Case Report
pISSN 1738-2637 / eISSN 2288-2928
J Korean Soc Radiol 2014;70(3):213-215
http://dx.doi.org/10.3348/jksr.2014.70.3.213

대한영상의학회지

Congenital Absence of the Azygos Vein with Persistent Left Superior Vena Cava: A Case Report

좌측 상대정맥을 동반한 선천성 기정맥 결손: 증례 보고

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Absence of the azygos vein is a very rare variant of venous tributary arrangement which has been reported only in few cases so far. We hereby introduce the chest radiographic and computed tomographic findings of the congenital absence of the azygos vein with bilateral superior vena cava, incidentally detected during a follow-up for rectal cancer. The hemiazygos vein is drained into persistent left superior vena cava via left superior intercostal vein, so called the "aortic nipple".

Index terms
Azygos Vein
Bilateral Superior Vena Cava
Hemiazygos Vein
Superior Intercostal Vein
Computed Tomography

Received November 10, 2013; **Accepted** February 5, 2014
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INTRODUCTION

Azygos vein has many variations in their venous tributaries. Among them, the absence of the azygos vein is a very rare form of the congenital venous anatomic variation, and only a few cases have been reported in the literature up to recently (1-3). We report the radiologic findings of the congenital absence of the azygos vein with hemiazygos vein, which drained into the left superior vena cava via superior intercostal vein (SICV).

CASE REPORT

A 65-year-old man diagnosed as rectal cancer visited our colorectal cancer clinic. He did not show any chest symptoms.

He underwent preoperative chest posteroanterior radiograph and postoperative contrast-enhanced chest computed tomography (CT) to monitor metastasis in the thorax. On the chest radiograph, azygos arch shadow was not visible in its usual location, and there was an aortic nipple, a focal bulge adjacent to the aortic arch, representing the left SICV (Fig. 1A). The chest CT showed no azygos vein at its usual location and the left SVC was present (Fig. 1B-D). Hemiazygos vein was slightly dilated and drained into the left SVC via prominent left SICV (Fig. 1D, E).

DISCUSSION

Because of the complexity of its developmental stages, the cardinal vein system could undergo a variety number of congenital anomalies during the embryonic period (4). While in the developmental period, the cardinal system consists of anterior and posterior cardinal veins. By the eighth week, right and left anterior cardinal veins become connected by an anastomotic duct, which later become the superior vena cava on right side (from right anterior cardinal vein and right common cardinal vein) and left brachiocephalic vein on left side (from left anterior cardinal vein) (3, 4). However, when the left common cardinal vein fails to be obliterated, it becomes the left SVC (4).

The second most important system of the great thoracic veins is the subcardinal veins that form the azygos and the hemiazygos veins (4). Azygos vein is corresponded by the right subcar-

dinal vein and hemiazygos vein by the left subcardinal vein. The connection of the right and left subcardinal veins usually forms at the level of the sixth or seventh thoracic vertebra (5). The left subcardinal vein undergoes obliteration cranial to the anastomotic site, or it may persist as the accessory hemiazygos vein. The accessory hemiazygos vein is connected to the left SICV medial to the distal aortic arch (6). The left SICV is developed from the embryonic posterior cardinal veins. The left SICV then courses anteriorly beside the aortic arch to meet the left brachiocephalic vein posteriorly (6).

A persistent left SVC is considered to be the most common anomalous systemic vein-to-cardiac connection which is seen in 0.3–0.5% of general population (7). Up to 90% of the people with persistent left SVC present with the right SVC as well. About 65% of the people with persistent left SVC have no left brachiocephalic vein or atrophic one. About 20% of them show left superior intercostal vein forming a communication between hemiazygos vein and the left SVC, producing a left hemiazygos arch like the current case (7).

There have been several case reports about the radiologic findings of the absence of azygos vein (1-3, 5-7). All of these cases showed no azygos vein at the normal anatomic position and the "aortic nipple" in the dependent drainage side (i.e., left SVC, left innominate vein) on the chest radiograph. Chest CT showed the "aortic nipple" and the agenesis of the azygos vein. It also showed some cases with the right SVC only (1) or the left SVC only (2, 3, 5), and the patients with bilateral SVC (6, 7) like in this case.

Absence of the azygos vein is a very rare congenital venous anomaly, but we should take this anomaly into account when the chest radiograph fails to show azygos arch shadow on its usual location and shows the "aortic nipple" on the lateral side of aortic arch. The chest CT is the best way to confirm the agenesis of the azygos vein and the associated thoracic venous anomalies based on the chest radiograph.

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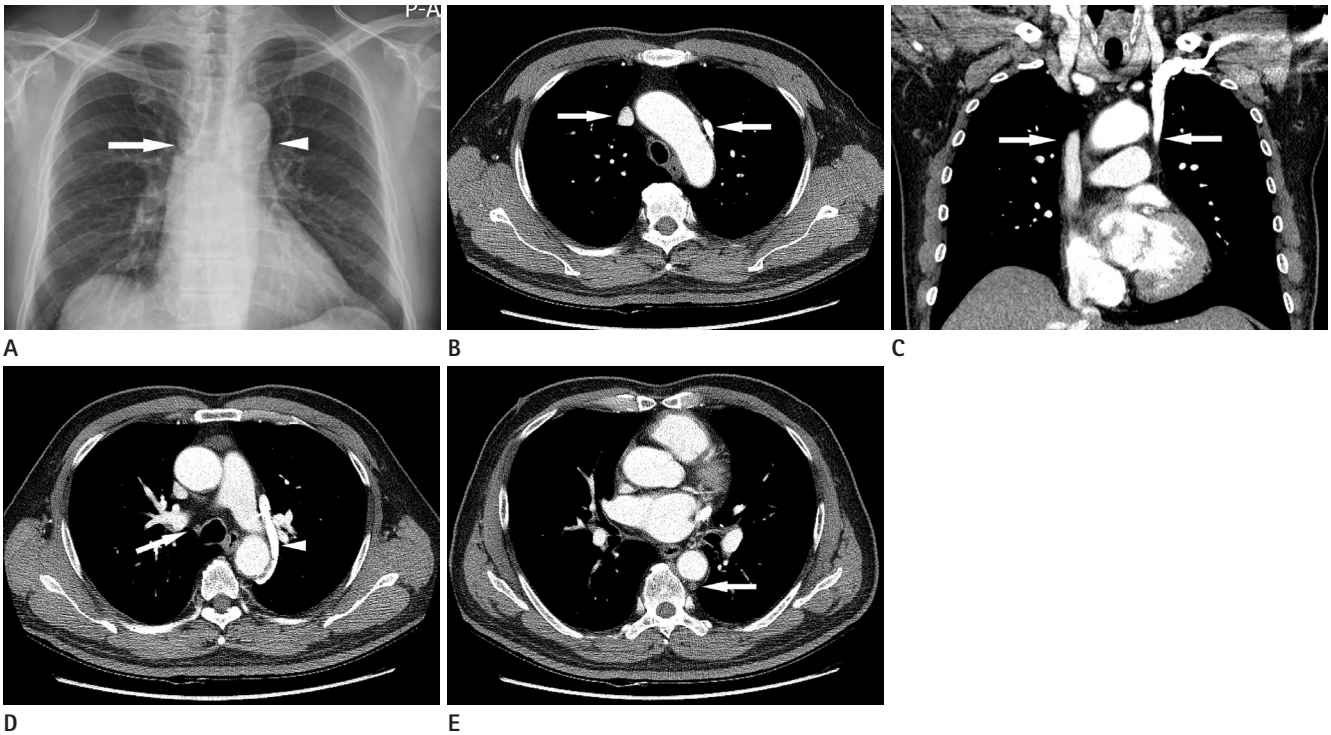

Fig. 1. A 65-year-old man with rectal cancer.
A. Chest radiograph shows no azygos arch shadow in its usual site (arrow) and shows focal bulge of aortic arch (aortic nipple) (arrowhead).
B, C. Contrast enhanced axial **(B)** and coronal **(C)** CT images show bilateral SVC (arrows).
D. Contrast enhanced axial CT scan also shows the superior intercostal vein, the "aortic nipple" (arrowhead), beside the aortic arch and the absence of the azygos vein at its usual site (arrow).
E. Contrast enhanced axial scan shows the slightly dilated hemiazygos vein (arrow).

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좌측 상대정맥을 동반한 선천성 기정맥 결손: 증례 보고

임영희 · 이정근 · 송인영 · 박정희

기정맥의 선천적 결손은 매우 드문 혈관 기형이다. 우리는 대장암 환자에서 기정맥이 없고, 한 편으로 반기정맥이 대동맥궁 측부의 좌측 상부늑간정맥, 이른바 "대동맥 유두"를 이루어 좌측 상대정맥(좌우 상대정맥이 있음)으로 혈류가 유입된 소견을 흉부 X-선 사진 및 흉부 전산화단층촬영 영상으로 관찰하였기에 보고한다.

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