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Dilated Accessory Hemiazygos Vein Mimicking Aortic Dissection in Setting of Absent Left Brachiocephalic Vein

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Abstract Although uncommon, aortic dissection can be rapidly fatal with a 20% out-of-hospital mortality which increases at a rate of 1 to 3% every hour [1]. An expeditious diagnosis of acute aortic dissection is therefore imperative. One of the most advantageous diagnostic modalities utilized in the setting of acute aortic dissection is a transesophageal echocardiogram (TEE). In an acute setting, TEE is preferred because of its ability to provide a timely and definitive diagnosis [2]. Despite its diagnostic convenience, clinicians must be aware that there are several entities that may mimic aortic dissection. One of these entities includes a dilated accessory hemiazygos vein. In this report, we present a case of a 66-year-old woman who was found to have a possible Stanford type B dissection in the descending thoracic aorta on a TEE exam in the context of work up prior to an elective electrophysiology study and ablation for paroxysmal atrial fibrillation. CT angiography, however, revealed a dilated accessory hemiazygos vein, a rare congenital vascular anomaly that could potentially mimic aortic dissection leading to misdiagnosis and potentially unnecessary interventions.

Keywords: *aortic dissection, transesophageal echocardiogram, hemiazygos vein, brachiocephalic vein, congenital vascular anomaly*

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1. Introduction

Congenital thoracic anomalies have become more prevalent with the increasing use of noninvasive imaging modalities. The azygous venous system serves as an accessory collateral pathway between the inferior and superior vena cava (IVC/SVC) system [3]. The azygous system is formed from embryological remnants of the posterior cardinal veins [4]. The azygous vein drains venous blood from the posterior thorax and abdomen into the SVC. It is formed by the union of the ascending lumbar vein with the right subcostal veins. The accessory hemiazygos vein and the hemiazygos vein drain the superior left hemithorax (specifically the left-sided fourth through eighth intercostal veins) into the azygos vein on the right. A rare variation of the accessory hemiazygos vein involves an anatomic connection with the left brachiocephalic vein, which is seen in approximately 1-2% of the population [5]. The hemiazygos vein is the inferior and left-sided counterpart to the right-sided azygous system which is responsible for draining the ninth to the eleventh intercostal veins as well as the subcostal vein. In the absence of a left brachiocephalic vein, the left internal jugular and left subclavian veins drain via the left superior intercostal vein into the left-sided accessory hemiazygos system which results in a dilated accessory hemiazygos vein [6]. On routine chest radiography, this gives the appearance of a widened mediastinum which may be misdiagnosed as hilar lymphadenopathy, retroperitoneal neoplasm or an aortic dissection [7]. Here we present a congenital anomaly of the azygous system which mimicked an aortic dissection on TEE.

2. Case Report

A 66-year-old obese female with essential hypertension, paroxysmal atrial fibrillation and atrial tachycardia was referred for a transesophageal echocardiogram prior to an elective electrophysiology study and ablation. On physical exam, she was morbidly obese with a body mass index of 37.7. Her cardiac exam was unremarkable with normal S1

and S2 heart sounds and absence of murmurs. The respiratory exam was unremarkable. Blood pressure and peripheral pulses were equal bilaterally. The chest X-ray was unremarkable. Mediastinum was within normal limits. Laboratory studies were unremarkable with the exception of mild normocytic anemia (hemoglobin of 10.2 g/dl).

Transthoracic echocardiography revealed a normal sized left ventricle with an estimated ejection fraction of 60-65%. The left ventricular wall thickness and diastolic parameters were normal. The right ventricle was dilated but demonstrated normal systolic function. The aortic root, sinotubular junction and ascending aortic diameters were within normal limits. The aortic valve was trileaflet with no significant regurgitation. No other significant valvulopathy was noted.

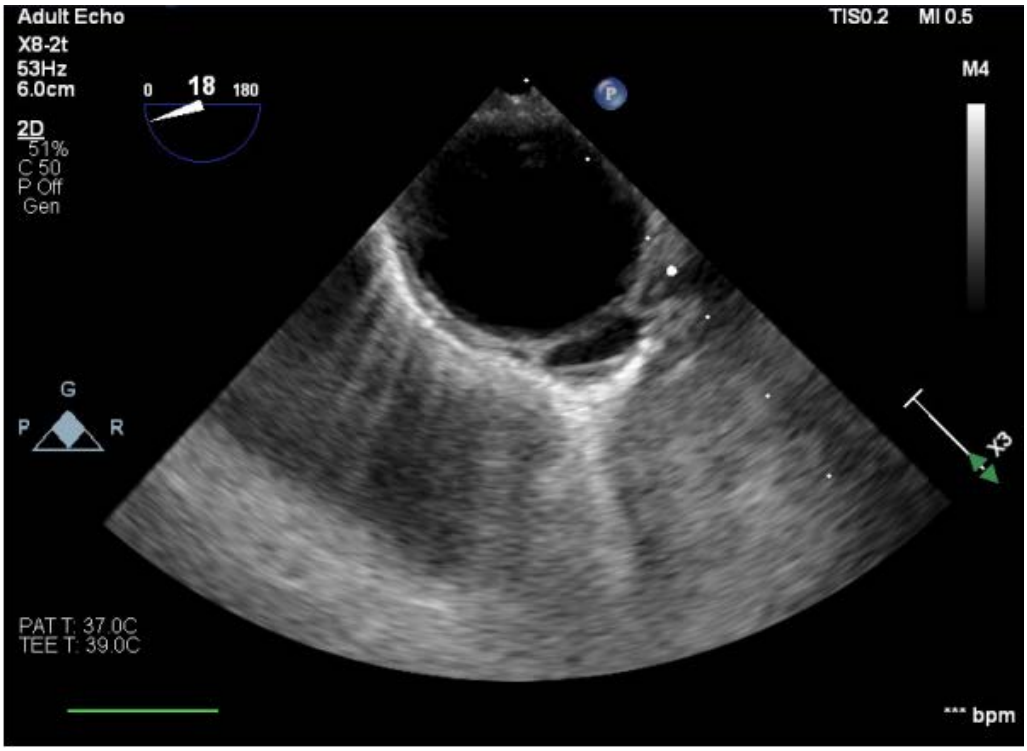

Figure 1. Descending thoracic aorta, with a posteriorly located echolucent lumen

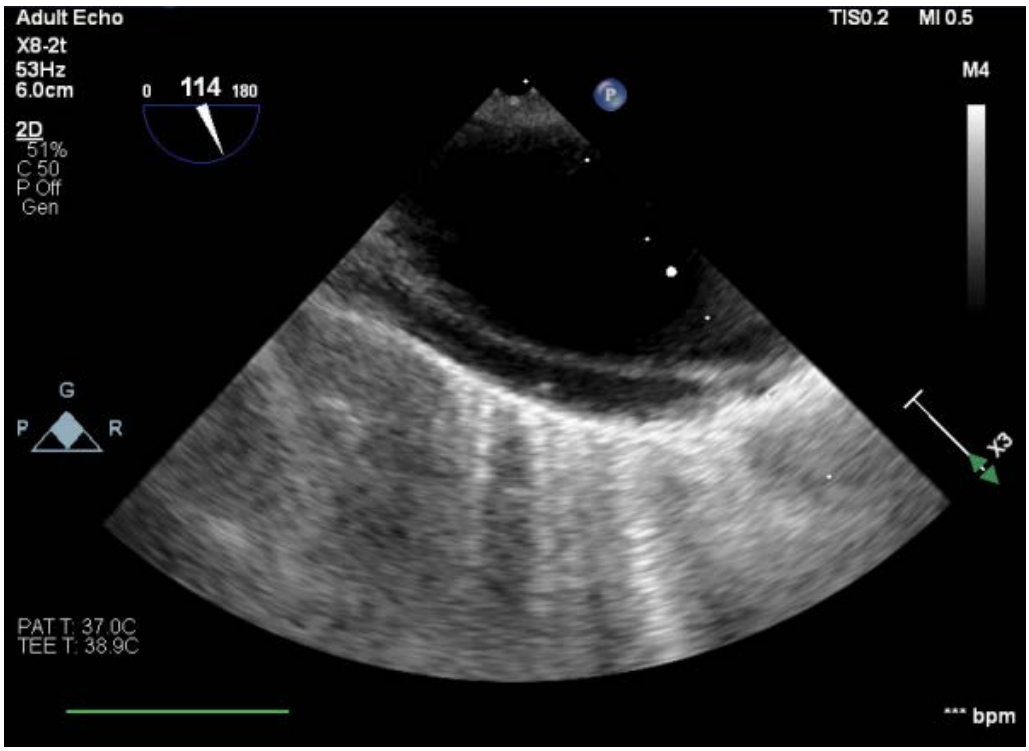

Figure 2. Long axis view demonstrating a suspected Stanford type B dissection Flap

The patient underwent a transesophageal echocardiogram in the EP lab under general anesthesia with endotracheal intubation due to high risk neck anatomy. The study confirmed normal left ventricular parameters and no significant valvulopathy. The left atrial appendage velocity was normal on pulse doppler imaging. There was no LAA thrombus. Evaluation of the descending thoracic aorta revealed an additional echolucent lumen located posteriorly (Figure 1, Figure 2). Color Doppler imaging demonstrated flow directed away from the probe (Figure 3). Pulse wave Doppler was suggestive of venous flow (Figure 4). A possible Stanford type B dissection in the descending thoracic aorta could not be excluded. No dissecting flap was noted in the ascending aorta. Additionally, a small sized atheroma was visualized in the proximal descending thoracic aorta. The patient underwent CT angiography for evaluation of suspected aortic syndrome.

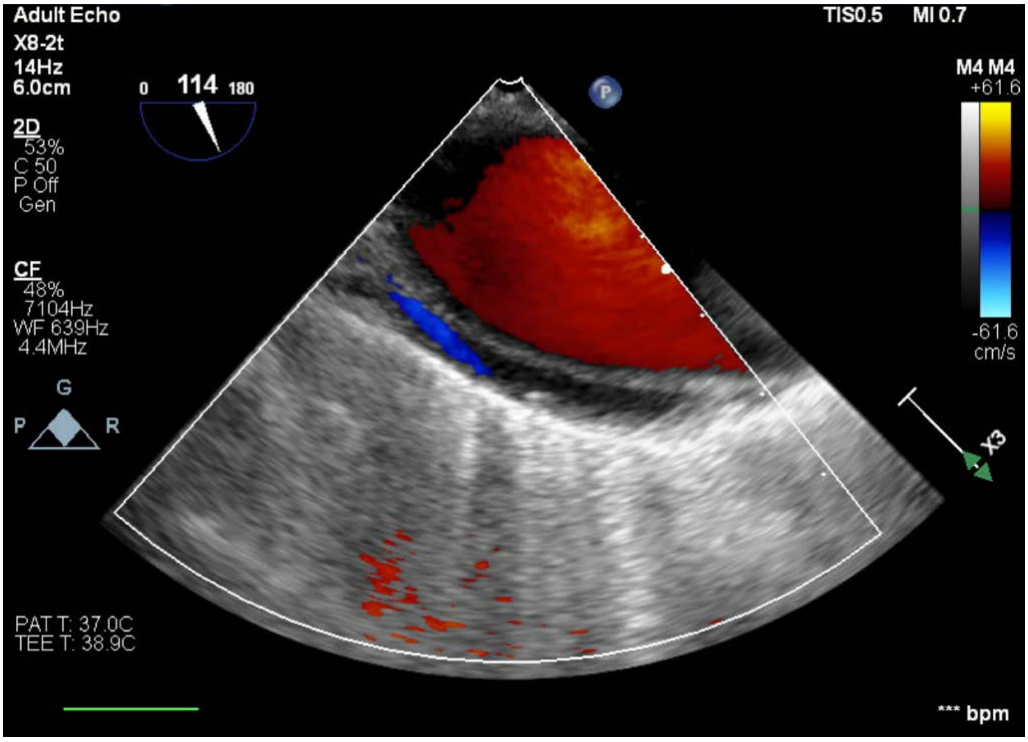

Figure 3. Color Doppler imaging demonstrating bidirectional flow

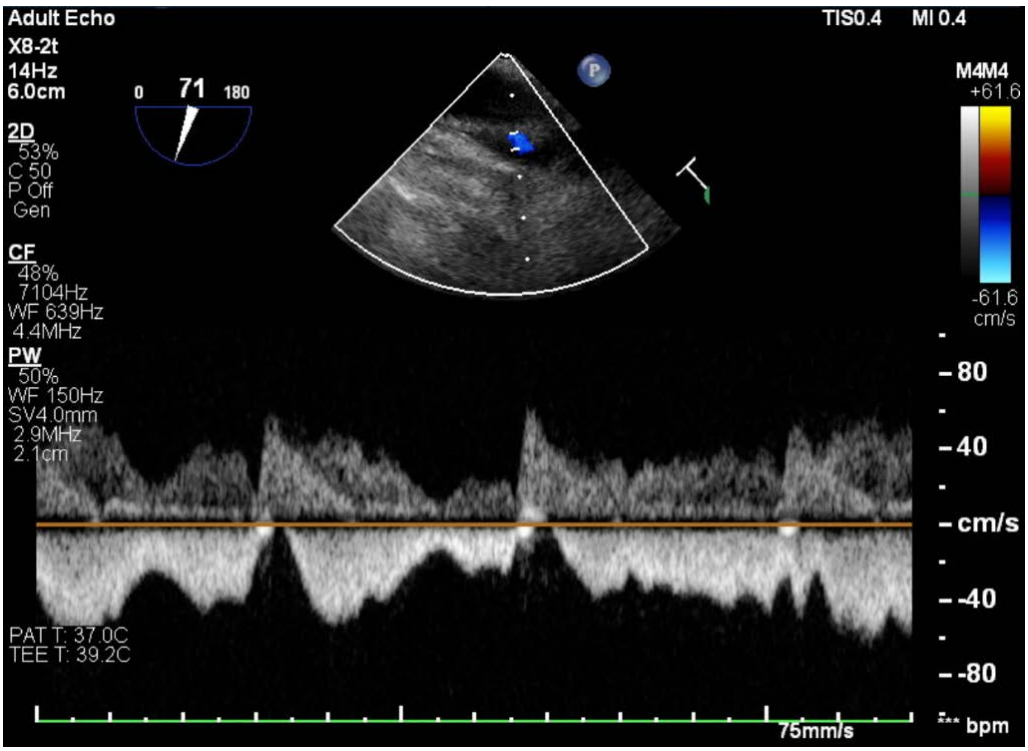

Figure 4. Pulse wave Doppler suggesting venous flow

CT angiography showed a normal descending aorta with no evidence of dissection, however it did reveal a dilated accessory hemiazygos vein (Figure 5, Figure 6, Figure 7). The left internal jugular and left subclavian were stenotic with a string-like appearance and there was an absent left brachiocephalic vein.

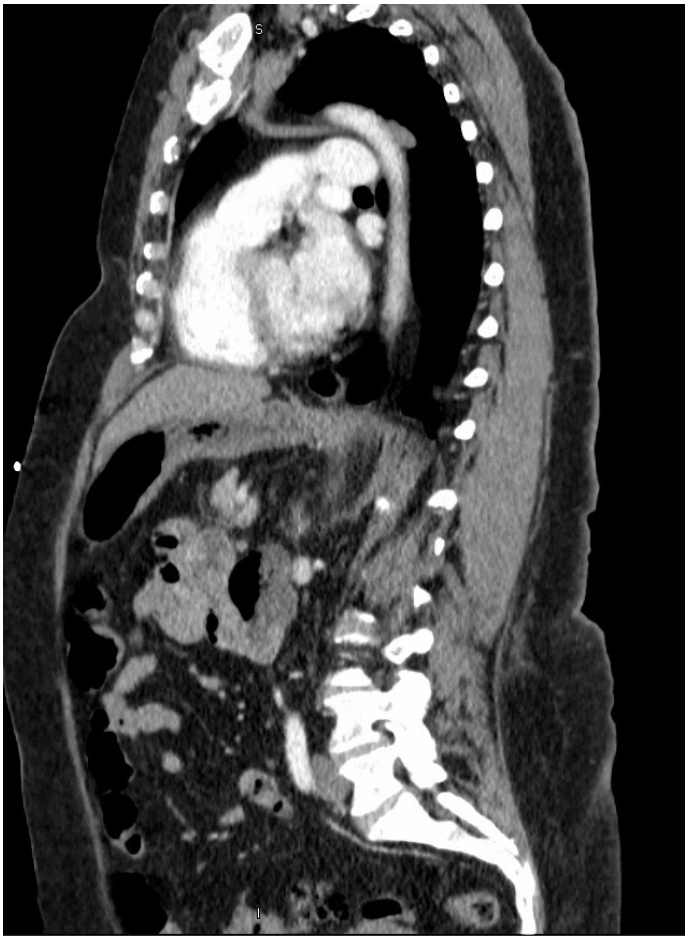

Figure 5. Sagittal view showing the accessory hemiazygos vein inferior to the descending thoracic aorta

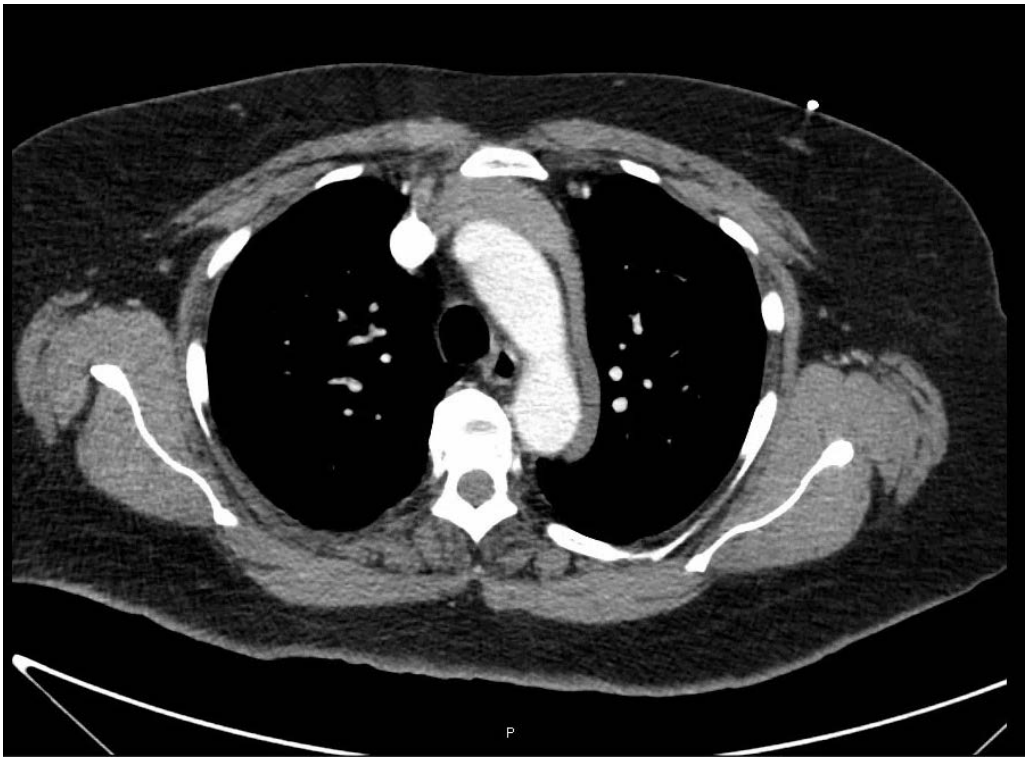

Figure 6. Transverse section showing dilated accessory hemiazygos vein

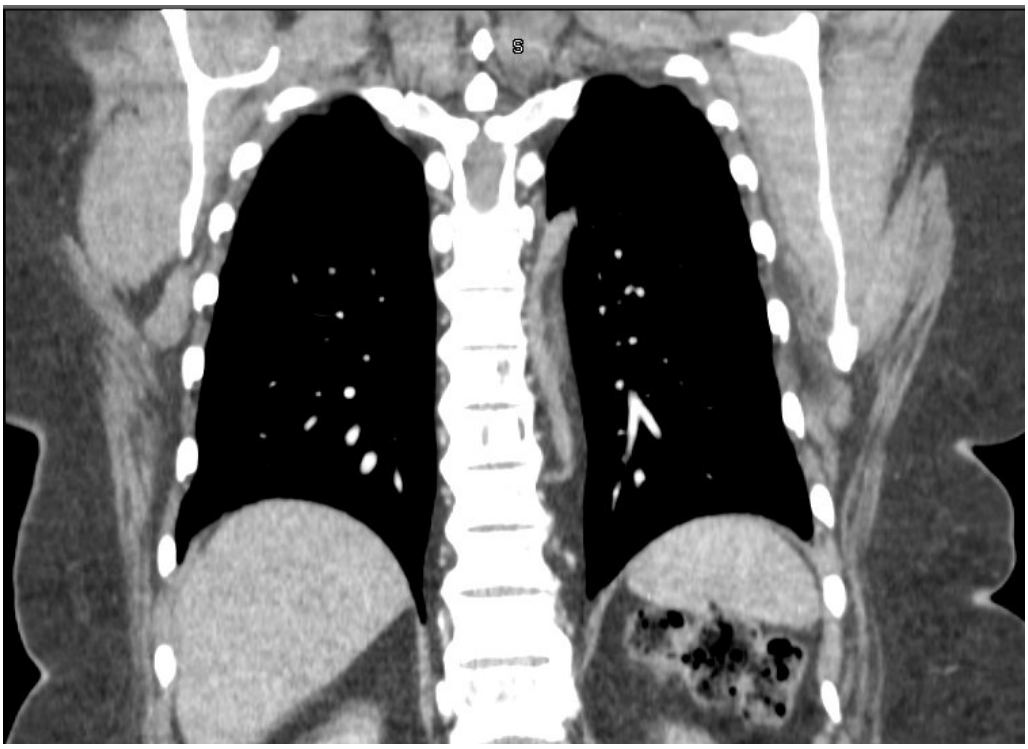

Figure 7. Coronal views showing dilated hemiazygos vein.

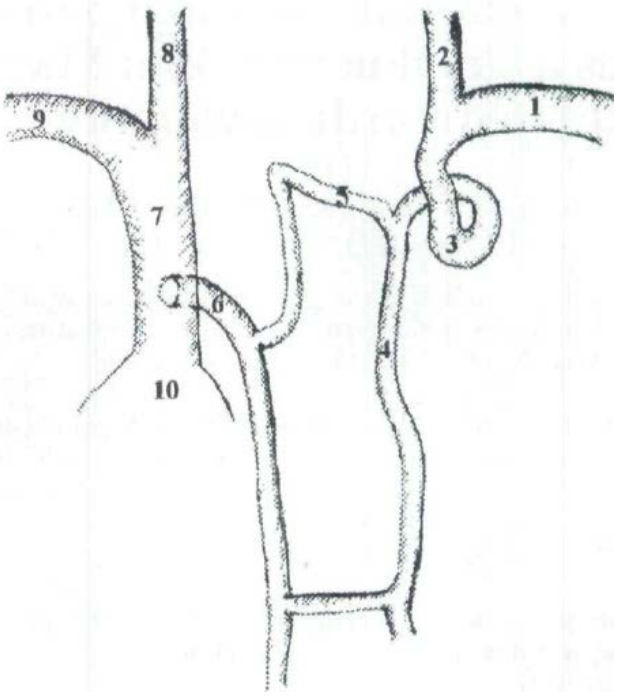

Figure 8. Anatomical representation of absent Left brachiocephalic vein. 1. Left subclavian vein 2. Left internal jugular vein 3. Left superior intercostal vein 4. Accessory Hemiazygos vein 5. Superior anastomosis 6. Azygos vein 7. Superior vena cava 8. Right internal jugular vein 9. Right subclavian vein

3. Discussion

An anomalous left brachiocephalic vein has an incidence of about 0.2 to 1% of all congenital cardiovascular anomalies [8]. There is limited data regarding the prevalence of left brachiocephalic vein anomalies with less than 100 reported cases [9]. Typically, the accessory hemiazygos vein courses vertically in the left retroperitoneal space between T5 to T8. The left fourth to seventh intercostal veins drain into the accessory hemiazygos system. The accessory hemiazygos vein either drains superiorly to the left superior intercostal vein (LSIV) and into the left brachiocephalic or merges with the right-sided azygos vein at the level of T7-T8 [10]. In our patient, CT angiography revealed the absence of the left brachiocephalic vein with severe stenosis of the left internal jugular and subclavian vein, which resulted in the dilation of the LSIV and subsequent dilation of the accessory hemiazygos vein (Figure 8).

TEE remains an efficient and timely diagnostic tool in acute aortic syndrome, however it is dependent on operator proficiency, with sensitivities ranging from 52 to 80% and specificities ranging from 87 to 99% [11]. To avoid diagnostic pitfalls, echocardiographers must be aware of various congenital anomalies of the azygous system. Endovascular procedures such as right heart catheterization, central venous cannulation via the left internal jugular vein and cardiac device implantation in the setting of such venous anomalies can result in higher failure rates and possible complications [12].

4. Conclusion

We presented a case of a 66-year-old woman with paroxysmal atrial fibrillation who was referred for a TEE prior to an elective electrophysiology study and ablation. TEE showed a possible Stanford type B dissection in the descending thoracic aorta. However, CT angiography showed a normal descending aorta with no evidence of dissection. It also revealed a dilated accessory hemiazygos vein, a rather rate congenital anomaly that might mimic aortic dissection on TTE. This is quite serious differential diagnosis for the echocardiographers must be aware of, in order to avoid misdiagnoses and unnecessary and potentially hazardous interventions.

Acknowledgements

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