Final Diagnosis of Resisting Asthma: Incomplete Double Aortic Arch with Distal Left Arch Atresia

Dirençli Astım Tanılı Hastaya Nihai Tanı: Distal Sol Ark Atrezili İnkomplet Çift Aortik Ark

İnkomplet Çift Aortik Ark / Incomplete Double Aortic Arch

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Özet

İnkomplet çift aortik ark nadir bir vasküler anomalidir. Bu vakada dispne ve disfaji şikayeti ile gelen astım yanlış tanılı inkomplet çift aortik ark olgusunun karakteristik görüntüleme bulguları sunulmuştur.

Anahtar Kelimeler

İnkomplet Çift Aortik Ark; Astım; Bilgisayarlı Tomografi

Abstract

Incomplete double aortic arch is a rare vascular anomaly. We describe a case of distinctive imaging features of incomplete double aortic arch misdiagnosed as asthma that admitted with dyspnea and dysphagia.

Keywords

Incomplete Double Aortic Arch; Asthma; Computed Tomography

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Introduction

Incomplete double aortic arch is a rare anomaly resulting from atresia. This anomaly is a rare cause of a potentially symptomatic vascular ring[1, 2]. Incomplete double aortic arch anomalies look like typical double aortic arches, but atresia of a fragment of the left arch is present, resulting in a non-patent fibrous cord[2]. This anatomic vascular anomaly may cause stridor, wheezing or dysphagia[3]. Contrast-enhanced CT is a important tool in evaluating patients with aortic arch abnormalities and it allows for multiplanar visualization of aortic anatomy while clearly depicting any associated mass effect on the adjacent trachea or esophagus. We describe distinctive imaging features in a case of incomplete double aortic arch misdiagnosed as asthma.

Case Report

We present a ten-year-old girl who admitted with dyspnea and dysphagia. Physical examination and history revealed cough, that didn't respond to medical asthma therapy. In order to show relation between the tracheobronchial tree and the abnormal vascular structure, contrast-enhanced multi-detector CT was performed. Axial MIP images showed right archus aorta, incomplete double aortic arch with left arch atresia and aortic diverticulum (Fig. 1). Three dimensional reconstructed CT images made it easier to see the narrowing of the tracheobronchial tree (Fig. 2).

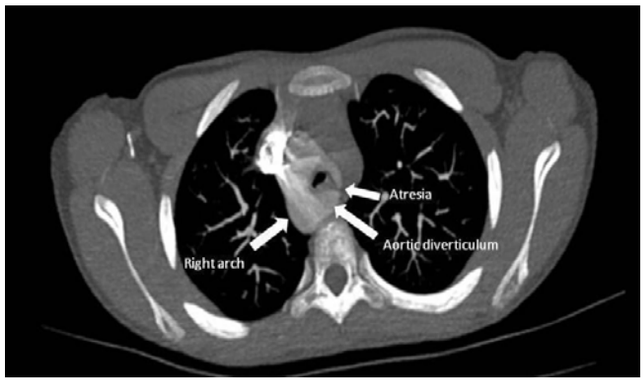


Figure 1. Axial MIP image show right archus aorta, incomplete double aortic arch with left arch atresia and aortic diverticulum

Discussion

All reported vascular rings represents less than 1-% of all congenital cardiovascular anomalies. Incomplete double aortic arch occurs because of atresia during the embryonic period at any point on either side of double aortic arch[2]. Incomplete double aortic arch include a non-patent fibrous cord connecting the descending aortic diverticulum[1]. This potential vascular ring usually shows itself with tracheoesophageal compression.

The lack of complete left arch easily distinguishes incomplete double aortic arch with distal left arch atresia from complete double arch. Incomplete double aortic arch include two types depending on localization of atresia. Atresia occurs distal to left ductus with resulting fibrous cord that inserts in descending aortic diverticulum (subtype 1). Atresia occurs between left subclavian artery and ductus with resulting fibrous cord. Both cord and left ductus insert in aortic diverticulum (subtype 2). These two forms of incomplete double aortic arch are indistinguishable by imaging, because the ductus and the fibrous cord are not visible on MR or CT images[1].

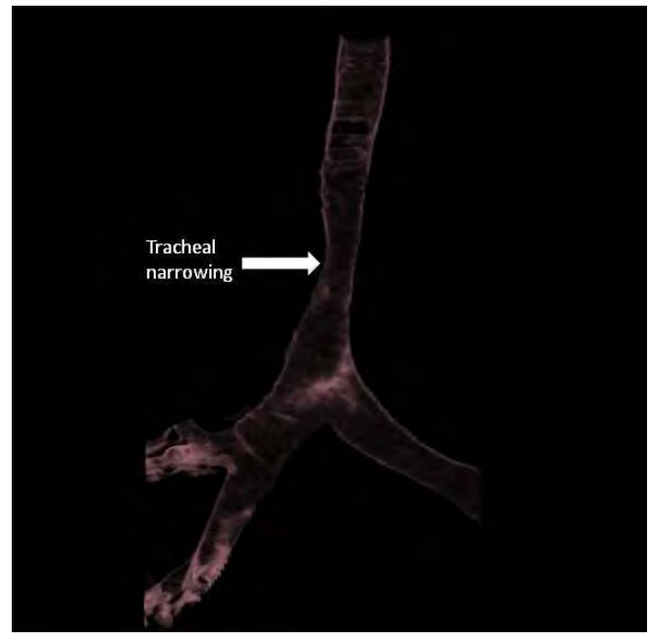


Figure 2. Three dimensional reconstructed CT image makes it easier to see the narrowing of the tracheobronchial tree

Relationship with adjacent structures of aortic arch can be accurately defined by CT and MR. These methods allow virtual demonstration of vessels with possibility of 3-D display. Additionally, the vessels are assessable by MR techniques not requiring intravenous contrast injection. In our case MR angiography could not be examined because of technical problems on the device. We tried to demonstrate 3-D definition and multiplanar reconstruction of the airway and aortic diverticulum with multidetector contrast-enhanced CT. Identification and demonstration of some of the anatomical features such as tracheal narrowing, esophageal compression, aortic diverticulum are important for the surgeon to be aware of anatomic possibilities and to predict the course of operation.

Most congenital abnormalities of the aortic arch cause respiratory symptoms and swallowing difficulties[4]. Main symptoms of our patient were also dysphagia and dyspnea. Some cases reported different presentations like heart failure, widening of the superior mediastinum on chest radiograph[5], recurring pneumonia[3], postprandial choking and respiratory distress[6]. Two cases with incomplete double aortic arch reported as misdiagnosed asthma in the literature[7, 8]. Similarly, the present case had been treated for asthma for 8 years but symptoms hadn't completely reduced.

In conclusion, vascular ring abnormalities such as incomplete double aortic arch with distal left arch atresia should be in mind for the differential diagnosis of asthma and dysphagia combination in early ages. Contrast-enhanced CT may provide reliable diagnostic information for therapy planning.

Competing interests

The authors declare that they have no competing interests.

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