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*Case Report / Olgu Sunumu*

# Cervical aortic arch

## *Servikal aortik ark*

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**ABSTRACT**

Cervical aortic arch is an unusual congenital anomaly. It is usually symptomatic in childhood, but sometimes this abnormality is diagnosed during radiological examination because of unrelated causes in asymptomatic adults. We aimed to report a case of cervical aortic arch and review of the literature which presented with cough and shortness of breath.

**Keywords:** Cervical, aortic arch, shortness of breath, dyspnea

**ÖZ**

Servikal aortik ark, nadir görülen bir konjenital anomalidir. Çocuklukta genellikle semptomatiktir, ancak asemptomatik erişkinlerde bazen bu anormallik farklı nedenlerden dolayı radyolojik görüntüleme sırasında teşhis edilir. Biz bu yazıda öksürük ve nefes darlığı ile başvuran servikal aortik ark olgusunu ve literatürün gözden geçirilmesini amaçladık.

**Anahtar Kelimeler:** Servikal, aortik ark, nefes darlığı, dispne

## INTRODUCTION

Cervical aortic arch (CAA) is a unusual congenital anomaly in which the ascending aorta elongate in such a way that the aortic arch is situated higher to its usual position (1) . The position of the arch different from lightly superior to normal to very high in the neck, lying on either side of the trachea . Patients is usually asymptomatic. But, it may cause cough and dyspnea, especially during exercise (2). We here report a cases of CAA, which is a rare case, presented with cough and shortness of breath.

## CASE REPORT

A 48-year-old female was admitted to emergency department suffering from cough and shortness of breath during both exercise. She was on salbutamol inhalation as well as budesonide/formoterol inhalation for 5 years and the symptoms did not ameliorate. Her blood pressure was 120/80 mmHg and all peripheral pulses were palpable. The breath and pulse rate were within normal limits. She had no significant family history. There was no history of change in voice, trauma, dysphagia, hemoptysis, or arm claudication. On inspection, the neck swelling appeared pulsatile. There was a pulsatile left supraclavicular and suprasternal mass. The palpation revealed an elongated soft neck swelling on the left side. There was no skin discoloration or erythema over the swelling. Routine laboratory investigations were normal. Chest radiograph did not reveal any information except a homogeneous shadow with sharp margins in the left apex and continuous with superior mediastinum (**Figure 1**).

Computed tomography (CT) revealed an elongated ascending aorta with cervical aortic arch and the tracheal compression (**Figure 2-3**). The arch was at the level of C-7 vertebra. The descending aorta was seen to cross the midline to descend on the left side. No vessel was seen arising from the aortic arch. The brachiocephalic trunks were absent and all major vessels were arising separately. No associated cardiac anomaly was evident on CT scan. The patient was referred to cardiovascular surgery with a diagnosis of CAA.

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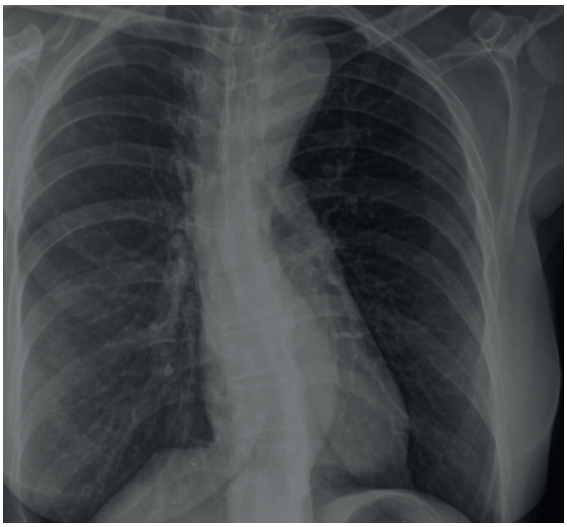

**Figure 1.** Chest radiograph shows homogeneous shadow of the aortic knob

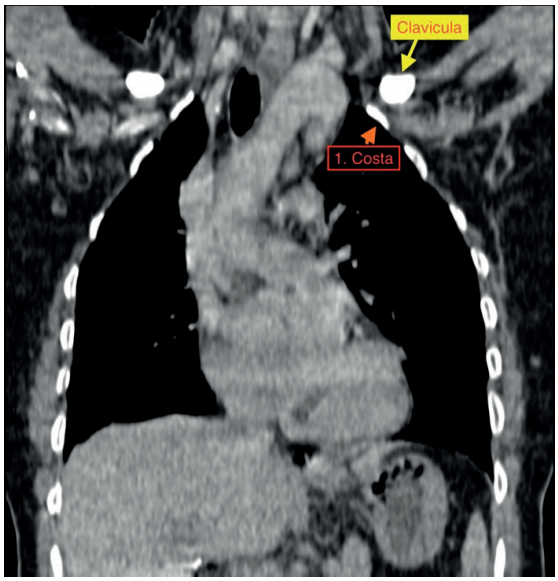


**Figure 2.** Computed tomography of the thorax shows cervical aortic arch at the level of C-7 vertebra

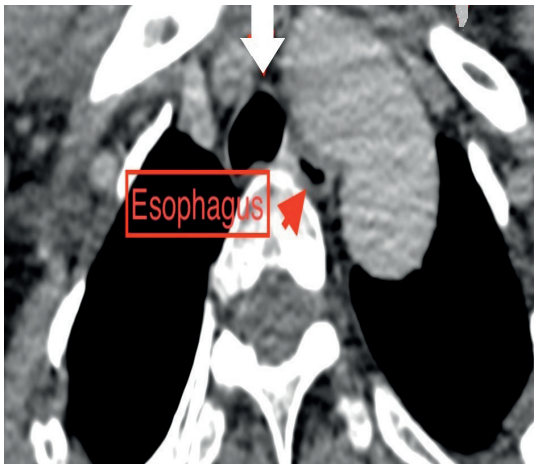


**Figure 3.** Computed tomography of the thorax, compressed tracheal lumen and cervical aortic arch is shown (white arrow: trachea)

## DISCUSSION

CAA is a rare congenital anomaly resulting from cephalic displacement of the aortic arch. Haughton identified five different forms on the basis of the aortic configuration, brachiocephalic branching and embryogenesis sequence. Type A has separate external and internal carotid artery branches from the aortic arch, type B has dual common carotid arteries, type C is a left cervical arch with right sided descending aorta and bicarotid trunk, type D has normal brachiocephalic branching, redundant transverse aorta and left-sided descending aorta, type E is a right cervical aortic arch with a right descending aorta and an aberrant left subclavian artery (3).

Most patients with this malformation are asymptomatic, but patients may present with esophageal and tracheal vascular compression indicative of dysphagia, dyspnea or frequent pulmonary infection. Dysphagia and respiratory distress are associated with the right cervical arc, while aneurysmal formations are more commonly associated with the left cervical arc. Physical examination reveals a large pulsatile mass in the supraclavicular area. CT, angiography and magnetic resonance angiography are useful for specific diagnosis (4). The case of our patient demonstrates the importance of suspicion of a vascular ring in patients with frequent respiratory symptoms. This congenital vascular abnormality and others are diagnosed optically by computed tomography or magnetic resonance imaging and angiography.

A variety of cervical arched associations include aneurysms, aortic coarctation, aortic pseudo-coarctation, congenital cardiac anomalies, Di George's syndrome and Turner's syndrome. Complications usually result from the compression of the brachial plexus resulting in aneurysmal dilatation, coarctation, dissection or cervical radiculopathy. These conditions require surgical management (5).

Investigations suggest that a cervical aortic arch is associated with deletions in the chromosome 22q11 and therefore may be included in the spectrum of defects known as catch 22. This syndrome, described by Wilson and colleagues in 1993, is characterized by cardiac defects, facial dysmorphic, thymic hypoplasia, cleft palate, hypocalcaemia and a deletion in chromosome 22 (2).

Treatment includes excision and thoracic re-localization and surgical correction. Other options include an endovascular stent to exclude aneurysms and dissections (6).

In conclusion, CAA might be considered in differential diagnosis of patients who admitted to emergency department suffering from cough and shortness of breath.

## ETHICAL DECLARATION

Patient approval was obtained.

## MATERIAL SUPPORT AND RELATIONSHIP

There is no person / organization to support the work financially and the authors have no conflict of interest.

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