

Segmental Agenesis of the Azygos Vein: A Case Report and Literature Review

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Abstract

Case Report

Congenital anomalies of the azygos venous system are uncommon and usually asymptomatic. This report describes a segmental agenesis of the superior azygos vein associated with an empty azygos fissure. We present the case of a 60-year-old woman with a suspected pulmonary mass on chest radiography. Contrast-enhanced thoracic computed tomography (CT) demonstrated an azygos fissure devoid of its usual vascular content. The normal azygos arch was absent above the level of T7–T8. A venous channel was identified along the left posterolateral aspect of the descending thoracic aorta, continuing superiorly above the aortic arch and draining into the left brachiocephalic vein. The inferior azygos segment appeared to communicate with the left-sided venous pathway around T7–T8, suggesting drainage through the hemiazygos and accessory hemiazygos system and the left superior intercostal vein. The coexistence of an empty azygos fissure and a suspected segmental agenesis of the superior azygos vein represents an unusual venous configuration that may reflect a complex developmental anomaly of the azygos system.

Keywords: Azygos vein, azygos fissure, agenesis, congenital anomaly, hemiazygos vein, left superior intercostal vein, computed tomography.

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INTRODUCTION

The azygos venous system is a network of veins that serves as an accessory pathway between the superior vena cava (SVC) and the inferior vena cava (IVC), draining the thoracic wall and part of the posterior mediastinum into the SVC [1]. This system comprises three main veins: the azygos, hemiazygos, and accessory hemiazygos veins.

The azygos vein originates at the junction of the right ascending lumbar and right subcostal vein and enters the thorax through the aortic hiatus. It then ascends along the right anterolateral aspect of the thoracic vertebrae, arches superior to the right main bronchus, and drains into the SVC at the T4–T5 level [2]. The hemiazygos vein ascends along the vertebral column on the opposite side behind the aorta, draining into the azygos vein at the T8–T9 level. After communicating with the accessory hemiazygos vein, the left superior intercostal vein (LSIV) arches anteriorly along the lateral margin of the aortic arch and joins the left brachiocephalic vein [3].

The azygos venous system develops from a complex embryologic network involving the posterior cardinal, supracardinal, and anterior cardinal veins. Failure of development of the cranial portion of the right supracardinal vein results in agenesis of the azygos vein [4,5]. The association of an empty azygos fissure with absence of the superior azygos segment has only rarely been discussed.

CASE REPORT

A 60-year-old female patient was referred for a contrast-enhanced thoracic CT scan to investigate thoracic pain and weight loss, with a suspected pulmonary mass on chest radiography. CT examination incidentally revealed an azygos fissure without its usual vascular content (Figure 1). No azygos arch was identified in the usual right paratracheal location.

A venous structure was visible only at the T7–T8 level, where it appeared to communicate with a prominent left-sided paravertebral vein. This vessel ascended along the posterior left border of the descending thoracic aorta, passed superior to the aortic

arch, and drained into the left brachiocephalic vein (Figure 2).

Based on its anatomical course, this left-sided venous pathway was considered to correspond to a prominent hemiazygos and accessory hemiazygos system, providing compensatory drainage in the absence

of the superior segment of the azygos vein through the left superior intercostal vein.

The superior vena cava was normally developed, and no additional vascular anomalies were identified. Furthermore, the patient had no history of pneumothorax, pleural effusion, thoracic surgery, or any other acquired condition that could account for secondary migration of the azygos vein.

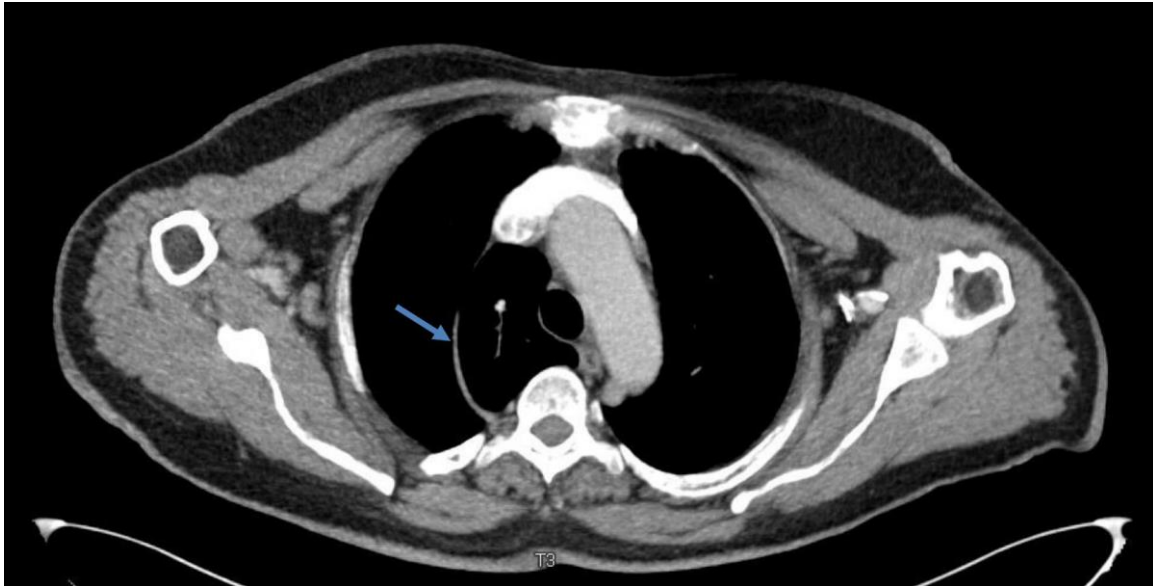


Figure 1: Axial chest CT image with MIP reconstruction showing an empty azygos fissure (arrow) and the absence of the azygos arch in its normal right paratracheal location



Figure 2: Axial chest CT image with MIP reconstruction at the level of the aortic arch showing a dilated left superior intercostal vein (arrow) draining into the left brachiocephalic vein, which provides compensatory drainage in the setting of agenesis of the superior segment of azygos vein

DISCUSSION

Congenital agenesis of the azygos vein is a rare anatomic variation of the thoracic venous system, usually discovered incidentally on cross-sectional imaging. It can occur in isolation or coexist with other venous anomalies, reflecting the intricate embryonic

development of this system [1]. The hemiazygos and accessory hemiazygos veins play important roles in hemodynamics by draining into the left brachiocephalic vein or the persistent left SVC via the prominent LSIV [6].

Only a limited number of cases have been reported in the literature. In 2000, Arslan *et al.*, reported what was considered the sixth case of azygos vein agenesis and the fourth case with detailed radiological findings [6]. They reported a complete agenesis of azygos vein and a venous drainage ensured by a prominent hemiazygos vein continuing as the LSIV before draining into the left brachiocephalic vein. More recently, Yeung reported another case of congenital absence of the azygos vein associated with enlargement of the left superior intercostal vein, resulting in the so-called "aortic nipple" sign on chest radiography [7]. Similar to our patient, venous return was ensured by the hemiazygos and accessory hemiazygos system.

A wide spectrum of congenital anomalies involving the azygos venous system has been described in the literature. These include azygos lobe, azygos continuation of the IVC, persistent left SVC, duplication of the SVC, and partial or complete agenesis of the azygos vein [2].

The azygos fissure is another uncommon congenital anomaly resulting from an abnormal migration of the azygos vein during embryogenesis. Instead of coursing medially over the apex of the right lung to join the SVC, the azygos vein penetrates the upper lobe of the right lung, carrying a double layer of pleura and giving rise to the azygos fissure and the so-called azygos lobe [8]. Despite its denomination, the azygos lobe does not constitute a true accessory pulmonary lobe, as it lacks an independent bronchovascular supply. Its prevalence has been estimated to range from 0.1% to 1% and it is usually detected incidentally on chest radiography or computed tomography [2].

Although the azygos fissure is generally considered a benign incidental finding, cases of empty azygos fissure have been reported. This entity is classically regarded as an acquired condition resulting from migration of the azygos vein following pneumothorax, pleural effusion, lung collapse, or thoracic surgery [9]. In the present case, the absence of an obvious acquired cause raises the possibility of a congenital origin. The coexistence of these findings highlights the complexity of congenital anomalies involving the azygos venous system and emphasizes the importance of careful assessment of mediastinal vascular anatomy on cross-sectional imaging.

CONCLUSION

This case highlights a potentially rare association between an empty azygos fissure and

segmental agenesis of the superior azygos vein. Recognition of such variations is essential to avoid diagnostic pitfalls and to facilitate thoracic surgical and interventional planning.

Declarations

Conflict of interest: None.

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