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Case of Isolated Absence of the Azygos Vein: Evaluation Using Photon-counting Detector CT

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The isolated absence of the azygos vein was incidentally found on computed tomography (CT) examination in a 60-year-old female. The exact anomaly can be evaluated on high-resolution images of 0.4-mm slice thickness with low keV using photon-counting detector CT. The azygos vein, including the azygos arch, was absent, and a mildly dilated hemiazygos vein flowed to the left brachiocephalic vein through the left superior intercostal vein. A hemiazygos vein connected the left renal vein at the level of the first lumbar vertebra. This patient was the second patient to undergo evaluation using volume rendering images. High-resolution maximum-intensity projection images were useful for assessing the anatomy. Radiation dose was decreased compared with that in conventional CT.

Key words: absence of azygos vein, congenital anomaly, photon-counting detector CT

INTRODUCTION

The azygos vein exhibits numerous variations; however, reports concerning the absence of the azygos vein (AAV) are rare [1–10]. AAV is associated with congenital vena cava anomalies, including the absence of the right superior vena cava (SVC), double SVC, and persistent left SVC [4–10]. Isolated AAV is a very rare congenital anomaly. Only three cases of isolated AAV with computed tomography (CT) evidence have been reported in the literature [1–3]. In these cases, the venous anatomy, including the hemiazygos and accessory hemiazygos veins, could not be evaluated in detail. In this report, isolated AAV was incidentally found; however, the venous anatomy could be evaluated using high-resolution images from photon-counting detector CT (PCD-CT).

CASE REPORT

A 60-year-old woman with a history of vulvar cancer underwent follow-up CT examination. Contrast-enhanced CT from the chest to the pelvis was performed using PCD-CT (NAEOTOM Alpha; Siemens Healthineers) by injecting 2 mL/kg of nonionic contrast material with a scanning delay of 120 s. The tube voltage was 120 kVp, and the detector configuration was 144 × 0.4 mm automatic tube modulation on PCD-CT. Scans were performed at an IQ level of 180 (quality reference mAs), pitch range of 0.8, and iterative reconstruction strength of 2. The volume CT dose index of contrast-enhanced CT was 11.6 mGy (body wight: 79 kg, body mass index: 33.5 kg/m^2). We evaluated two-dimensional images, which included multiplanar projections of 0.4-mm slice thickness. Volume rendering and maximum-intensity projection images of the vessels were reviewed using 40-keV images.

Contrast-enhanced CT images revealed the absence of the full range of the right azygos vein, including the azygos arch (Fig. 1a–d). A mildly dilated hemiazygos vein flowed to the left brachiocephalic vein through the left superior intercostal vein (Fig. 1a–c). The right superior intercostal vein flowed into the SVC (Fig. 1a). Other right-sided intercostal veins flowed into the hemiazygos vein (Fig. 1a). The left hemiazygos vein was connected to the left renal vein at the level of the first lumbar vertebra (Fig. 1a, c, e). The left subcostal vein was connected to the hemiazygos vein (Fig. 1e). No connection between the hemiazygos vein and the left ascending lumbar vein was observed. The inferior and superior venae cavae were normal. No arterial anomaly was observed. Medical records showed no congenital heart disease. Plain chest X-ray revealed a normal aortic arch without a nipple (Fig. 2).

DISCUSSION

The azygos vein (azygos = unpaired) is mostly observed on the right side of the vertebral column [11, 12]. The azygos system comprises three interconnected major veins: the azygos vein, hemiazygos vein, and accessory hemiazygos veins, which drain the posterior

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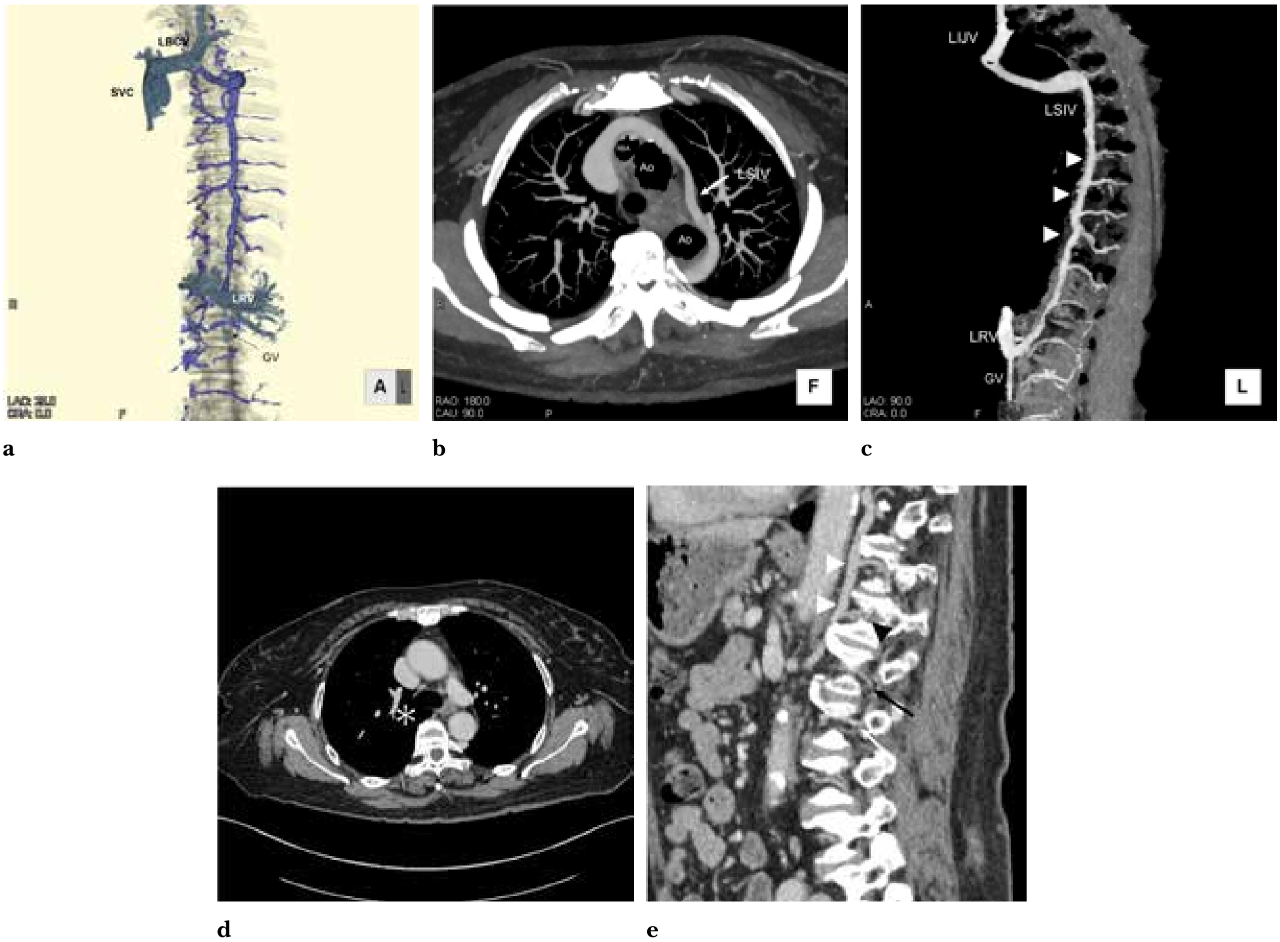


Fig. 1(a) Volume rendering image of the oblique anterior view shows the overall view of the hemiazygos system where the hemiazygos vein connects to the superior intercostal vein. SVC: superior vena cava, LBCV: left brachiocephalic vein, LRV: left renal vein, GV: gonadal vein, purple vessels: hemiazygos, intercostal and lumbar veins.

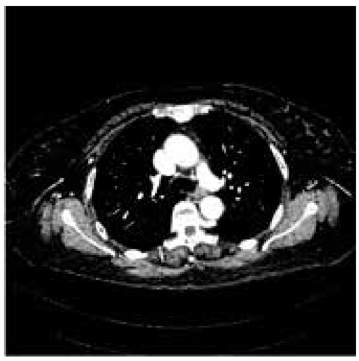
(b) Partial maximum-intensity projection image shows a mildly dilated left superior intercostal vein. Arterial density of the right brachiocephalic artery and aorta is removed from the image. LSIV: left superior intercostal vein, RBA: right brachiocephalic artery, Ao: aorta.

(c) Maximum-intensity projection image of the overall view of the hemiazygos system. White arrowheads show the hemiazygos vein. LIJV: left internal jugular vein, LSIV: left superior intercostal vein, LRV: left renal vein, GV: gonadal vein.

(d) Axial contrast-enhanced CT image (1-mm slice thickness, 70 keV) shows the absence of the azygos arch (asterisk).

(e) Sagittal image shows the first lumbar vein (black arrow) and the second lumbar vein (white arrow). No ascending lumbar vein between the first and second lumbar veins. The subcostal vein (black arrowhead) connects to the hemiazygos vein (white arrowheads).

Fig. 2 Plain chest radiograph shows normal anatomy without "aortic nipple" enlargement.



Supplemental file for review
An axial image at same level of figure 1c (0.4mm slice thickness using 40 keV.) has higher density with contrast as compared with fig. 1d.

wall of the thorax and abdomen. Developmental abnormalities of the azygos vein are due to the complex embryological origin of this venous system. The right subcardinal vein corresponds to the azygos vein, and the left subcardinal vein corresponds to the hemiazygos vein [2, 11]. An anastomosis develops between the right and left subcardinal veins, usually at the level of the sixth or seventh thoracic vertebra. The left subcardinal vein undergoes complete or partial atrophy cranial to the anastomosis, or it may persist as the accessory hemiazygos vein. The accessory hemiazygos vein meets the left superior intercostal vein medial to the distal aortic arch; the left superior intercostal vein is derived from the embryonic posterior cardinal vein [2, 3, 11, 12]. The left superior intercostal vein then turns anteriorly adjacent to the aortic arch to empty into the left brachiocephalic vein posteriorly [2, 11]. The left superior intercostal vein appears on frontal chest films as a small "nipple" projecting from the lateral aspect of the aortic knob [1, 2, 13]. The finding of "aortic nipple" enlargement on chest radiographs due to dilatation of the left superior intercostal vein on AAV is a well-known radiological feature [1, 2]. In AAV, the hemiazygos vein plays an important role in hemodynamics, particularly draining almost all the right and left intercostal veins. In our case, chest radiography at the present time was normal without "aortic nipple" enlargement; however, volume rendering and axial images revealed a mildly dilated left superior intercostal vein. If the inferior vena cava is stenosed by para-aortic lymph node metastasis in a patient with cancer, such as our case, the hemiazygos system may be dilated as collateral vessels, and an "aortic nipple" may appear on chest radiographs. In this case, knowledge of AAV and the hemiazygos system is important.

The cause of congenital AAV is uncertain. Most patients with AAV with or without SVC anomalies present with no symptoms [1–10]. Atari *et al.* reported a case of isolated AAV in a patient with lung cancer [3]. The patient underwent right upper lobectomy for lung cancer; therefore, preoperative three-dimensional angiography provided strong support for confirming abnormal vessel structures during thoracic surgery [3].

Patients with the absence of the right SVC are associated with AAV. Central venous catheterization in such patients is difficult because of its relatively tortuous course [4]. Manipulation of a guidewire can cause hemodynamic instability, arrhythmia, perforation of the heart, and tamponade [4]. In case of isolated AAV, a pacemaker and central venous line from a left-sided vein should be placed with caution because the guidewire could migrate into the superior intercostal vein. When stenosis of the inferior vena cava occurs because of thrombosis or a tumor, the azygos system is important as a collateral vascular system.

Three cases of isolated AAV underwent contrast-enhanced CT, and only one case was evaluated using volume rendering images [1–3]. AAV in this case report was incidentally found during follow-up of cancer using routine contrast-enhanced CT. Examination was fortunately performed using PCD-CT. To the best of our knowledge, this is the first case of isolated AAV to undergo evaluation using PCD-CT. PCD-CT is a novel CT protocol that solves many limitations of conventional energy-integrated detectors [1–7]. It uses semiconductor materials to generate electronic signals from incident X-ray photons. The beneficial characteristics of PCDs include the absence of electronic noise, increased iodine signal-to-noise ratio, improved spatial resolution, and full-time multi-energy imaging [14–16]. Promising PCD-CT applications involve anatomical imaging, where exquisite spatial resolution adds clinical value, and applications require multi-energy data simultaneously with high spatial and/or temporal resolution [14–16]. PCD-CT can provide full-time multi-energy data. Because we could obtain high-resolution images of 0.4-mm slice thickness at 40 keV, an exact anatomical diagnosis was possible. The left-sided azygos venous system (hemiazygos vein) was evaluated using ultrathin slice thickness with high contrast. High-resolution two-dimensional images, including maximum projection reconstruction, multiplanar, and volume rendering images, can be used. Normally, the azygos and hemiazygos veins are formed by the union of the ascending lumbar and subcostal veins [17, 18]. The azygos venous system does not directly connect to the ascending lumbar vein because the azygos venous system is located at the medial sympathetic line and the ascending lumbar vein is located at the precostal line [18]. In our case, the hemiazygos vein was connected to the subcostal vein, whereas the ascending lumbar vein did not connect to the hemiazygos vein. In PCD-CT, the radiation dose is decreased compared with that in conventional CT. The radiation dose of this CT examination decreased by 24% compared to conventional CT examination in previous hospital despite gaining weight of 10 kg.

Isolated AAV is a very rare congenital anomaly; however, it can be incidentally detected using non-invasive radiological examinations, such as CT and magnetic resonance imaging. Therefore, determining the exact anatomical diagnosis and knowledge of AAV are important before performing interventional procedures, such as central venous line placement and pacemaker implantation. When a central venous line from the left jugular vein is inserted, the risk of catheter migration into a dilated left superior intercostal vein and hemiazygos vein may increase.

In conclusion, we reported the first case of isolated AAV evaluated using PCD-CT. The exact anatomy of AAV can be diagnosed using high-resolution images from PCD-CT.

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PATIENT CONSENT

Informed consent for publication of the case was obtained from the patient.

COMPETING INTERESTS

The authors declare that they have no relevant conflicts of interest.

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