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CASE REPORT PEER REVIEWED | OPEN ACCESS

Absence of azygos vein with persistence of left vitelline vein

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ABSTRACT

Anatomical anomalies often reflect congenital malformations or developmental defects. We describe a cadaver in which the azygos vein was absent, coupled with the presence of an accessory atrio-hepatic venous connection that we attribute to persistence of the embryologic left vitelline vein. The formation of the azygos vein from the supracardinal vein, and the regression of the left vitelline vein, both occur around seventh week of development. The left vitelline vein connects the left and quadrate lobes of the liver to the developing heart. Although the absence of the azygos vein and adult persistence of the left vitelline vein have been reported independently, this is the first description of these two variants in combination.

Keywords: Azygos, Congenital variation, Left hepatic vein, Vitelline

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INTRODUCTION

The azygos vein drains the right posterior thoracic and abdominal wall via several tributaries including the segmental posterior right intercostal veins. It is located in the posterior mediastinum, and it arches anteriorly over the right main bronchus to join the superior vena cava. The hemiazygos vein, which drains portions of the left thoracic wall, crosses the midline around T8-T9 and empties into the azygos vein [1].

Early in development, the vitelline system of veins drains the yolk sac, and supplies the initial nourishment of the fetus and to the developing heart. The vitelline veins ultimately contribute to formation of the portal system, liver sinusoids, and portions of the inferior vena cava. The left vitelline vein initially connected the liver to the sinus venosus, which later develops into the right atrium and coronary sinus. This connection is lost around seventh week of development [2].

CASE REPORT

During routine dissection of the chest cavity of an 87-year-old female cadaver at the New York Institute of Technology College of Osteopathic Medicine no azygos

vein was found to be present. The intercostal veins did not enter into a collecting vein on the right side (azygos), but rather independently crossed over the spine to enter the left chest and drain into a large vessel (hemi-azygos) (Figure 1). This enlarged hemi-azygos vein drained proximally into the left brachiocephalic vein. No other venous anomalies were identified in the chest and on subsequent examination of the abdomen, no anomalies were found in the inferior vena cava system.

When the heart was removed from the pericardial sac, an accessory opening (13.20 mm by 10.44 mm) was identified to the left side of the caval orifice of the right atrium (Figure 2). This opening communicated through the diaphragm and ended in the liver parenchyma. This anomalous cardiac opening was located on diaphragmatic surface of the right atrium, adjacent to the branch point of the right marginal artery from the right coronary artery. This vein was dissected, and it terminated in the left and quadrate lobes of the liver. This vascular connection between the liver and right atrium resembled the original pathway of the left vitelline vein. Histological examinations of the structure at three distinct places; the entrance to the right atrium, the crossing through the diaphragm, and in the left lobe of the liver demonstrated normal venous endothelium and no cellular abnormalities of note in the adjacent liver parenchyma. The inferior vena cava was in its expected position and the hepatic vein entered it appropriately. Further examination of the inferior vena cava in the abdomen was unremarkable down to and including the bifurcation at the common iliac veins.

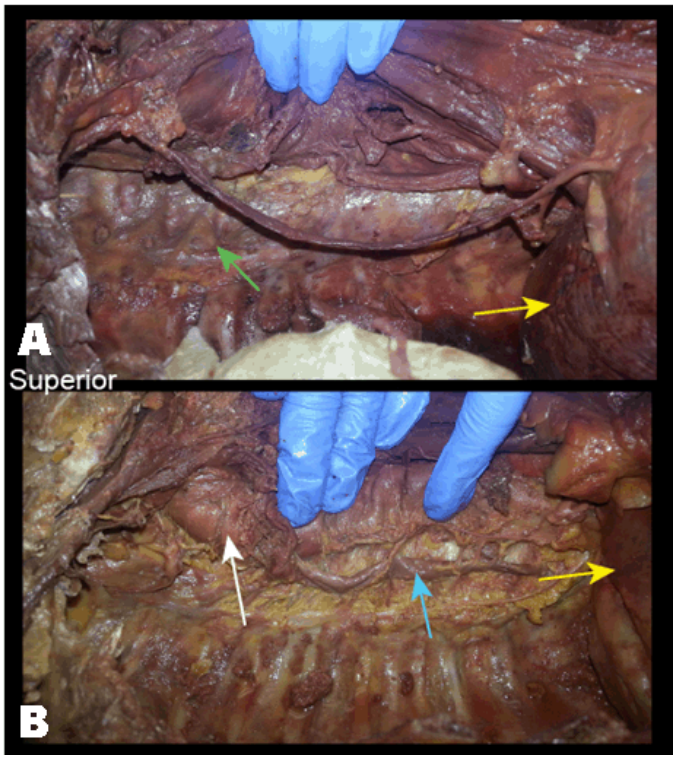

Figure 1: (A) The absence of the azygos vein and (B) Enlarged hemiazygos vein. The right posterior intercostal veins (green arrow) cross over at midline and drain into the hemiazygos vein. The hemiazygos vein (blue arrow) runs lateroposterior to the aorta (white arrow) and drains proximally into the left brachiocephalic vein. The diaphragm is labeled with a yellow arrow.

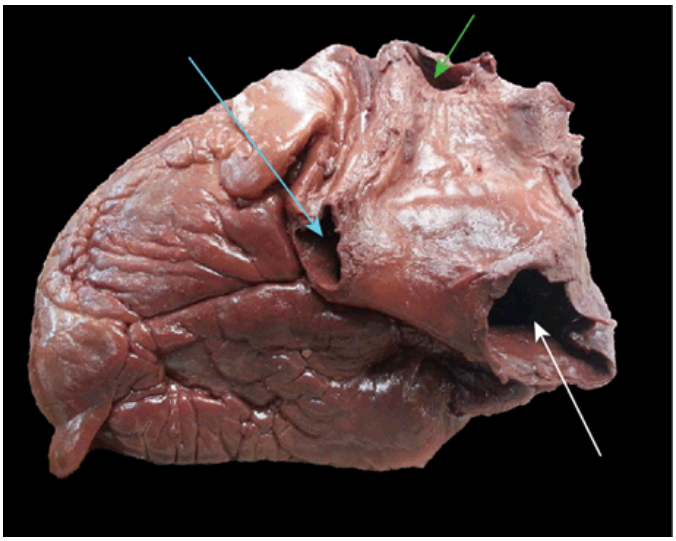

Figure 2: The openings into the right atrium. The opening of the superior vena cava (green arrow) measured 23.78 x 26.81 mm. The opening of the inferior vena cava (white arrow) measured 13.54 x 13.54 mm. The accessory opening (blue arrow) measured 13.20 x 10.44 mm.

DISCUSSION

The vitelline system of veins arises from the developing yolk sac, and ultimately drains the gastrointestinal tract and organs. The vitelline system is intimately associated with the liver parenchyma, and it gives rise to the sinusoids. The vitelline plexus condenses into the left and right vitelline veins, which drain from the liver into the right and left sinus horns respectively [3]. The connection between the left vitelline vein and the developing heart (hepato-cardiac channel) regresses around seventh week. Additionally, the left vitelline vein, in the intra hepatic region, forms the left hepatic vein that with the right and middle hepatic veins drain the liver parenchyma into the inferior vena cava [1, 2]. The right vitelline vein derivatives are maintained in the adult as the connection between the liver and heart, forming the proximal-most portion of the inferior vena cava and right hepatic vein [4]. The portions of the vitelline system caudal to the liver develop into the portal system of veins.

The posterior and anterior cardinal veins are the earliest system to drain the developing head, neck, and body. Around week-7, a pair of supracardinal veins arises from the posterior cardinals and grows medially

to drain the body wall via the intercostal veins [5]. The left supracardinal vein does not connect to the developing heart, but instead drains into the right supracardinal vein, which anastomoses with the superior vena cava. The left supracardinal vein ultimately forms the hemiazygos vein, and the right supracardinal vein forms the azygos vein [4].

We believe the combination of the absent azygos vein and persistent left vitelline vein resulted from an embryogenic event occurring around seventh week of development. During this crucial time period, the left vitelline vein normally disintegrates, and the azygos vein develops.

There have been several reports in literature describing a radiographic or cadaveric absence of the azygos vein [6–10]. This is often associated with dilation of the hemiazygos vein, draining into an enlarged left superior intercostal vein, which is commonly referred to as the "aortic nipple" [6, 8, 9]. Also consistent with the presently describing variation, several cases have been described where the left hepatic vein opened directly into the right atrium, often independent of the coronary sinus [11–15]. The left hepatic vein is a derivative of the left vitelline vein, and the terminology is synonymous with "persistent left vitelline vein" [13]. While independently, both anomalies have been described, a review of literature has failed to reveal prior description of the combination of an absent azygos vein with a persistent left vitelline vein.

CONCLUSION

The development of the azygos vein from the supracardinal vein, and the regression of the left vitelline vein are embryonic events that occur around seventh week. We describe an anomalous absence of azygos vein and persistence of left vitelline vein; both anatomic variations have been described separately, however, this is the first description of these variants in combination. We believe this combined vascular anomaly resulted from an embryogenic event occurring around week-7 of development and propose the following mechanism for the combined occurrence. Many vascular developmental events are dependent upon hemodynamic or pressure phenomenon. If there were a failure for the right anterior and posterior cardinal veins to combine properly to form the right common cardinal vein, then the azygous vein would fail to develop. This would also lead to a pressure backup into the vitelline plexus and then alter the hemodynamics in the remainder of the vitelline system and left horn of the sinus venosus, leading to enlargement of the forming hemi-azygos system and left hepato-cardiac channel (left hepatic vein).

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Bennett Futterman – Substantial contributions to conception and design, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published
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Guarantor
The corresponding author is the guarantor of submission.

Conflict of Interest
Authors declare no conflict of interest.

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SUGGESTED READING

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