. Double SVC is benign condition unless it is associated with other cardiac anomalies. If double SVC with PLSVC draining to left atrium is detected, further cardiac evaluation with imaging methods becomes is necessary.

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This paper has been checked for language accuracy by JOSAM editors.
The National Library of Medicine (NLM) citation style guide has been used in this paper.

Suggested citation: Patrias K. Citing medicine: the NLM style guide for authors, editors, and publishers [Internet]. 2nd ed. Wendling DL, technical editor. Bethesda (MD): National Library of Medicine (US); 2007-[updated 2015 Oct 2; cited Year Month Day]. Available from: http://www.nlm.nih.gov/citingmedicine