Mediastinal Distortions: A Rare Case of Congenital Anatomic Relocations of Mediastinal Structures and Vascular Anomalies in Combination with Heterotopic Conduction in Pulmonary Veins and Sick Sinus Syndrome

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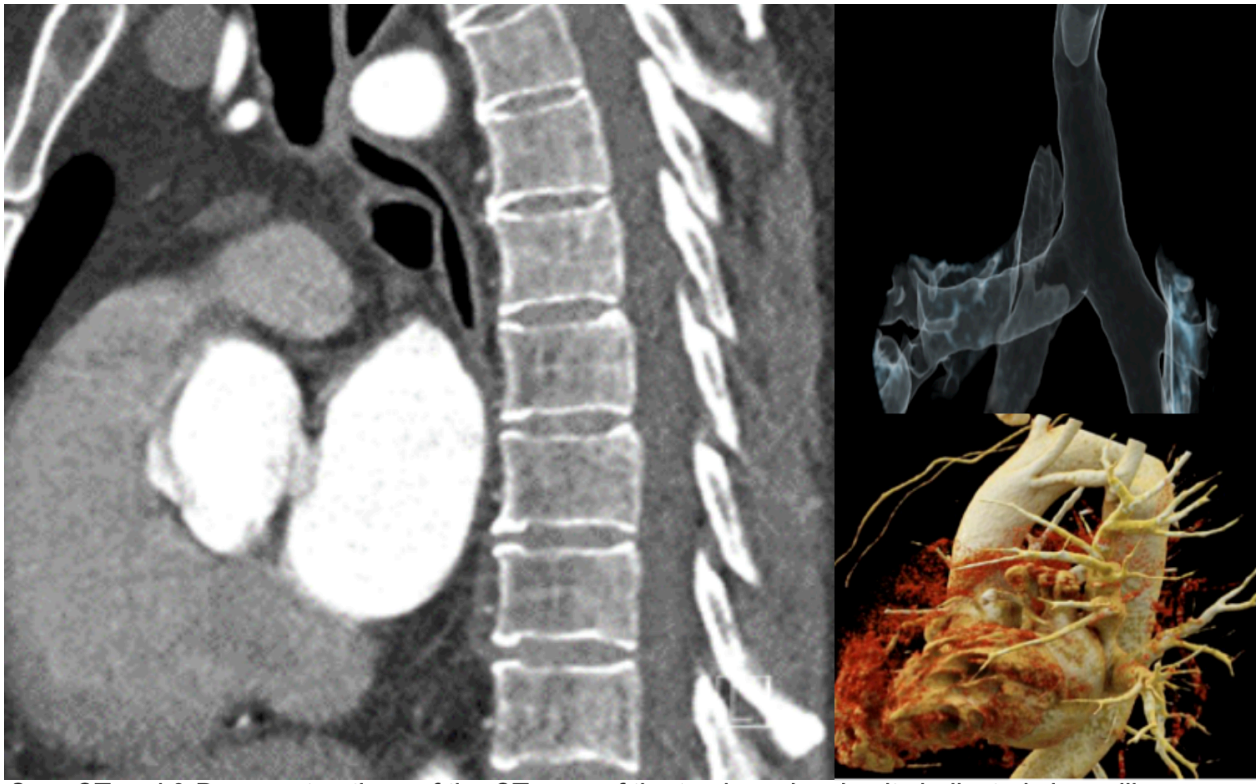
Abstract

The case we present involves an atypically developed thoracic aorta, resulting in the consecutive displacement of the trachea and esophagus, combined with the unusual formation of vascular bridges between the ascending and descending thoracic aorta and the arising supraaortic branches. This represents a pathological entity further associated with a cardiac condition of Sick Sinus Syndrome and heterotopy of the excitation center in the pulmonary veins. Multiple descriptions of the combination of dextrocardia and Sick Sinus Syndrome have been reported by Aurora Bakalli et al. 2021 (1), as well as the combination of dextrocardia, persistent left superior vena cava, and Sick Sinus Syndrome by Gangliang Guo et al. 2017 (2), and another case of dextrocardia and Sick Sinus Syndrome by Junqian Luo et al. 2022 (3). Such combinations of anomalies have not been reported in larger statistical collections to date.



Sag.-CT and 3-D reconstructions of the CT scan of the trachea showing its indicated clamp-like narrowing caused by the course of the aorta.

Keywords: *Malrotated thoracic aorta, bridging artery connecting ascending and descending aorta, embracing semicircular relocated and slighty compressed trachea and esophagus, heterotopic conduction in pulmonary veins, Sick Sinus Syndrome*

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Introduction

Dystopic mediastinal vascular structures, such as the right-descending thoracic aorta or the right subclavian artery running dorsally to the esophagus, also known as Arteria lusoria, which induces dysphagia due to esophageal compression, are the most frequently described aberrant mediastinal vessels, occurring in 0.5-2.5% of cases (4,5).

The right-descending thoracic aorta was diagnosed with a frequency of 0.012-0.018% in 2018 by Toshihiko Hayashi et al. in a study of 27 cases from three Japanese university hospitals using angio-CT (6).

The aorta displaced from its normal position, with the ascending thoracic aorta and aortic arch shifted to the midline and the descending thoracic aorta descending on the right, results in the esophagus being displaced, as well as slight compression and lateral displacement of the trachea to the left, and further distally to the right in its course. Additionally, it is semi-circularly encompassed on the left side by a previously unnamed artery that bridges the ascending and descending

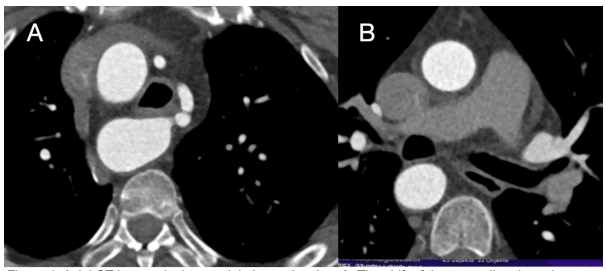

Figure 1: Axial CT images in the arterial phase showing: A: The shift of the ascending thoracic aorta and aortic arch to the midline and the trachea to the left. B: The right-descending thoracic aorta

aorta. Two supraaortic branches arise from this atypical vessel.

This anatomy is extremely unusual and has not yet been described in combination with a positional anomaly of the thoracic aorta. Furthermore, the patient presents a combination of Sick Sinus Syndrome with a pulmonary excitation center dystopia.

The occurrence of clinically relevant aortic arch pathologies has been detailed by Minhee Hwang et al. in 2023. Out of 348 patients examined using angio-CT and/or MRI, 29 patients (8.3%) had aortic arch anomalies, of which 13 patients were clinically symptomatic (13/29), corresponding to an occurrence rate of 44.8% (9). For further information on existing congenital, partially complex aortic arch anomalies, refer to the work of Shenli Li et al. 2019 in Cardiovasc Pathol. In this study, anomalies were classified into sub-groups I-V according to the Stewart classification of 1964 (10).

Case Presentation

Emergency inpatient admission of a 55-year-old patient who presented to our emergency department in March 2024 with recurrent highly symptomatic atrial fibrillation. This was preceded by a cryoablation of a heterotopic conduction system in the pulmonary veins in November 2022 at an external hospital. The patient is known to have Sick Sinus Syndrome with a pathological sinus node recovery time, for which rhythm-controlling therapy or beta-blocker therapy was no longer feasible from a cardiological perspective at our clinic.

Further imaging diagnostics:

As part of further cardiological evaluation using imaging diagnostics, cardiac CT imaging was performed using MDCT: The MDCT of the heart

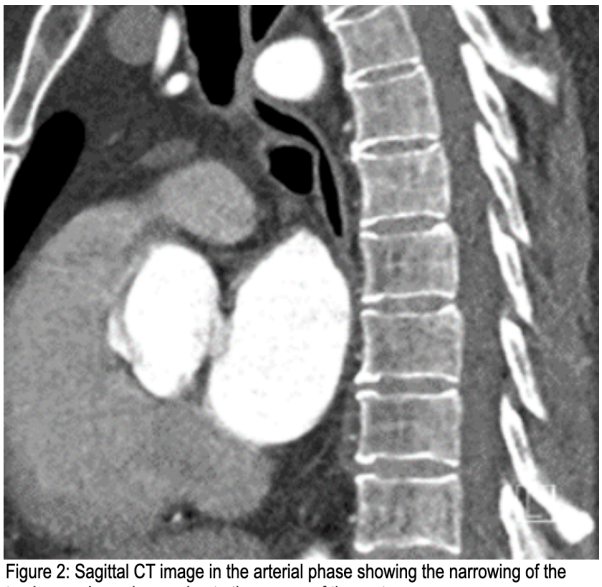

Figure 2: Sagittal CT image in the arterial phase showing the narrowing of the trachea and esophagus due to the course of the aorta.

showed calcifying plaques in the left main coronary artery and the LAD. Further analysis and reconstructions of the mediastinum revealed a midline shift of the ascending thoracic aorta and aortic arch, with a right-descending thoracic aorta in its further course (Figure 1). This aortic course results in an indicated clamp-like narrowing of the trachea, with a rightward elevation and leftward displacement (Figure 1 & 3), causing the craniocaudal course of the trachea to resemble the tortuous path of the thoracic aorta. Additional slight narrowing of the trachea is observed due to an atypical vessel encircling it semi-circularly on the left side, bridging the ascending and descending aorta. Three supraaortic branches arise from this vessel, as seen in the 3-D

reconstructions (Figure 4). These vessels are likely the left subclavian artery and the brachiocephalic trunk. No other misplacements or atypical findings are present, and the pulmonary veins

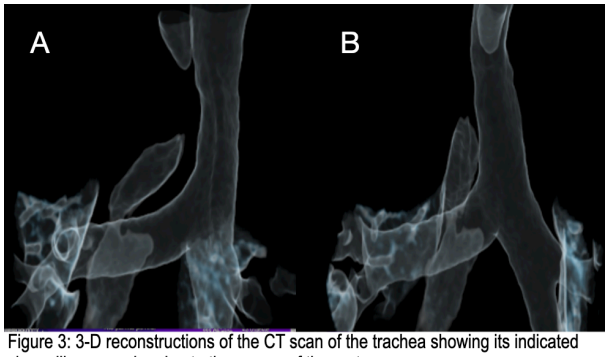

Figure 3: 3-D reconstructions of the CT scan of the trachea showing its indicated clamp-like narrowing due to the course of the aorta.

drain correctly into the left atrium as seen on the MDCT of the heart.

The esophagus, similar to the trachea, appears slightly tortuous and displaced due to the atypical course of the thoracic aorta. In the proximal third, there is a stenosis caused by the vascular bridge, resulting in the clinical symptom of mild dysphagia (Figure 2).

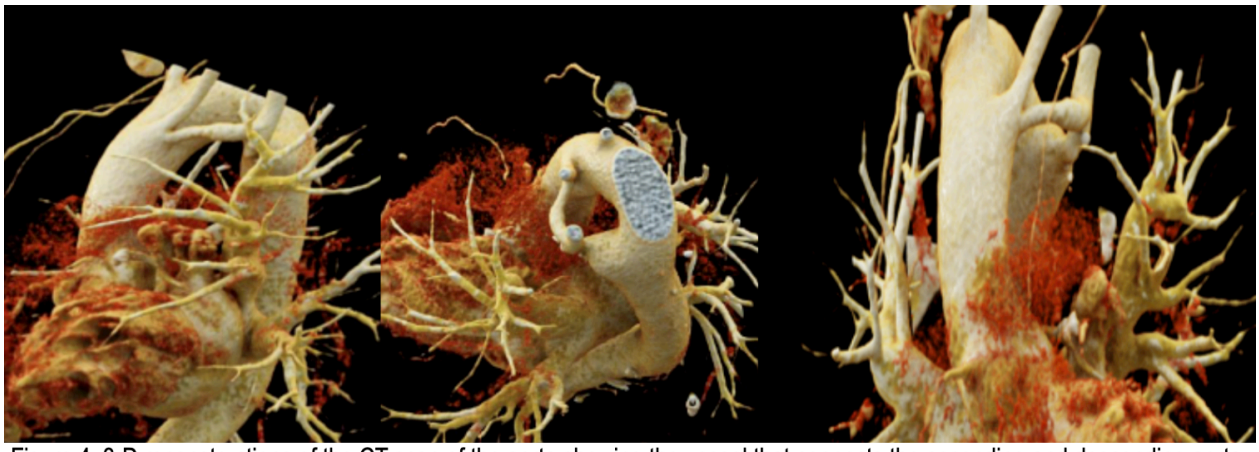

Figure 4: 3-D reconstructions of the CT scan of the aorta showing the vessel that connects the ascending and descending aorta, as well as the various supraaortic branches.

Discussion

In our case, the thoracic aorta with its atypical course, resulting in the consecutive displacement of the trachea and esophagus, combined with the unusual formation of vascular bridges between the ascending and descending thoracic aorta and the arising supraaortic branches, represents a pathological entity further associated with a cardiac condition of Sick Sinus Syndrome and heterotopy of the excitation center in the pulmonary veins. Multiple descriptions of the combination of dextrocardia and Sick Sinus Syndrome have been reported by Aurora Bakalli et al. 2021 (1), as well as the combination of dextrocardia, persistent left superior vena cava, and Sick Sinus Syndrome by Gangliang Guo et al. 2017 (2), and another case of dextrocardia and Sick Sinus Syndrome by Junqian Luo et al. 2022 (3). Such combinations of anomalies have not been reported in larger statistical collections to date. Only individual case reports exist.

In our case of a 55-year-old patient, we observe an unusual combination of several anomalies and dystopias:

1. A vascular anomaly involving the formation of a vascular bridge between the dystopic ascending and descending thoracic aorta, with supraaortic branches arising from it.
2. A midline position of the ascending aorta and aortic arch with a right-descending thoracic aorta.
3. A slight displacement of the trachea and a localized clamp-like vascular-induced narrowing of its lumen due to the unusual vascular bridge between the ascending and descending aorta.
4. A displacement of the esophagus and its vascular-induced luminal narrowing by the described vascular bridge between the ascending and descending thoracic aorta, caused by the atypical course of the thoracic aorta.
5. A heterotopic excitation center in the pulmonary venous system, with normally draining pulmonary veins into the left atrium.
6. The presence of Sick Sinus Syndrome.

To our knowledge and based on our PubMed search, this rare combination of anatomical vascular variations, dystopia of the aortic arch and its branches, a vascular bridge between the ascending and descending thoracic aorta causing tracheal and esophageal displacement and

dysphagia, has not been previously described (Tab.1). Such cases are usually described with the presence of a Lusoria artery (4,5).

Unusual combination of several anomalies and dystopias
1. Vascular bridge between the dystopic ascending and descending thoracic aorta, with supraaortic branches arising
2. Midline position of the ascending aorta and aortic arch with a right-descending thoracic aorta
3. Displacement of the trachea and a localized clamp-like vascular-induced narrowing of its lumen
4. Displacement of the esophagus and its vascular-induced luminal narrowing leading to dysphagia
5. Heterotopic excitation center in the pulmonary venous system, with normally draining pulmonary veins into the left atrium
6. Sick Sinus Syndrome

Tab.1: Rare case of congenital anatomic relocations of mediastinal structures and vascular anomalies in combination with heterotopic conduction in pulmonary veins and Sick Sinus Syndrome.

According to Pschyrembel's depiction of various vascular courses, vascular anomalies in the aortic arch are described with a frequency of 1% (7). The treatment of the heterotopic excitation center in the pulmonary venous system had already been performed once at an external hospital by cryoablation and, according to our cardiologists, could not be repeated.

To gain some understanding of the occurrence and frequency of a secondary excitation center in the pulmonary veins causing atrial fibrillation, refer to the work of Takahashi Nakashima et al. 2023 (8). He reports on 113 patients with a total of 441 pulmonary vein isolations (PVIs) with existing atrial fibrillation. Clinically, this pathological electrophysiological combination of Sick Sinus Syndrome with a heterotopy of a secondary excitation center in the pulmonary veins was the reason for the emergency inpatient admission of the patient with existing atrial fibrillation.

The isolated occurrence of Sick Sinus Syndrome is described by Paul N. Jensen et al. 2014 in a compilation of 20,572 patients with a frequency of 0.8% per 1000 persons examined per year in his 17-year follow-up study. A lower incidence was found among colored people compared to caucasians. Right bundle branch block, hypertension, and obesity were the most commonly described comorbidities among his patients. A combination or parts of the anomalies described in our case were not found in the very large cohort of this study (7), highlighting the uniqueness of our case.

Further treatment strategy:
The patient's clinical course during hospitalization was satisfactory after successful cardiological stabilization through medication, allowing for a timely discharge. Close outpatient monitoring, initially by the primary care physician, has been arranged with the patient due to this extremely unusual combination of anatomical and electrophysiological cardiac conditions.

Conclusion

To our knowledge and based on a comprehensive PubMed search, this case showcases an exceptionally rare and previously undescribed combination of anatomical vascular variations. It involves the dystopia of the aortic arch and its branches, an unusual vascular bridge between the ascending and descending thoracic aorta, and the consequent displacement of the trachea and esophagus.

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Conflict of interest:

The authors declare that there were no conflicts of interest within the meaning of the recommendations of the International Committee of Medical Journal Editors when the article was written. Further statements and declaration:

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