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Case Report

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Isolated Left Subclavian Artery Associated with Right Aortic Arch: A Case Report

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ABSTRACT

An 18-day-old infant with a rare vascular anomaly characterized by an isolated left subclavian artery (ILSA) and a right aortic arch presented with respiratory distress. To gain a deeper understanding of this unusual combination, a literature review was conducted.

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Abbreviation

ILSCA: Isolated Left Subclavian Artery
RAA: Right Aortic Arch
CT: Computed Tomography
CHD: Congenital Heart Defects
d-TGA: dextro-Transposition of Great Arteries
VSD: Ventricular Septal Defect
PDA: Patent Ductus Arteriosus

Introduction

Isolation of the subclavian artery is a rare aortic arch anomaly, defined as a loss of continuity between the subclavian artery and the aorta, with a persistent connection to the pulmonary artery via a patent or non-patent ductus arteriosus [1, 2, 3]. Isolated left subclavian artery (ILSA) with right aortic arch is a rare association with an incidence of 0.8% in all right-sided aortic arch anomalies [4, 5]. It is typically associated with other congenital heart diseases, and clinical manifestations are variable [6].

We present the case of a newborn with RAA and isolated LSCA discovered during a thoracic CT scan

Case Report

An 18-day-old, 2.7 kg full-term baby girl hospitalized in the pediatric department for respiratory distress on unilateral left choanal atresia, a thoracic radiography has objectified a left pulmonary cystic image. The newborn was sent to our radiology department for a thoracic angioscan, which objectified the presence of a right aortic arch with an isolated left subclavian artery communicating with the trunk of the pulmonary artery via a patent ductus arteriosus; it also objectified the persistence of the left superior vena cava. In the parenchymatous window, she has right condensation of infectious origin with absence of pulmonary cystic lesion (figure 1)

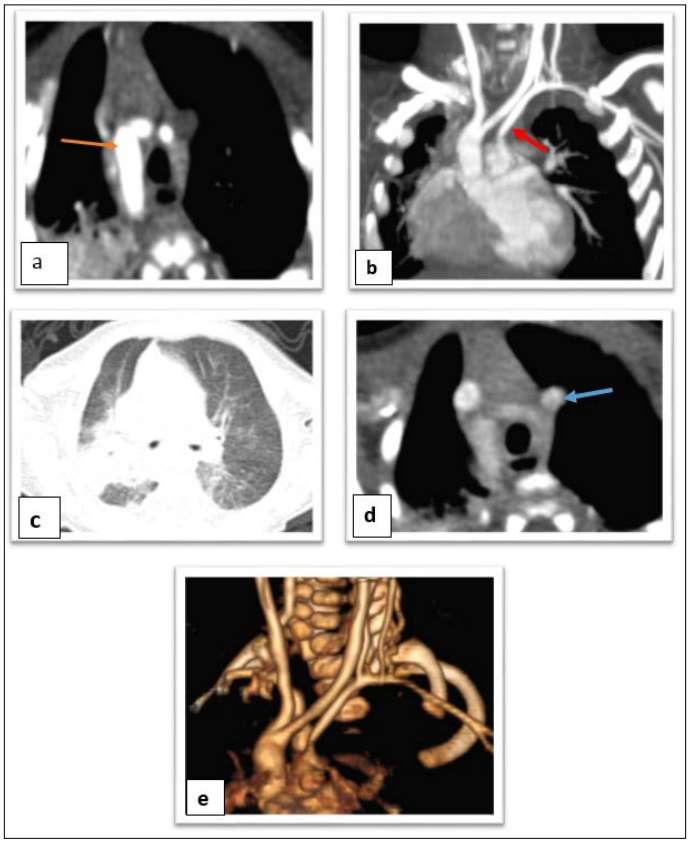

Figure1 : Computed tomography (CT) :a: right aortic arch(orange arrow); b: isolated left subclavian artery communicating with the trunk of the pulmonary artery via a patent ductus arteriosus(red arrow); c: right pulmonary condensation on paarenchymatous window; d: persitence of the left superior vena cava(blue arrow): e: 3D reconstriction

Discussion

Isolated left subclavian artery is a rare congenital malformation affecting the aortic arch system and is present in fewer than 1% of patients with right-sided aortic arches [4]. In this condition, the left subclavian artery is disconnected from the aorta and is exclusively connected to the pulmonary artery through the ductus arteriosus, which may or may not be patent [3]. The developmental basis for this malformation is described by the double aortic arch model, which posits that it results from two interruptions in the aortic arch: one between the left common carotid artery and the left subclavian artery and another between the left ductus arteriosus and the aortic root. This leads to a right aortic arch with the branches of the left common carotid artery, right common carotid artery, and right subclavian artery. The left subclavian artery is supplied by mediastinal or thoracic collateral vessels and receives blood retrogradely from the left vertebral artery [5, 7].

Embryologically, the double aortic arch model introduced by Edwards, explain clearly the development of isolated subclavian artery [1, 8]. In this model, the aortic sac is connected to the dorsal aorta via six paired aortic arches. Normally, the first two pairs of aortic arches form part of the vasculature of the face. The third pair of aortic arches forms the common carotid arteries, while the fourth aortic arches give rise to the part of the aorta between the common carotid and subclavian arteries. Each proximal sixth aortic arch forms the right and left pulmonary arteries, while the distal sixth arches form the ductus arteriosus on each side [9, 10]. Finally, the seven intersegmental arteries connect the vertebral arteries to the ipsilateral dorsal aorta at each level during development, but only the seventh pair of intersegmental arteries persists at each side as the subclavian arteries. The normal left aortic arch develops from interruption of the dorsal segment of the right arch between the right subclavian artery and the descending aorta with regression of the right ductus arteriosus, along with persistence of the left aortic arch and left ductus arteriosus. Accordingly, RAA with a mirror-image branching pattern occurs when there is involution of the right ductus arteriosus and the left dorsal arch between the LSCA and descending aorta [10].

In isolated left subclavian artery, there are two interruptions in the aortic arch. The first occurs between the left common carotid artery and the left subclavian artery (LSCA), while the second occurs between the left ductus arteriosus and the aortic root. This leads to a right-sided aortic arch with an isolated LSCA (figure 2). The branches of the right aortic arch are arranged in the order of the left common carotid artery, right common carotid artery, and right subclavian artery. The LSCA is disconnected from the aorta and connected to the pulmonary artery through the ductus arteriosus, which may be patent or closed [11, 12]. The left vertebral artery supplies blood retrogradely to the LSCA, which may also be supplied by mediastinal or thoracic collateral vessels [6, 10, 13].

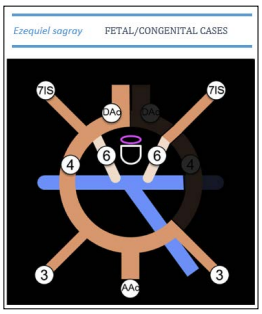

Figure 2: Diagram of theoretical double aortic arch, showing involution of left fourth aortic arch and left descending aorta (DAo), leaving the isolated LSCA supplied by the left ductus arteriosus.AAo, ascending aorta

The majority of patients with isolated left subclavian artery having an association with another congenital heart disease [1, 3]. A review of the literature by Alhuzaimi et al of 52 cases of isolated LSCA showed the association with another CHD in 84% of which the most frequent are tetralogy of Fallot, right-sided PDA, and large VSD and dextro-transposition of great arteries (d-TGA).

Clinically, patients with ILSCA are often asymptomatic; when they are symptomatic, it can be during the neonatal period or later in life. Patients may present with a subclavian steal syndrome secondary to vertebrobasilar insufficiency and manifesting as paroxysmal vertigo, headache, or syncope [14, 15]. They may also present with symptoms related to the hypoperfusion of the left upper limb, such as paresthesia, coldness, pain, weakness, or even hypotrophy of the limb [6, 10, 13].

The same review of the literature by Alhuzaimi et al showed that the average age of diagnosis is 4.7 years, 48% of patients are asymptomatic, 36% have reduced pulse or blood pressure or both, 6% have limb claudication or ischemia, 10% with neurological symptoms and 4% with respiratory symptoms [5].

Transthoracic echography is the first-line investigation, it can show the right aortic arch with the subclavian artery isolated, in addition, it allows making the hemodynamic study, and if it is performed early, it allows seeing the PDA. CT scan angiography and MRI can be the second-line imaging modalities especially after the closure of ductus arteriosus, to better delineate the course and connections of the LSCA [5, 6].

For treatment, surgical repair is indicated in patients with associated congenital heart disease. For asymptomatic patients, surgical treatment is discussed on a case-by-case basis depending on the benefits and risks [14, 15]. Regular monitoring may be the strategy of choice, but when surgical treatment is indicated; the preferred approach is reimplantation of the subclavian artery, either through anastomosis or autologous saphenous vein graft placement, to the aortic root or the common carotid artery [5, 6, 10].

Conclusion

ILSC with right aortic arch is a rare congenital anomaly, often asymptomatic, the symptoms may be related to limb hypoperfusion or vertebrobasilar insufficiency, early diagnosis is essential to monitor patients and discuss surgical indications.

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