REVIEW ARTICLE

Azygos Venous System Variations: A Clinical Review

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ABSTRACT

Aim: To study and analyze the common as well as rare anatomical variations of the azygos venous system along with their embryological aspect available in the literature.

Background: The azygos system of veins comprises azygos, hemiazygos, and accessory hemiazygos veins that act as a connecting channel between the inferior vena cava and superior vena cava.

Results: The review of research studies of the azygos venous system in the literature and in the cadaveric studies demonstrated a wide range of variations in the origin, course, number of tributaries, and termination. The results may contribute to the anatomical knowledge and training of students as well as clinicians in addition to supporting future research.

Conclusion: Anatomical variations of the azygos venous system can affect patient outcomes indirectly. Therefore, anatomical knowledge of all possible variations of the azygos venous system should be clearly understood.

Clinical significance: Each anomaly of the azygos venous system along with its embryology and clinical significance is of utmost importance for radiologists, surgeons, and clinicians in their respective fields.

Keywords: Azygos, Embryology, Review, Radiologists.

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BACKGROUND

Azygos venous system involves three major veins such as azygos, hemiazygos, and accessory hemiazygos veins draining the major part of the posterior abdominal and thoracic wall.¹ The azygos vein is usually positioned right of the vertebral column and forms anatomical connection of the superior and inferior vena cava.^{2,3} The azygos vein is formed by the union of the right ascending lumbar vein and right subcostal vein at T12 vertebra.^{4,5} It ascends in the posterior mediastinum and arches over the hilum of the right lung and opens into the posterior aspect of the superior vena cava, just superior to its pericardial incorporation.⁶ In some cases, it may drain into the right brachiocephalic or subclavian vein or into the intrapericardial part of superior vena cava of the right atrium.⁷ Azygos vein may show normal physiological enlargement in supine position and in overhydration or mid-trimester of pregnancy or in pathological conditions like renal failure.⁸⁻¹⁰ The hemiazygos vein usually originates from a common trunk formed by the union of the left ascending lumbar vein and the left subcostal vein. It ascends in the posterior mediastinum on the left side of the vertebral column and crosses to the right side and terminates in the azygos vein at the level of ninth thoracic vertebra.¹¹ The accessory hemiazygos is located in the fourth or fifth left intercostal space. It descends on the left side of the vertebral column and terminates in the azygos vein at the eighth thoracic vertebra¹² or in 1–2% of cases drains into the left brachiocephalic vein.^{13,14}

Results

The present study is done to review all the possible congenital as well as acquired variations of azygos venous system reported in the literature. Embryological anomalies of systemic venous system comprise 0.07–8.7% in general population and are rarely causing symptoms. They are accidentally detected during routine examination done for other reasons. The most common congenital

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anomalies are the complete absence of azygos vein, aortic nipple, and azygos continuation of the inferior vena cava.^{15–18}

Positional Changes with Age

As age advances, the vertebral column reacts to mechanical stress and strains by osteophytosis. In thoracic vertebral column, the aorta on the left side inhibits formation of osteophytes on that side, but in lower thoracic and lumbar region, as the aorta is present in midline, osteophytes develop symmetrically on both sides. The asymmetrical development of osteophytes on the right side in the thoracic region deviates the azygos vein to the left side.¹⁹

Anomalies of Azygos Vein Associated with Anomalies of Superior Vena Cava and Inferior Vena Cava

Occasionally, the anomalies of the azygos vein are associated with the anomalies of superior and inferior vena cava. A rare case of complete absence of azygos vein on right as well as left side was diagnosed. It was associated with the presence of a left superior vena cava as diagnosed on computed tomography.²⁰ Sometimes, there is no superior vena cava either on right or left side and its association with azygos continuation of inferior vena cava has been reported.²¹

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Anomalies of Hemiazygos and Accessory Hemiazygos Veins

Sometimes, hemiazygos or accessory hemiazygos may be absent. They may also show variation in their formation, course, termination, or tributaries. An interazygos, which is transverse segment of the hemiazygos crossing the vertebral column may pass between the esophagus and descending thoracic aorta.²² The absence of hemiazygos vein may be associated with the presence of transvertebral communication passing in front of the aorta known as pre-aortic interazygos vein²³ or absence of hemiazygos associated with presence of two parallel paravertebral longitudinal venous channels known as right and left azygos vein²⁴ has been reported.

Anomalous Tributaries

A very rare anomaly showing the opening of all pulmonary veins into the azygos vein on echocardiography has been reported.²⁵

Agenesis of the azygos vein occurs when superior segment of the right supra cardinal vein failed to develop. The hemiazygos and accessory hemiazygos vein present on the left side took over the function of azygos vein and the condition remained asymptomatic.²⁶

Anomalous Positions of Azygos Vein

Variations in the position of the azygos vein can cause intraoperative complications for the surgeons. An anomalous position of azygos vein covering the sympathetic chain can indirectly cause a significant risk in endoscopic thoracic sympathectomy.²⁷ The azygos vein in the midline position may possibly be due to advancing age.²⁸

Idiopathic Azygos Aneurysm

It may mimic mediastinal mass and has to be confirmed radiographically.²⁹

Azygos Lobe and Aortic Nipple

Azygos lobe is seen in 0.4–1% of the population. It indicates the incomplete medial migration of the right posterior cardinal vein that is the precursor of the azygos vein. This anomaly can be diagnosed on CT scan that shows azygos vein more laterally than usual anterior arching before draining into the superior vena cava.³⁰ Aortic nipple is seen in about 10% of the normal patients and occurs when the left superior intercostal vein draining left second, third, and fourth left-posterior intercostal veins drain into the left brachiocephalic vein forming aortic nipple. It can be seen as a soft tissue density adjacent to the lateral border of the aortic knob on the frontal radiograph and as a contrast-filled vessel along the left lateral border of the aorta. It can be associated with the congenital absence of azygos vein.³¹

Reversed Azygos Venous System

A rare variation showing reversed azygos system with azygos shifted to the left side and ultimately draining into the left brachiocephalic vein. The accessory hemiazygos and hemiazygos vein were seen on the right side of the vertebral column.³²

Multiple Transvertebral Communication

Multiple transvertebral connections between azygos and hemiazygos veins were seen instead of two.³³

Congenital Anomalies Associated with Other Medical Conditions

Azygos continuation with infrahepatic interruption of the inferior vena cava is a congenital anomaly (prevalence 0.6%) in which there occurs failure of the union of hepatic and prerenal segments. The venous blood is shunted from supra–subcardinal anastomosis through the azygos vein.³⁴ This anomaly may be seen in cases of other congenital anomalies like congenital heart disease, asplenia, or polysplenia syndrome.¹⁰ This congenital condition is of utmost importance in cardiac catheterization procedures like balloon dilatation, stent placement, or even in other surgical procedures like azygos vein ligation during thoracotomy or portocaval depression surgery.³⁵

Acquired Variations

The contour and size of the azygos vein and mediastinal pedicle on chest X-ray can provide useful hemodynamic information in clinical condition like over-infusion or renal failure with consequent fluid overload.^{8,9} There occurs 0.5 cm variation in the vascular pedicle size corresponding to an increase of 1L of circulating fluid.

DISCUSSION

The results obtained in the present review validate the proposed hypothesis regarding wide variable patterns of venous drainage system. Azygos system of veins largely represents the postcardinal veins in an embryo, which persist only at the commencement of the azygos system after birth.³⁶ The complex development of veins can cause many variations.⁶ Thoracic portion of the azygos vein is the most vulnerable part for wide variations of azygos venous system.³⁷ Anomalies of the azygos venous system are important, especially when CT of the mediastinum is performed as these can be easily confused with an aneurysm, lymphadenopathy, or other mediastinal masses.³⁸⁻⁴⁰ Anatomical variations of azygos vein can be classified into three types. Type I two vertical parallel veins with no connection between them. Type II – an anastomotic connect ion dorsal to the aorta between azygos vein and hemiazygos vein. Type III - a single azygos vein located at the midline.⁴¹ Type II is considered as the most common variant and Type I is taken as the rarest one. 33,40,42-44 Our present review of literature demonstrated that the most common prevertebral transverse anastomosis between azygos and hemiazygos vein is at the level of ninth thoracic vertebra and anastomosis between accessory hemiazygos and azygos vein is at the level of the 8th thoracic vertebra. A study done on 32 human fetuses demonstrated the vertebral level of communication of the azygos vein, and hemiazygos vein was the eighth thoracic vertebra (35.7%) and accessory hemiazygos vein was at the fourth thoracic vertebra (41.6%).⁴⁵ A rare anomaly of the azygos venous system consisted of preaortic interazygos vein that was seen as a vascular structure surrounding the left anterior portion of the thoracic aorta at the level of eighth thoracic vertebra on enhanced computed tomography examination. It was continuous on the right side with azygos vein and with hemiazygos vein on the left side. This rare anomaly can mimic other surgical mediastinal masses.⁴⁶

CONCLUSION

The wide knowledge of all the possible variations of the azygos venous system being reviewed and highlighted under different headings is of utmost importance to the radiologists performing

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venous cannulations, cardiac echography, and other vascular radiology procedures. It is also important to cardiothoracic surgeons to minimize the chances of iatrogenic hemorrhage.

CLINICAL **S**IGNIFICANCE

Normal anatomy as well as all the possible variations should be kept in mind during clinical diagnostic procedures like echocardiography, other radiological tests, and also during cardiothoracic surgery. This can lead to iatrogenic bleeding or radiological misdiagnosis. Venous catheterization can be misplaced in the anomalies of azygos vein involving the dilatation of its terminal parts and in turn leading to severe bleeding.⁴⁷ The anomalies of the azygos venous system can affect the surgery of esophageal atresia.48 The azygos system of veins acts as an important accessory venous pathway that provides collateral circulation between superior vena cava and inferior vena cava, particularly in increased venous flow, increased venous pressure, or interruption.⁴ During insertion of devices into the azygos vein, the tip of the central venous catheter should be placed in the superior vena cava between the azygos vein ending into the superior vena cava and atriocaval junction. Sometimes, the tip may get dislocated inside the azygos vein. Implantable devices in azygos vein can cause thrombosis at the tip and fistula formation due to friction between vein and airways.^{49,50} Transvenous pacemaker implantation can also introduce superior vena cava syndrome due to thrombosis of the tip.⁵¹

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