The unusual connection between the right-sided aortic arch and bronchial asthma

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Abstract

This work describes a rare clinical case of anomalous positioning of the aortic arch in a female patient with asthma. It highlights literature suggesting that this anomaly may sometimes mimic bronchial asthma, underscoring the importance of conducting bronchodilator reversibility tests more frequently to identify underlying asthma. The patient, a woman presenting with asthma symptoms, was found to have a Right-Sided Aortic Arch (RSAA), which is an unusual finding in the absence of congenital heart disease. Given the rarity of this presentation, it underscores the need for thorough evaluation and consideration of alternative diagnoses in patients with atypical asthma symptoms. This case emphasizes the importance of comprehensive diagnostic approaches, including spirometry and bronchodilator testing, to accurately diagnose and manage patients with asthma-like symptoms associated with rare anatomical variations such as RSAA.

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Introduction

According to the Global Initiative for Asthma (GINA) 2023 document, bronchial asthma is a heterogeneous disease characterized mainly by chronic airway inflammation. It is defined by the history of respiratory symptoms such as wheezing, dyspnea, chest tightness, and cough, which vary in intensity over time, together with the variable limitation of airflow on exhalation, which may become persistent over time. It is usually associated with Airway Hyper-Responsiveness (AHR) and inflammation, but these features are neither necessary nor sufficient for the diagnosis. The diagnosis of asthma is based on the history of characteristic symptoms and the presence of variable airflow limitation during exhalation, which must be confirmed by bronchodilator reversibility testing or other tests.[1] Variable airflow obstruction and AHR are hallmarks of asthma, with proximal airway wall thickening correlating with AHR in asthma. These findings underscore asthma's chronic obstructive nature and highlight the potential role of airway remodeling in promoting AHR.[2] Anomalies like Right-Sided Arcus Aorta (RSAA), with an incidence of 0.1-0.2%, may mimic bronchial asthma, especially during exercise. RSAA, often asymptomatic, can cause dyspnea and dysphagia. Seven patients with RSAA were retrospectively evaluated; four had symptomatic tracheal/esophageal compression. Misdiagnosis occurred in two patients who received asthma treatment, despite unresponsive symptoms. Diagnosis relied on chest radiography and was confirmed by thorax Computed Tomography (CT) / Magnetic Resonance Imaging (MRI).[3] A Caucasian woman with bronchial asthma presented a rare case of concurrent RSAA anomaly. This anomaly was detected during diagnostic evaluation for respiratory symptoms.

Case Report

A 63-year-old Caucasian woman came to my outpatient clinic in April 2024, with a history of bronchial asthma and sporadic allergic rhinoconjunctivitis. A chest X-ray (Figure 1) revealed an incidental finding of a RSAA. She presented with typical asthma

symptoms but reported non-adherence to prescribed therapy, including Inhaled Corticosteroids (ICS) / Long-Acting Beta Agonists (LABA). Spirometry showed reversible airflow obstruction. Blood analysis revealed hypercholesterolemia and vitamin D deficiency. An endocrinology visit diagnosed multinodular goiter with a cold nodule and hyperglycemia. Physical examination showed diminished tactile fremitus, decreased vesicular murmur, bilateral wheezes, and minimal crackles. Proposed management included pharmacotherapy adjustments for asthma (ICS/LABA twice/day) and hypercholesterolemia (switch therapy with rosuvastatin), along with recommendations for further cardiac evaluation. During the pulmonology visit, the patient exhibited normal chest morphology with decreased tactile fremitus, clear pulmonary resonance, and diminished vesicular murmur. Spirometry (Table 1) confirmed reversible airflow obstruction, consistent with non-adherent allergic bronchial asthma (Figure 2). Pharmacotherapy adjustments were recommended, including medication changes for hypercholesterolemia and initiation of asthma maintenance therapy. Follow-up spirometry after one month of treatment and a cardiology consultation for echocardiography and vascular assessment were advised. These assessments aimed to optimize asthma management, address comorbidities and ensure cardiovascular health in this complex clinical presentation.

Discussion

Anomalies in the anterior mediastinum often present diagnostic challenges, as they can mimic symptoms of common respiratory conditions such as bronchial asthma. This is notably observed in the spirometric curves, with aspects of variable intrathoracic airway obstruction. Several case reports in the literature have highlighted this phenomenon, illustrating how anterior mediastinal anomalies can induce a characteristic spirometric profile that closely mimics bronchial asthma.[4] In cases where right aortic arch transposition symptoms develop in infancy, surgical intervention provides a straightforward solution to alleviate symptoms effectively. This approach ensures symptom resolution, offering relief from dysphagia, chronic cough, and other associated respiratory

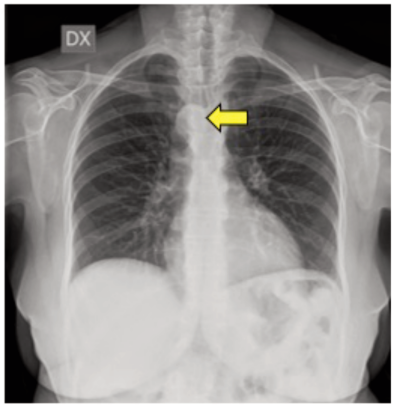

Figure 1. In the figure, it is possible to observe the presence of right-sided aortic arch positioning, indicated by the yellow arrow, associated with accentuation of the bronchovascular pattern. Therefore, it seemed necessary to advise the patient to undergo a Cardiology evaluation to exclude congenital vascular anomalies.

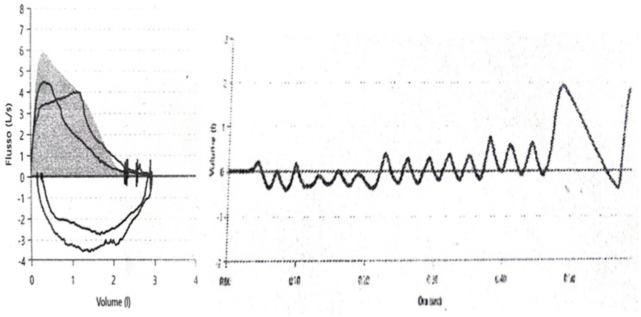

Figure 2. In the second test, there is a variable intrathoracic obstruction, that indicates the concomitance of both pathologies: asthma and right-sided aortic arch.

Table 1. Global Spirometry with PulmOne MiniBox+. Global Spirometric values indicating the presence of bronchial asthma and air trapping in expiratory flow (RV and RV/TLC upper to 100% predicted value). As demonstrated, there is a mild obstructive deficit reversible after the administration of 400 mcg of salbutamol, with a percentage change in Forced Expiratory Volume in 1 second (FEV1) of 15% of the predicted value, corresponding to 290 mL in absolute value. Additionally, there is a change in the Tiffeneau index percentage (FEV1/FVC pre: 63.89-80% of predicted value *vs* 74.06-93% of predicted value post-bronchodilator).

Spirometric parameters calculated	Predicted Value (pre-BD)	Predicted Value (post-BD)	Percentage change from predicted value
FEV1	1,89 L - 87%	2,18 L - 100%	+15%
FVC	2,84 L - 103%	2,94 L - 107%	+4%
FEV1/FVC	63,89% - 80% predicted value	74,06% - 93% of predicted value	+16%
FEF 25-75	0,86 L/sec – 43%	1,69 L/sec – 85%	+97%
PEF	4,46 L/sec – 81%	4,00 L/sec – 72%	-11%
TLC	5,32 L – 117%	/	/
RV	2,48 L – 153%	/	/
RV/TLC	46,68% - 133%	/	/
IC	1,99 L – 91%	/	/

FEV1, Forced Expiratory Volume in 1 second; FVC, Forced Vital Capacity; FEV1/FVC, Forced Expiratory Volume in 1 second to Forced Vital Capacity ratio; FEF 25-75, Forced Expiratory Flow 25-75%; PEF, Peak Expiratory Flow; TLC, Total Lung Capacity; RV, Residual Volume; RV/TLC, Residual Volume to Total Lung Capacity ratio; IC, Inspiratory Capacity; BD, Bronchodilator.

issues.[5] The incidence and prevalence of RSAA anomalies, such as Double Aortic Arch (DAA) and Right Aortic Arch with Left-Sided Ductus Arteriosus (RAALSA), are relatively low in the general population, typically accounting for less than 1% of congenital heart defects. These anomalies are often asymptomatic, making diagnosis challenging and frequently discovered incidentally during imaging studies. However, when present, they are commonly associated with other congenital cardiac anomalies, which can complicate diagnosis and clinical management.[6] Aortic arch anomalies arise from deviations in embryonic aorta development. Normally, the aortic arch forms from the truncus arteriosus and divides into left and right dorsal vessels, which later merge into a single descending structure. The left 4th aortic arch becomes part of the distal arch, while the right 4th aortic arch forms the proximal segment of the right subclavian artery. Persistence of the right dorsal aorta can lead to anomalies like double aortic arches or vascular rings. A persistent RSAA, occurring in 0.1% of adults, is often associated with a complete vascular ring, causing respiratory compromission.[7] Symptomatic cases of RSAA in the anterior mediastinum have been reported in the medical literature, with symptoms such as exertional dyspnea and chronic cough that persist despite optimal asthma treatment.[8,9] The intensity and nature of the functional symptoms generated by this condition can simulate other pathologies. The difficulty in diagnosis arises because confirmation remains elusive, although the functional symptoms suggest it. Imaging is crucial in diagnosing aortic arch anomalies, specifying the type of anomaly, its relationship with respiratory and esophageal structures, and associated abnormalities. Diagnostic methods such as echocardiography, angiography, and multiplanar reconstruction CT scans are valuable for accurate diagnosis, aiding in surgical decision-making.[10] RSAA and bronchial asthma are distinct conditions, but one can mimic the other. In this clinical case, both conditions coexist without the aortic arch simulating asthma.

Conclusions

Anomalies like the RSAA in the anterior mediastinum can mimic symptoms of bronchial asthma, posing diagnostic challenges. Spirometric patterns often resemble asthma, complicating diagnosis. Surgical intervention is needed for symptomatic cases to alleviate symptoms effectively. Despite their rarity, these anomalies significantly affect patient health and quality of life. Accurate diagnosis involves thorough evaluation, including echocardiography and CT scans. Awareness of these anomalies and their clinical presentations is crucial for timely diagnosis and proper management, ensuring optimal patient outcomes and avoiding unnecessary treatments for asthma-like symptoms associated with uncommon anatomical variations. In this clinical case, contrary to previous literature, the association between asthma and RSAA positioning in the mediastinum is evident. This emphasizes the importance of investigating such cases with spirometry and bronchodilator reversibility testing.

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