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Incidental finding of situs ambiguous frustrates attempt at pulmonary vein isolation

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Abstract

We present a case of a 76-year-old woman undergoing elective pulmonary vein isolation. The procedure had to be terminated early due to an incidental finding of an interrupted inferior vena cava with azygos continuation. Computer tomography showed further visceral anomalies and revealed a case of situs ambiguous. Knowledge of anomalies of the vascular system and of the thoracic and abdominal organs is essential before planning trans-catheter procedures to prevent frustrated treatments and complications.

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Introduction

Catheter-based pulmonary vein isolation is an evidence-based treatment to achieve control of heart rhythm in patients with paroxysmal or persistent atrial fibrillation [1]. Several vascular or anatomic anomalies can lead to difficulties in access to the heart and further complicate specific treatments of cardiovascular diseases and cardiac arrhythmia.

Case report

A 76-year-old woman was admitted for an elective pulmonary vein isolation (PVI) due to persistent atrial fibrillation. During the procedure the diagnostic catheter performed an unusual loop where the right atrium was to be expected (Figure 1). An anomaly of the inferior vena cava was suspected which led to an early termination of the PVI. Computer tomography confirmed the inferior vena cava entering the superior vena cava directly and a diagnosis of azygos continuation due to an interrupted inferior vena cava was made (Figure 2). The hepatic veins drained directly into the right atrium. Additionally, asplenia, a centrally-located liver and an atypical localization of the stomach in the right upper abdomen was found (Figure 3). The patient denied any type of (exertional) dyspnea, reduced performance compared to peers or a higher frequency of bacterial infections. The past medical history was unremarkable, except that hilar tuberculosis was once suspected in a past, routine chest x-ray. The sputum results were always negative however, and treatment was not necessary. Of note, the daughter of the patient was born with three kidneys. No further anatomic anomalies in the family are known.

Discussion

Laterality defects include Situs Inversus (SI) and situs ambiguous (SA) [2]. Any laterality defect other than SI can be included in the Heterotaxy Syndromes (HS) [2]. In patients with HS or SA a misarrangement of the thoracoabdominal organs regarding the left-right axis can be found [3]. An association with cardiac malformations is common [3]. Laterality defects usually develop in early stages of the embryogenic period and several genetic markers such as ZIC3, LEFTYA, CRYPTIC and ACVR2B are related to causing heterotaxy syndromes [4-6]. Maternal diabetes and maternal cocaine-intake during the first trimester can also result in laterality defects [7]. HS includes left and right atrial isomer-

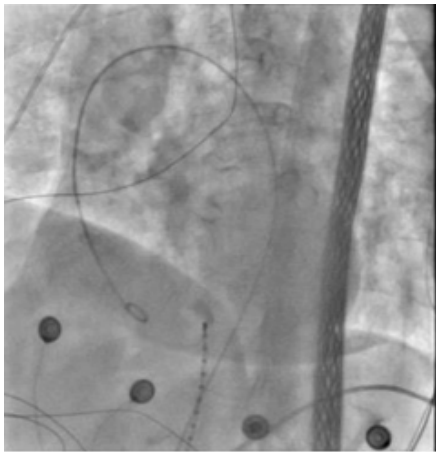

Figure 1: Unusual loop of the diagnostic catheter during the procedure.

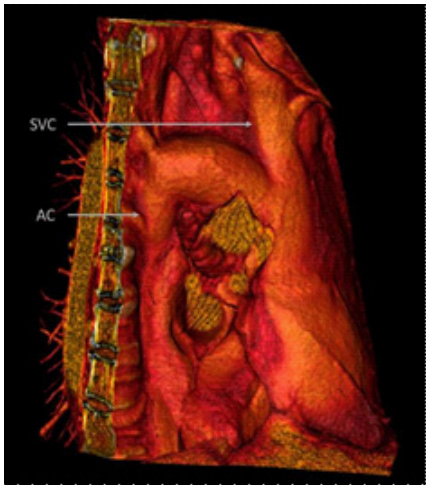

Figure 2: Three-dimensional reconstruction of the inferior vena cava anomaly.

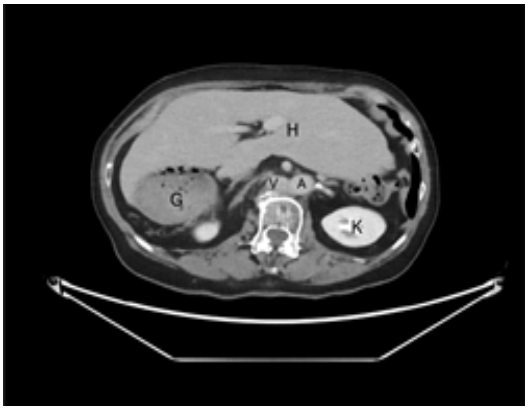

Figure 3: Computer tomography showing an absence of the spleen, a centrally-located liver **(H)** and a right sided stomach **(G)**. K (kidney), V (inferior vena cava), A (aorta abdominalis).

ism. In cases of left atrial isomerism mostly levocardia, left atrial morphology on both atrial sides, intermediate atrioventricular septal defects, polysplenia, an inferior vena cava interruption with azygos continuation and bilateral two-lobed lungs are typical [8-10]. In contrast, patients with right atrial isomerism usually show dextrocardia or mesocardia, right atrial morphology on both atrial sides, common atrioventicular valve/atrium or complete atrioventricular septal defects, bilateral three-lobed lungs, pulmonary stenosis or atresia and asplenia [8,10]. In both cases, a centrally located liver and a right, left or centrally sided stomach are possible [8]. In cases of right atrial isomerism, cardiac malformations are mostly more severe than in cases of left atrial isomerism [11]. An interrupted inferior vena cava with azygous continuation refers to a failed anastomose of the right subcardinal vein with the vitelline vein [12]. Dilatation of the azygos vein can lead to misdiagnoses as described in the present case [12]. Possible alternative approachs for PVI could be the left subclavian vein, a combined approach of subclavian vein and internal jugular vein or a transhepatic access [13-15]. Furthermore, a minimal invasive surgical ablation of the atrial fibrillation could be considerated [16].

Conclusion

The presented case is the first case of an asymptomatic elderly patient with an incidental finding of an interrupted vena cava inferior with azygos continuation, asplenia and the absence of pulmonary or cardiac malformations. Consequently, the anatomic anomalies in our case are heterogenic and cannot be specifically ordered to left or right atrial isomerism and, according to the literature should not be seen as a rule [17]. In heterotaxy syndromes it is important to individually assess each organ [18]. Pre-procedural performance of a computer tomography can lead to significant extra-cardiac findings [19]. All treating physicians need to be aware about possible variations in anatomy, malformations or misarrangements of thoracoabdominal organs and the accompanying vascular system.

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