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Case Report

Azygos and hemiazygos continuation: An occasional finding in emergency department

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ABSTRACT

Interruption of the inferior vena cava (IVC) with azygos continuation is a rare congenital anomaly, in which the IVC is interrupted below the hepatic vein and venous return beyond this point is restored by the dilated azygos and hemiazygos veins draining into the superior vena cava. A case of the interruption of the IVC with azygos/emmyazygos continuation for the absence of the hepatic segment of IVC, left renal vein duplication, and polysplenia is reported. The embryologic, clinical, and radiological significance are discussed.

The diagnosis is suggested by X-ray, but contrast-enhanced multidetector computed tomography is the method of choice to diagnose this venous anomaly and reveals the aberrant vascular structures.

Awareness of different congenital anomalies of IVC is important to surgeons and cardiologists and is necessary for radiologists to avoid diagnostic pitfalls and for preoperative planning; they should be remembered because they can influence several surgical interventions and endovascular procedures. Accidental ligation of the azygos vein is fatal and cardiac catheterization using the lower extremity vein is troublesome in patients with this condition.

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Introduction

Developmental variations of inferior vena cava (IVC) have prevalence in general population comprised between 0.07% and 8.7% and rarely cause symptoms. They are usually detected during routine examinations performed for other

reasons. The most frequent congenital abnormalities include an azygos fissure, absence of the azygos vein, aortic nipple, azygos continuation of the IVC, and partial venous return [1–5].

Failure of the union between the hepatic and prerenal segments during embryologic development results in the so-called “intrahepatic interruption of the IVC with azygos

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continuation.” Consequently, blood is shunted from the supra-subcardinal anastomosis through the retrocaval azygos vein, which is usually mildly dilated; its prevalence in general population is about 0.5%–2.1% [2–8].

This congenital anomaly could be isolated, but often it is part of more complex syndrome including for example cardiac malformations, asplenia, and polysplenia syndrome. Interruption of the IVC with azygos continuation is the second most common abnormality associated with polysplenia syndrome [1,5,9,14,15].

Currently largest spreading of contrast-enhanced multidetector computed tomography (CE-MDCT) and magnetic resonance imaging angiogram have made it easier to detect congenital abnormalities of IVC. The radiological finding is usual incidental recognized during examinations conducted for different clinical indications, without any connection to the vascular alteration. However, patients with this confirmation of venous vascular tree have a predisposition in developing deep venous and renal veins. Indeed this congenital anomaly in venous draining system lead to vascular stasis in caudal districts of venous vascular tree inducing a pressure increase in caudal venous vessels; azygos continuation is founded in 5% of patients younger than 30 years old having events of deep venous thrombosis not otherways explained. In conclusion, it is important to detect this anomaly before patients undergo cardiac catheterism, azygos ligation during thoracotomy, or porto-caval decompression surgery [9–11,13].

Case report

A 52-year-old woman was admitted to the Emergency Department for acute dyspnea and respiratory distress on April 2015. Patient history was free of any type of disease. Initially clinical suspicion of pulmonary thromboembolism (TEP) was posed; while D-dimer sampling resulted normal, hemo-gas-analysis parameters demonstrated a mild alteration of pCO₂ and pO₂. Conducted into the Radiology Department the patient undergone initially chest radiography.

Plain film showed a right paratracheal mass-like widening of mediastinum (Figs. 1a and b) well observed in the postero-anterior projection; the lateral view was unhelpful. This finding lead to a neoplastic disease or a mass-like mediastinal syndrome or vascular anomalies but anyway to rule out TEP, it was planned a contrast-enhanced chest CT.

During the basal contrast-free examination it was detected the enlargement of azygos adjacent to the descending thoracic aorta and dilated emyazygos veins; besides in caudal slices was observed a mass-like image in left hypochondrium. Therefore, the radiologist decided to extend contrast medium evaluation to upper abdomen. CE-MDCT and postprocessing images took away every doubt.

CE-MDCT imaging was performed in emergency and excluded the presence of TEP above all.

On CT examination the azygos vein was easily identified when the tubular structure, the intense contrast enhancement to the same degree as the aorta, and the continuity with the superior vena cava through the azygos arch were seen. The

mass-like aspect at chest X-ray examination was due to abnormal enlargement of azygos vein (maximum caliber about 20 mm; the normal caliber is about 3–7 mm) caused by congenital absence of infrahepatic segment of IVC (Figs. 2a–c). The dilated hemiazygos vein (maximum caliber about 10 mm; the normal caliber is about 2–5 mm) and the azygos-hemiazygos junction is well observed in CE-MDCT axial images (Fig. 2b).

Multiplanar reconstructions (MPR) and volume rendering reconstructions images explained better the anatomic relationship of vascular structures and depicted other associated findings. Absence of hepatic segment of IVC was noted; it continued as a dilated, right-sided azygos vein.

Sovahepatic veins drained directly in right atrium, while right renal vein and IVC conducted their blood in azygos vein (Figs. 2a–c; Figs. 3a–d).

Moreover, no additional congenital anomalies were seen or heart disease, except a polylobulated spleen, specifically almost 6 splenules (polysplenia) posterior to the stomach (Figs. 5a–d).

Another common vascular anomaly was found in upper abdomen: duplication of left renal vein, depicted in normal retroaortic side (Figs. 4 and 5c and d).

Discussion

IVC and azygos system originate by a complex embryogenetic process in which each segment of IVC arises differently from primitive vessels.

Cardinal venous system is the principal blood drainage tool of fetal's body. It is composed by:

- Posterior cardinal vein
- Anterior cardinal vein
- Subcardinal vein
- Superior cardinal vein
- Common cardinal vein

Anastomosis formation and regression of these conduits give rise to definitive venous vessels between 4th and 8th week of gestation [1].

IVC is embryogenically divided in 4 segments recognizing each one a different development:

- Hepatic segment: derives from hepatic sinusoids formed by vitelline veins.
- Prerenal segment: derives from right supracardinal vein.
- Renal segment: derives from anastomosis between subcardinal and supracardinal vein.
- Postrenal segment: derives from right supracardinal vein.

The azygos and hemiazygos veins originate from the last portion of the posterior cardinal veins [2]. Anomalous persistence, regression, and anastomoses of the vitelline, posterior cardinal, subcardinal, and supracardinal veins, several classic congenital variations, including IVC absence or duplication, left-sided IVC, anomalous continuation of the IVC to the thorax, and retrocaval ureter, can occur alone or in combination [3].

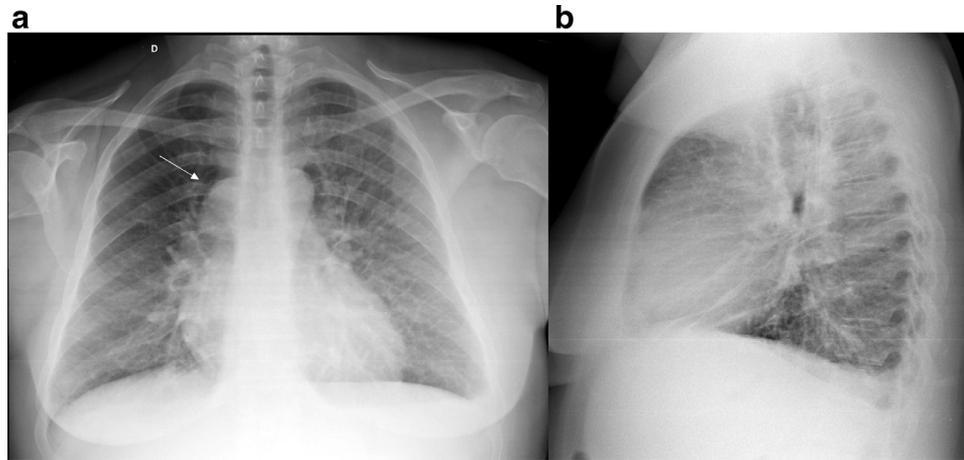


Fig. 1 – (a and b) A 52-year-old woman arrived to emergency department with acute dyspnea and respiratory distress. Chest plain radiography shows on postero-anterior projection an enlargement of the azygos vein arch on the right superior mediastinum (white arrow). It can simulate a mediastinal mass or lymphadenopathy.

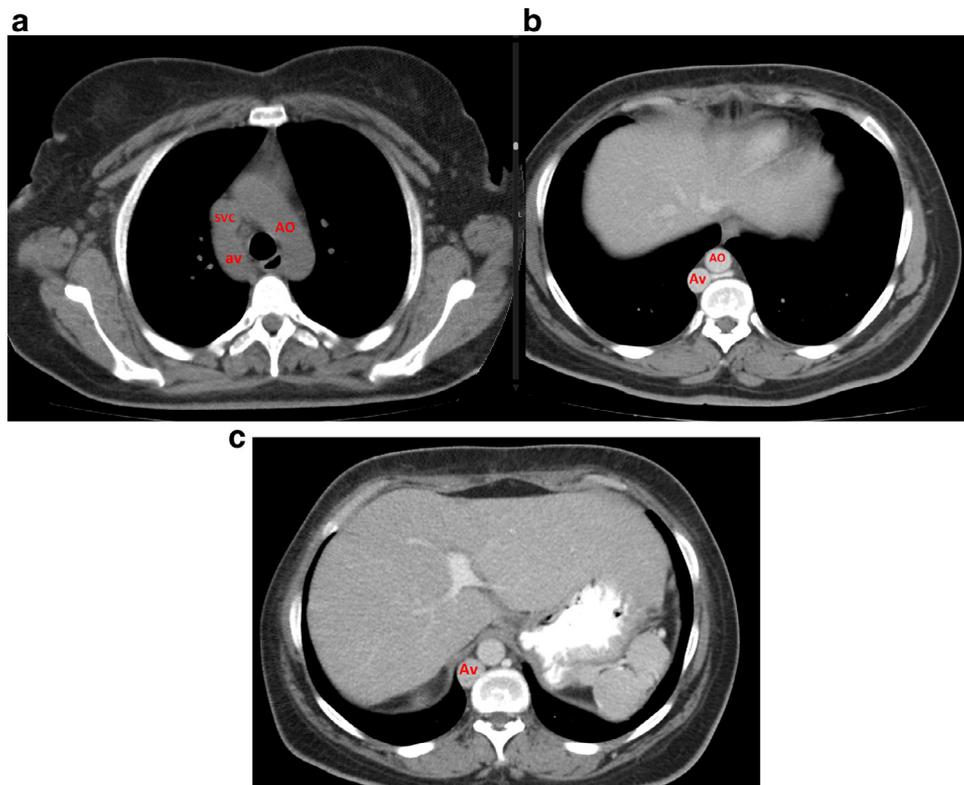


Fig. 2 – Basal CT indicates the dilated azygos vein (Av) that joins the superior vena cava (SVC) (a); contrast-enhanced MDCT scans obtained at the level of the diaphragmatic crus and the level of the azygos vein arch shows the enlarged azygos vein (Av) draining into the superior vena cava and dilated emiazygos vein draining into azygos vein (b and c). The IVC is not identified with the liver.

Failure in right subcardinal-hepatic anastomosis causes a discontinuation of IVC with or without azygos continuation [4,5]. IVC anomalies have a prevalence of 0.07%-8.7% in general population, while azygos or emiazygos continuation reported prevalence in about 1.5% (range 0.2%-3%) [5,6].

Usually this vascular anomaly is asymptomatic and encountered incidentally; however, its identification is important for planning vascular interventions and to avoid their being mistaken for diseases [7]. It is a potential mimicker of aortic pathology. Because these enlarged vessels lie parallel to

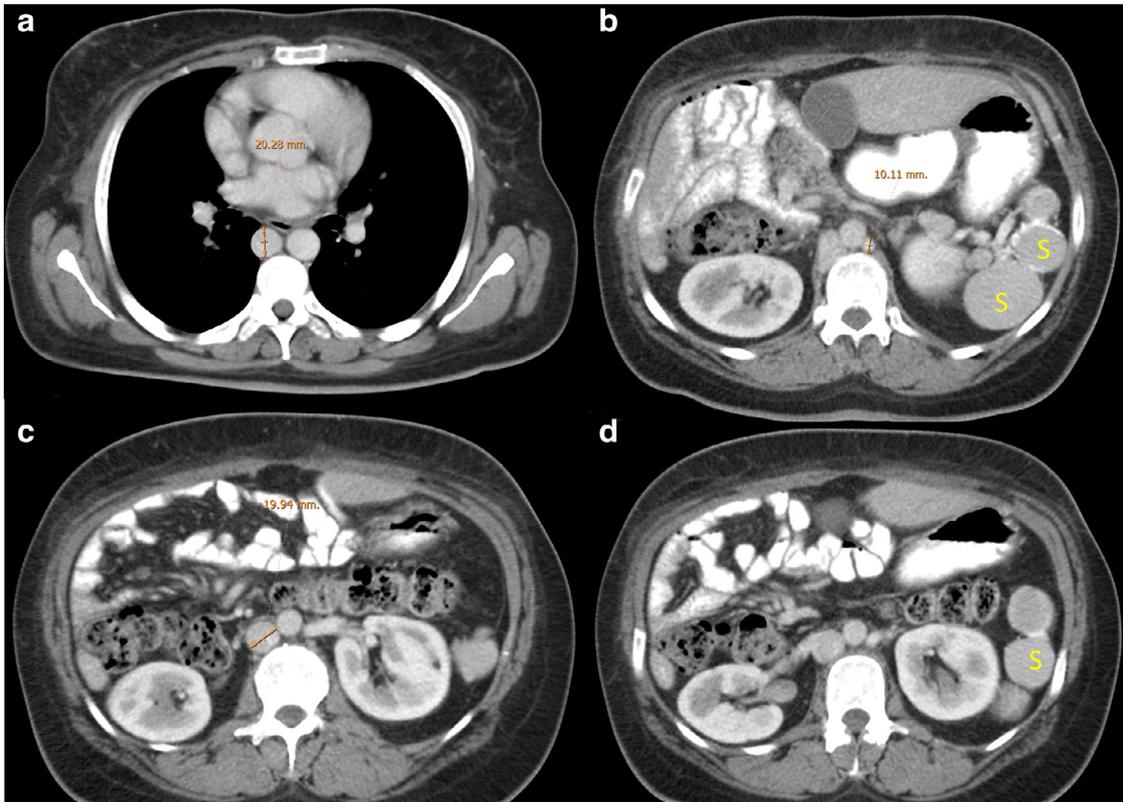


Fig. 3 – Contrast-enhanced MDCT documents an enlargement of the azygos vein about 20 mm. Normal caliber is around 3-7 mm. Emyazygos vein is dilated also with caliber about 10 mm (b-d). In the left ipocondrium are observed some small spleens (S) (polysplenia) (b-d).

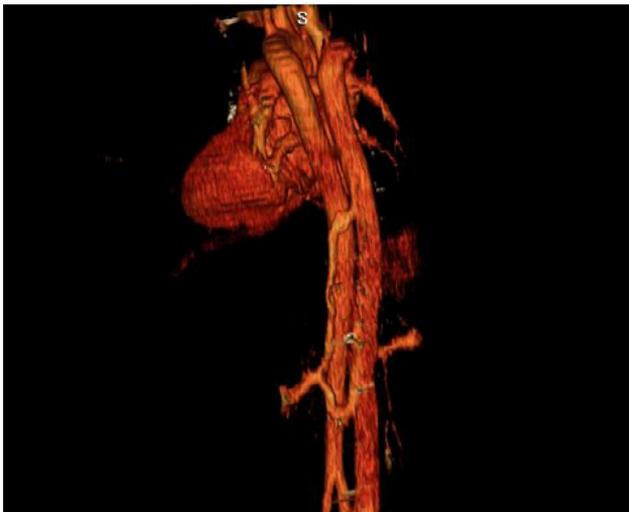


Fig. 4 – MDCT volume rendering reconstruction image shows parallel course of dilated azygos vein and thoracic aorta with emyazygos vein drained into the azygos vein. MDCT, multidetector computed tomography.

the descending thoracic aorta, they may be mistaken for aortic pathology (dissection, aneurysm, or rupture) during transesophageal echocardiography [8].

Azygos dilatation, due to its discharging function, could produce a right mediastinal widening at chest radiography, causing problems in differential diagnosis. For example, azygos and hemiazygos enlargement could be misinterpreted as right paratracheal mass or retrocrural adenopathy [9].

Azygos continuation is often part of a complex malformative syndrome including congenital heart anomalies, polysplenia syndrome, and situs anomalies [10,11]. Abnormal arrangement of thoraco-abdominal visceral and vascular structures in association with dysmorphism, is called heterotaxy syndrome or situs ambiguous syndrome which can be subdivided into polysplenia and asplenia syndromes. Because the heterotaxy polysplenia syndrome has no single pathognomonic feature, in every case comprehensive description of all clinical and radiological features is mandatory. The most consistently seen features associated with this syndrome are polysplenia, right sided stomach, interrupted IVC with azygos continuation, bilateral bilobed lungs, midline symmetrical liver, and intestinal malrotation.

Further IVC anomalies seems to induce a predisposition to deep vein thrombosis, pulmonary thromboembolism, and renal veins thrombosis, causing pressure increasing in venous vascular tree [11–13,16–18], and while these vascular malformations are rarely associated with thrombosis of the iliac and femoral veins, especially in young patients, these have been recognized as a possible risk factors for deep vein thrombosis. An interrupted IVC may cause increase blood pressure in

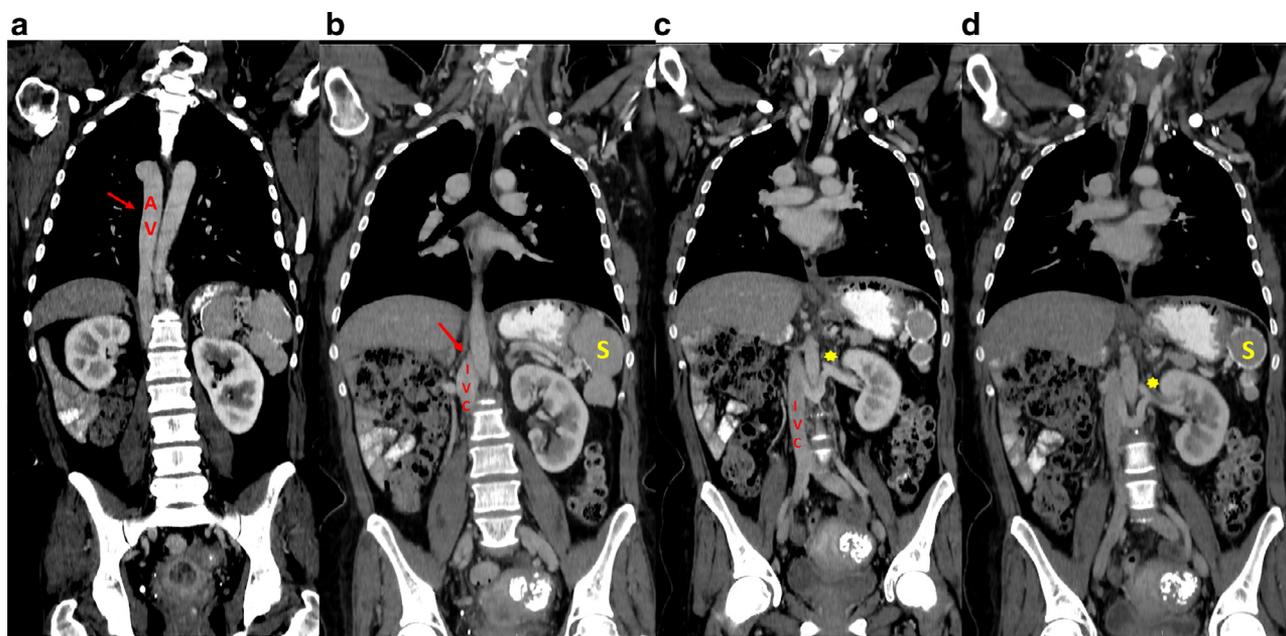


Fig. 5 – MDCT on MPR reconstructions coronal images showed a dilated azygos vein accompanied by continuation of interrupted IVC (arrows on a and b); another vessel anomaly is observed (stars): left renal vein duplication draining in the IVC. There are associated some small spleens (S) (polysplenia) visualized posterior to the stomach (c and d). MDCT, multidetector computed tomography; MPR, multiplanar reconstructions.

the lower extremity veins in the presence of insufficient collateral vessels; eventually venous stasis predisposes to deep vein thrombosis and to PTE [17].

Although most of these isolated anomalies are detected incidentally, they can have clinical implications in certain circumstances. The dilated azygos vein may be misinterpreted as a paracardiac or mediastinal mass on chest radiography. The anomaly may be associated with recurrent deep vein thrombosis of the lower limbs or with sick sinus syndrome.

There can be procedural difficulties during right heart catheterization, electrophysiological studies, cardiopulmonary bypass surgery, femoral vein catheter advancement, IVC filter placement, and temporary pacing through the transfemoral route. Awareness of the existence of these anomalies before temporary or permanent pacing can help in deciding about following an alternate route for pacing, which may avoid undue delay in an otherwise urgently needed procedure. Preoperative knowledge of this anomaly is essential because interrupted IVC can cause many problems in planning cardiopulmonary procedures and a correct diagnosis is important to avoid critical difficulties in these delicate operations. Moreover, it can lead to life-threatening complication during abdominal surgery, since ligation of the azygos vein, the only major route of venous drainage from below the diaphragm, results in death [18]. Noninvasive imaging modalities such as MDCT and magnetic resonance imaging are the most reliable methods for identification of these anomalies and differentiating it from pathologic mediastinal lesions and other vascular anomalies [18].

Currently, diffusion of modern MDCT makes diagnosis easier. Especially in emergency radiology MDCT with endovenous injection of contrast medium and postprocessing reconstruc-

tions (MIP, MPR, and volume rendering) allows a detailed study of venous system. Moreover, it has a high sensitivity in detection of pulmonary thromboembolism that should be excluded in patient with this type of malformation [13–18].

Conclusion

Although the presence of combination of these venous variations in a patient is extremely rare, it should be recognized in order to eliminate the risk for severe hemorrhage during abdominal and thoracic surgeries or the risk for recurrent embolism during the placement of an IVC filter in vascular interventional procedures. In addition, awareness of the different anomalies of the IVC is necessary for radiologists to prevent misinterpretation of aberrant vessels as paravertebral lymph node enlargement and mediastinal masses.

Conventional radiology may be the first crucial step leading to suspicion of azygos pathway changes (especially in the case of azygos continuation, hemodynamic changes, or catheter misplacement). Only by knowing the diagnostic keys to these relatively rare variations can we suspect the variation and successively confirm the diagnosis with contrast-enhanced CT scans.

Multidetector CT technique is the preferred method for imaging the congenital vascular anomalies of IVC since it is less costly, less invasive than conventional angiography, fast, easily applicable, and reliable in terms of identification of thoraco-abdominal vascular structures. Furthermore, MDCT imaging enables the acquisition of high-spatial-resolution volumetric image data during a single breath hold with the

possibility of two-dimensional (2D) and three-dimensional (3D) image after processing, which allows the visualization of complex vascular malformations in an understandable way.

Compliance with ethical standards

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

REFERENCES

- [1] Malaki M, Willis AP, Jones RG. Congenital anomalies of the inferior vena cava. *Clin Radiol* 2012;67(2):165–71.
- [2] Piciucchi S, Barone D, Sanna S, Dubini A, Goodman LR, Oboldi D, et al. The azygos vein pathway: an overview from anatomical variations to pathological changes. *Insights Imaging* 2014;5(5):619–28.
- [3] Smillie P. Imaging Evaluation of the Inferior Vena Cava. *Radiographics: A Review Publication of the Radiological Society of North America, Inc*; 2015. p. 578–92.
- [4] Kandpal H, Sharma R, Gamangatti S, Srivastava DN, Vashisht S. Imaging the inferior vena cava: a road less traveled. *Radiographics* 2008;28(3):669–89.
- [5] Awais M, Rehman A, Baloch NUA, Salam B. Multiplanar imaging of inferior vena cava variants. *Abdom Imaging* 2015;40(1):159–66.
- [6] Obernosterer A, Aschauer M, Schnedl W, Lipp RW. Anomalies of the inferior vena cava in patients with iliac venous thrombosis. *Ann Intern Med* 2002;136:37–41.
- [7] Koc Z, Oguzkurt L. Interruption or congenital stenosis of the inferior vena cava: prevalence, imaging, and clinical findings. *Eur J Radiol* 2007;62(2):257–66.
- [8] Blanchard DG, Sobel JL, Hope J, Raisinghani A, Keramati S, DeMaria AN. Infrahepatic interruption of the inferior vena cava with azygos continuation: a potential mimicker of aortic pathology. *J Am Soc Echocardiogr*. 1998;11(11):1078–83 Nov.
- [9] Kandpal H, Sharma R, Gamangatti S, Srivastava DN, Vashisht S. Imaging the inferior vena cava: a road less traveled. *Radiographics* 2008;28(3):669–89.
- [10] Schultz CL, Morrison S, Bryan PJ. Azygous continuation of the inferior vena cava: demonstration by NMR imaging. *J Comput Assist Tomogr* 1984;8:774–6.
- [11] Skeik N, Wickstrom KK, Schumacher CW, Sullivan TM. Infrahepatic inferior vena cava agenesis with bilateral vein thrombosis. *Ann Vasc Surg* 2013;27 973.e19–973.e23.
- [12] Del Prete A, Cavaliere C, Di Pietto F, De Ritis R. Situs inversus with levocardia, infrahepatic interruption of the inferior vena cava, and azygos continuation: a case report. *Surg Radiol Anat Surg Radiol Anat* 2015 [Epub ahead of print].
- [13] Trigaux JP, Jamart J, Van Beers B, Goncette L, Pringot J. Pulmonary sequestration. Visualization of an enlarged azygos system by CT. *Acta Radiol* 1995;36(3):265–9 (May 1995).
- [14] Rameshbabu CS, Gupta KK, Qasim M, Gupta OP. Heterotaxy polysplenia syndrome in an adult with unique vascular anomalies: case report with review of literature. *J Radiol Case Rep* 2015;9(7):22–37 Jul 31.
- [15] Coco D, Cecchini S, Leanza S, Ricci S, et al. Inferior vena cava duplication: incidental case in a young woman. *Case Rep Radiol* 2016;2016:3071873 Published online 2016 Apr 27.
- [16] Bass JE1, Redwine MD, Kramer LA, Huynh PT, Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics* 2000;20(3):639–52 May-Jun.
- [17] Okur A, Intepe YS, Serin HI, Yıldırım U, Mavili E. Recurrent pulmonary embolism in an asthmatic patient who had interrupted inferior vena cava with azygos continuation. *Turk Kardiyol Dern Ars* 2014;42(3):277–80 Apr.
- [18] Saito T, Watanabe M, Kojima T, Matsumura T, et al. Successful blood sampling through azygos continuation with interrupted inferior vena cava. A case report and review of the literature. *Int Heart J* 2011;52(5):327–30.