Transthoracic Echocardiography Evaluation of Left Cervical Aortic Arch with Aneurysm Formation

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Noninvasive echocardiography can have invaluable information prior to studies such as computed tomography or invasive angiography. A 23-year old man presented to our hospital with a chest x-ray showing widened upper mediastinum. Echocardiography demonstrated color flow jets in opposite directions, compatible with tortuous aneurysmal dilatation of the aortic arch at the cervical position. The 640-slice computed tomography later confirmed the developmental anomaly of cervical aortic arch.

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Introduction

Cervical aortic arch (CAA) was first described by Reid in 1914 and approximately 150 cases have been reported in literature up to 2009 [1]. A rarer finding is CAA with aneurysm formation, which constitutes about 20% of all cases [2]. The cause of this congenital anomaly is the persistence of second or third dorsal arch, confluence of third and fourth dorsal arches, or failure of normal caudal migration of the fully developed fourth arch [2,3]. Other congenital anomalies such as tetralogy of Fallot, ventricular septal defect or double-outlet right ventricle are associated in 30% of CAs [4]. Diagnosis of CAA usually required aortography, computed tomography, magnetic resonance angiography, and, rarely, echocardiography [5].

Case report

A 23-year-old male was referred to our cardiovascular surgery department for evaluation of a widened upper mediastinum noted on the service enlistment chest x-ray (Fig. 1). The patient described no physical discomfort except left facial...
and left upper arm anhidrosis since age 10 and mild left arm weakness with exertion since age 14. Upon further questioning, the patient revealed pulsatile senation at left supraclavicular area after heavy exercise. On physical examination, he appeared comfortable and had no obvious abnormal facies or external abnormality around the left neck area. Hair at the left axilla and left arm showed equal distribution and density as right side. Auscultation of the chest revealed regular heart beat with normal S1 and S2. Echocardiography from the suprasternal approach showed aortic arch with aneurysmal dilatations above normal position (Fig. 2). On color Doppler, two opposite color flow jets were visualized (Fig. 3), suggestive of tortuous nature of the arch. A 640-slice CT was ordered and revealed aortic arch aneurysm extending above the clavicle before becoming the descending aorta. The largest aneurysm measured dimension at 80 mm / C2 \(44 \text{ mm} \times 75 \text{ mm}\). Cardiac catheterization showed a normal coronary artery and essentially confirmed the CT finding. The patient subsequently underwent surgical intervention with resection of distal arch (cervical arch) and proximal descending aorta, interposition of 20 mm woven graft, and reimplantation of the left subclavian artery with a Goretex ring 8 mm from the ascending aorta. The patient was well after the surgery and follow-up CT performed 45 days later revealed graft insertion between the ascending aorta and left subclavian artery and a stationary aneurysm of the distal aortic arch (4.6 cm) with development of mural thrombus.

**Discussion**

CAA is defined by aortic arch situated cranially above the clavicle [2]. Major clinical findings include palpable neck mass, dysphagia, wheezing, coughing, stridor, choking, apneic spells, and recurrent pulmonary infection, even traumatic aortic rupture, or no symptoms at all, depending on the location and extent with or without aneurysm formation [6,7]. The patient usually presents between the ages of 20 and 40 years with equal sex distribution with aneurysm formation predominantly occurring in women [8].

In 1948, Edward’s hypothesis described concepts of embryological double aortic arch system with an aortic arch and a ductus arteriosus on each side [9,10]. The right carotid and subclavian arteries arise from the right arch; the left carotid and subclavian arteries originate from the left; and the descending aorta is in the midline. Normal arch system results from interruption of the dorsal segment of right arch between the right subclavian artery and descending aorta, with regression of the right ductus arteriosus. Interruption of this arch system at different locations can explain the various aortic arch anomalies, including CAA as a result. Our case is the result of CAA with aneurysm formation between the left common carotid artery and left subclavian artery (Fig. 4), most likely to be the result of a confluence of the third and fourth aortic arches and the failure of the arch descent into the thorax. Initially described by Haughton et al [11], there are five distinct forms of CAA, which are classified according to configuration of the aorta, sequence of brachiocephalic branching, and embryogenesis. In brief, Type A has separate external and internal carotid artery branches from the aortic arch. Type B is similar but has dual common carotid arteries. Type C is a left cervical arch with a right-sided descending aorta and bicarotid trunk. Type D is a left cervical arch with a normal branching pattern, redundant transverse aorta and left-sided (often hypoplastic) descending aorta. Type E is a right cervical arch with a right descending aorta and an aberrant left subclavian artery [11]. Type D is the most common form of CAA and corresponds to our situation.

Our case of young male is unusual given the rare aneurysm formation (female-predominant and constituting 20% of all cases) on the distal end of a CAA. The patient was not observed with an obvious palpable mass but instead presented with history of ipsilateral anhidrosis and weakness on...
exertion. Echocardiography, once implemented in the study of a right-side CAA, positioned the probe at the right side of the neck only showed two-dimensional picture [5]; no color Doppler or pulse wave velocity was available. In our evaluation of this patient, echocardiography offered unprecedented hemodynamic information by noninvasive diagnostic means, in addition to visualization of the tortuous nature of the cervically-positioned aortic arch. To attain a better picture of aortic arch and its anomalies, thorough assessment through the suprasternal approach and at the neck in selected clinical scenarios is essential. In summary, comprehensive color Doppler echocardiography provides important knowledge of cardiovascular hemodynamics and cannot be replaced by other noninvasive modalities.

References


Fig. 3 (A) Color Doppler echocardiography showed color flow jet inside the lumen of the tortuous aortic arch. (B) Pulse-wave (PW) Doppler interrogation showed color flow directing toward the transducer with jet velocity 1.75 m/s. (C) PW Doppler showed color flow directing away from the transducer with jet velocity 2.25 m/s. The increased velocities suggested pseudo-coarctation with maximum pressure gradient of 20.25 mmHg at distal aortic arch.

Fig. 4 Computed tomography showed cervical aortic arch with aneurysm formation at different angles. Note the proximal end of aneurysm formed between left common carotid artery (LCCA) and left subclavian artery (LSA). RIA = right innominate artery.