Double Aortic Arch Mimics the Clinical Characteristics of Severe Reactive Airway Disease in a Pediatric Patient

Osman Yesilbas[1] Hazar Dogus Kus[2] Güntülü Şik[3] Agop Citak[3] Bahar Temur[4] Can Yilmaz Yozgat[5] Hafize Otcu Temur[6] Erkan Cakir[7] Yilmaz Yozgat[8]

[1] Department of Pediatric Critical Care Medicine, Bezmialem Vakif University, Istanbul, Turkey
[2] Department of Pediatrics, Bezmialem Vakif University, Istanbul, Turkey
[3] Department of Pediatric Critical Care Medicine, Mehmet Ali Aydınlar University, Faculty of Medicine, Atakent Acıbadem Hospital, Istanbul, Turkey
[4] Department of Cardiovascular Surgery, Mehmet Ali Aydınlar University, Faculty of Medicine, Atakent Acıbadem Hospital, Istanbul, Turkey
[5] Faculty of Medicine, Bezmialem Vakif University, Istanbul, Turkey
[6] Department of Radiology, Bezmialem Vakif University, Istanbul, Turkey
[7] Department of Pediatric Pulmonology, Bezmialem Vakif University, Istanbul, Turkey
[8] Department of Pediatric Cardiology, Bezmialem Vakif University, Istanbul, Turkey.

Address for correspondence Yilmaz Yozgat, MBBS, MD, Department of Pediatric Cardiology, Adnan Menderes Bulvarı, Vatan Caddesi, Bezmialem Vakif University, Istanbul 34093, Turkey (e-mail: yozgatyilmaz@yahoo.com).

J Pediatr Intensive Care 2020;9:141–144.

Abstract

Reactive airway disease is a prevalent condition that can be detected in the early infancy period. The condition might also deteriorate into asthma in some cases. If infants do not respond to the treatment of persistent wheeze and coughing, other rare causes should be investigated. The complete form of vascular ring is an extremely uncommon congenital cardiovascular abnormality. Double aortic arch constitutes the most significant portion of the complete vascular ring anomalies. Clinical manifestations of the anomaly are mainly respiratory due to the tracheal compression and mimicking the conditions of asthma. There have not been many reports about the clinical presentations of double aortic arch being remarkably similar to the same clinical manifestations of asthma in the literature. As far as we can be sure, there have not been any reported cases about severe reactive airway disease that caused a patient to have a life-threatening condition in the pediatric intensive care unit. Herein, we present a 5-month-old girl who had double aortic arch. Her anatomical aberration was diagnosed by three-dimensional computed tomography angiography of thorax, and the anomaly mimicked the clinical characteristics of life-threatening severe reactive airway disease.

Keywords
- double aortic arch
- reactive airway disease
- respiratory insufficiency
- childhood

Introduction

A complete vascular ring is a relatively rare congenital cardiovascular malformation representing ~0.8 to 2% of all congenital cardiovascular malformations. The branches of the anomaly encircle the esophagus and apply immense pressure onto upper airway tracks.[1–3] The vascular ring has two aortic arches, both may be patent, or one of them

received
October 13, 2019
accepted after revision
November 23, 2019
published online
January 8, 2020

DOI https://doi.org/10.1055/s-0039-1700952.
ISSN 2146-4618.

may be atretic. Fifty-five to seventy percent of double aortic arch (DAA) is right dominant, 20 to 35% left dominant, and 5 to 10% balanced.[4] In most cases, the clinical presentation of DAA is often occurred early in life, usually within the first 6 months starting in the neonatal period. The symptoms are mainly respiratory due to the tracheal compression. Early diagnosis needs high clinical suspicion because symptoms may mimic asthma.[2–4] Herein, we present a 5-month-old girl who had DAA. Her anatomical aberration was diagnosed by thoracic aorta computed tomography angiography (CTA), and the anomaly mimicked the clinical characteristics of life-threatening severe reactive airway disease (RAD). DAA may go with severe tracheal obstruction that mimics the RAD.

Case Report

A 5-month-old girl was admitted to the emergency room (ER) with an extreme exacerbation of RAD (mild cyanosis, tachypnea, intercostal, and subcostal retractions) and respiration insufficiency. The patient was immediately transferred into the pediatric intensive care unit (PICU). The patient did not have any allergies. The patient was not taking any medications at the time of her admission into the ER. According to the patient's medical history, the patient had already been hospitalized in the inpatient clinic two times before her admission into our ER. During all of her hospitalizations, the patient had used nebulized salbutamol and budesonide. The patient was born full-term, via a cesarean delivery. The patient's birth weight was 3.25 kg. There was no family history of any relevant medical or surgical issue. Physical examination in the ER revealed significant tachypnea and both intercostal and subcostal retractions. Her pulse rate and blood pressure were 110 beats per minute and 100/70 mm Hg, respectively. Her oxygen saturation was 90% in room air. On pulmonary auscultation, her expiration was prolonged. Cardiovascular exam and rest of the systemic exam were unremarkable.

Laboratory investigations and acute phase markers appeared to be normal. However, the patient had respiratory acidosis (pH; 7.25, pCO_2; 68 mm Hg, hCO_3; 28 mEq/L). Chest X-ray revealed air confinement and the patient's ribs, which were flat, and her diaphragm were flattened bilaterally (►**Fig. 1**). Noninvasive respiratory support of nasal cannula was started with a high flow of oxygen for the patient's respiratory insufficiency. The patient was prescribed with nebulized salbutamol, budesonide, intravenous magnesium, and methylprednisolone as a preemptive measure due to the patient's history and physical examination. Also, chest X-ray was consistent with the clinical manifestations of exacerbated RAD. Despite the medical management for RAD with salbutamol, budesonide, intravenous magnesium, methylprednisolone, and oxygen supplementation with nasal cannula, her respiratory finding did not improve. Due to worsening respiratory distress and increased blood carbon dioxide level, bilevel positive airway pressure, noninvasive respiratory support was started. Ketamine was used as a sedative during the procedure.

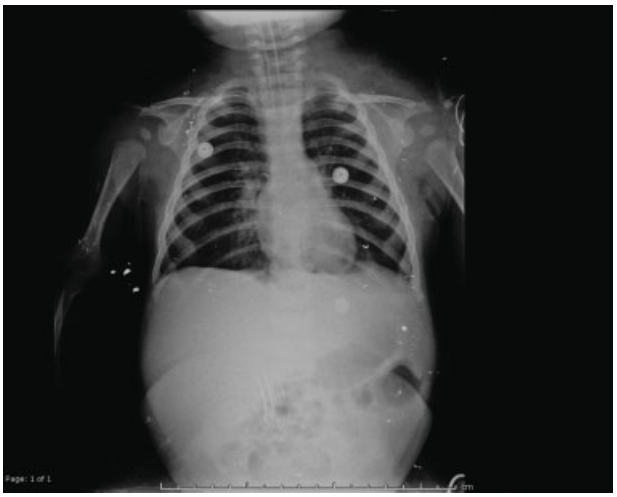

Fig. 1 Chest X-ray showing air confinement and the diaphragm was flattened bilaterally.

Despite all these treatments, the findings of respiratory failure had not been regressed at all. The patient's CO_2 levels in her blood gas gradually increased to 98 mm Hg. Despite the 48 hours treatment of bilevel positive airway pressure, the condition of the patient still persisted. Then, the patient was intubated immediately. All mechanical ventilation methods were used to remedy the obstructive airway condition as well. Despite all of these efforts and the intravenous aminophylline treatment, the patient's CO_2 gas level gradually increased up to 145 mmHg. Transthoracic echocardiography (TTE) was unremarkable and did not show vascular ring. We did not observe any significant improvement in physical examination, blood gas, and radiological findings of the patient, so our team decided to perform diagnostic bedside bronchoscopy on the fifth day of the admission into the PICU. The bronchoscopy results were quite intriguing. Approximately 1 cm above the carina, there was severe tracheal narrowing with near complete occlusion of trachea. One of the differential diagnoses of tracheal obstruction was congenital vascular anomaly. Three-dimensional CTA of thorax was performed for a definitive diagnosis. CTA showed DAA joining as a single right descending aorta. DAA was right dominant. Also, the trachea and esophagus were severely compressed by DAA (►**Figs. 2** and **3**).

The patient was operated for the correction of DAA by a cardiovascular surgeon on the seventh PICU day. The operative course was uneventful, and the patient was brought to the PICU while she was still intubated. The patient's respiratory symptoms did not immediately improve after the surgery due to the development of tracheomalacia, so therefore the patient was extubated 3 days after the surgery and weaned off respiratory support on the tenth day of PICU admission. Also, the patient had mild respiratory problems, and she was discharged on the fifteenth day of PICU admission.

Discussion

RAD is a prevalent condition that can be detected in the early infancy period. Wheezing is a loud, whistling sound

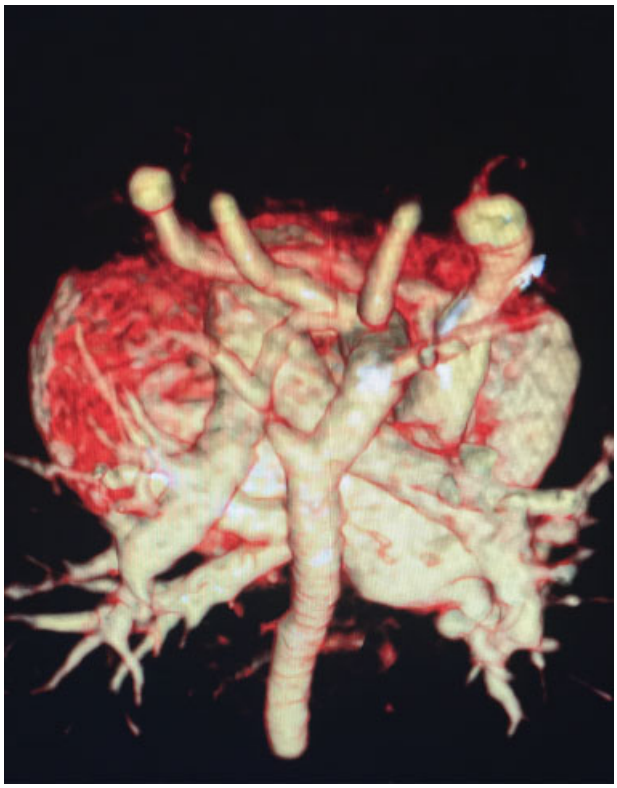

Fig. 2 Three-dimensional thorax computed tomography angiography showing the superior view of the double aortic arch.

that happens when smaller airways are narrowed by the presence of bronchospasm or the swelling of the mucosal lining. The condition also manifests itself with excessive amounts of secretions or if a foreign body is inhaled.

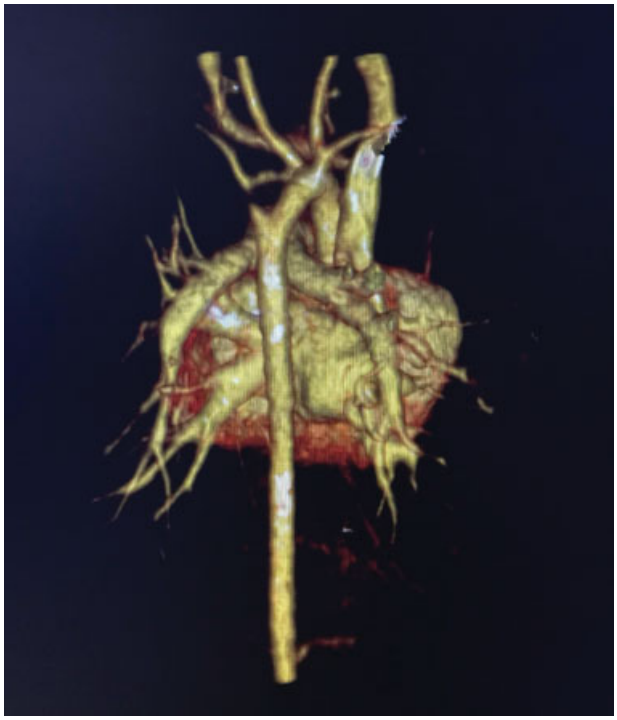

Fig. 3 Three-dimensional thorax computed tomography angiography showing the posterior view of the double aortic arch.

Clinicians mostly hear the state on the expiration phase due to the consequence of critical airway obstruction. The clinical manifestation of wheezing is extremely similar to the clinical presentations of allergies, asthma, and gastroesophageal reflux disease. All of these conditions should be kept in mind for infant's wheeze management. The condition might also deteriorate into asthma in some cases. If infants do not respond to the treatment of persistent wheeze and coughing, other rare causes should be investigated. Other etiologic factors should be investigated carefully for patients who are below a year old and do not respond to the treatment well.[2] The complete form of vascular ring is an extremely uncommon congenital cardiovascular abnormality.[5,6] The symptoms of the condition are mainly respiratory due to the tracheal compression, and the condition might mimic the clinical symptoms of asthma. There are a few reports in the literature about patients with DAA being mistreated for asthma in their long-term follow-ups.[2–5] As far as we can be sure, this will be the first case reported in the literature who had a deleterious version of RAD, which worsened into a life-threatening condition, and eventually, the patient had to be hospitalized in the PICU.

Piloni et al[3] reported a 20-year-old patient, and Lone et al[4] reported a 60-year-old chronic patient who had similar conditions as our patient. Despite the advent of modern medical techniques and treatments, both cases' clinical conditions did not improve at all. The cases' initial diagnoses of tracheal compression were made via bronchoscopy. The condition of DAA for both cases was confirmed by thorax CTA. Our case presented with similar clinical features of both cases, which were already mentioned above. The diagnostic bedside bronchoscopy was done, and the tracheal compression was noted in our patient. Three-dimensional thorax CTA was performed for a definitive diagnosis. However, the condition of DAA could not be detected on an echocardiographic examination of our case. The optimal treatment of options for these types of cases is usually the surgical division of the nondominant branch of DAA from the dominant branch of DAA.[7,8] Our patient was operated on the seventh day of her hospitalization. The compression of the trachea and esophagus due to the DAA was observed during the surgery, and the posterior aortic arch was divided to release the obstruction.

Conclusion

TTE is the primary noninvasive diagnostic tool for an initial evaluation of cardiovascular anomalies. TTE is not an optimal diagnostic tool to evaluate great vessels. Especially in the case of DAA, CTA is a far better tool than TTE for assessing great vascular anomalies and airways. Clinical symptoms of DAA conspicuously show themselves due to the compression of the trachea. Physicians, especially pediatric pulmonologists, should keep in mind the condition of DAA to evaluate the different forms of asthma-like conditions that do not respond to the treatment of asthma well.

Note
All authors participated in creating content for the manuscript, editing, and provided final approval for submission. No undisclosed authors contributed to the manuscript.

Ethical Approval
This article does not contain any studies with human participants or animals performed by any of the authors. Institutional Review Board approval was not required for this case report.

Funding
None.

Conflict of Interest
None declared.

Acknowledgments
The authors would like to thank parents and patient who granted the permission to publish this case report.

References

1 Lei WT, Lin HH, Tsai MC, et al. The effects of macrolides in children with reactive airway disease: a systematic review and meta-analysis of randomized controlled trials. Drug Des Devel Ther 2018;12:3825–3845
2 Javad H, Al-Sineidi K, Abdelmogheth AA, et al. Vascular anomalies in children misdiagnosed with asthma: case series. Sultan Qaboos Univ Med J 2015;15(01):e136–e139
3 Piloni D, Tirelli C, Domenica RD, et al. Asthma-like symptoms: is it always a pulmonary issue? Multidiscip Respir Med 2018; 13:21
4 Lone GN, Rathore SS, Malik JA, Ashraf HZ, Qadri AA. Double aortic arch masquerading as bronchial asthma for five decades. Asian Cardiovasc Thorac Ann 2012;20(03):338–340
5 Zhang Q, Fu Z, Dai J, Geng G, Fu W, Tian D. Recurrent wheezing and cough caused by double aortic arch, not asthma. Case Rep Cardiol 2017;2017:8079851
6 Balfour-Lynn I. Difficult asthma: beyond the guidelines. Arch Dis Child 1999;80(02):201–206
7 Kliegman RM, Stanton B, Geme J, Schor NF, Behrman RE. Annals of Internal Medicine, vol. 19. Philadelphia: Elsevier; 2011
8 Calabrese C, Corcione N, Di Spirito V, et al. Recurrent respiratory infections caused by a double aortic arch: the diagnostic role of spirometry. Respir Med Case Rep 2013;8(01):47–50