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Double Aortic Arch: An Unusual Cause of Laryngeal Dyspnea in Children - Case Report

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Abstract*: The double aortic arch is a rare anomaly of vascular malformations of the aorta, which develops in embryonic life. The clinical picture is usually silent, if not dominated by respiratory and/or digestive signs, particularly laryngeal dyspnea or dysphagia in newborns, infants and young children. Thoracic angioscan is the gold standard for diagnosis and treatment. The treatment is only surgical. We report the case of a 5-month-old infant, admitted for acute dyspnea with laryngeal stridor, whose interrogation reveals repetitive bouts of cyanosis during feedings, and the clinical examination reveals sibilant rales on auscultation. The radiological explorations carried out, in particular; a thoracic CT scan showed a double aortic arch joined by a thin ligament associated with a retroesophageal diverticulum encircling and stenosing the tracheo-esophageal axis. The baby was referred to a cardiovascular surgery department for possible surgical management.* ***Objective:*** *This clinical case challenges us to keep in mind embryological vascular malformations of the type of double aortic arch as an etiology, although rare, of acute dyspnea with laryngeal stridor in the newborn.*

Keywords: Double aortic arch, laryngeal dyspnea, Stridor.

Introduction

The double aortic arch is a rare anomaly of vascular malformations of the aorta; it develops in embryonic life. It is secondary to the lack of involution of the dorsal caudal aorta. **The clinical picture** is usually silent, if not dominated by respiratory and digestive signs, particularly laryngeal dyspnea or dysphagia in newborns, infants, and young children. **The thoracic angioscanner** is the gold standard in the diagnosis as well as in the therapeutic choice. **The treatment is only surgical** and consists of lifting the compression on the tracheo-esophageal axis.

We report an observation of double aortic arch in a dyspneic infant, with the contribution of imaging to diagnose this anomaly.

Patient and observation

Male infant, 05 months old, from a pregnancy carried to term, medical delivery by vaginal route, APGAR 10/10, without the notion of consanguinity or a particular personal or family history. Admitted to the pediatric emergency room for the management of acute laryngeal dyspnea.

The interrogation reveals the notion of a neonatal stridor with cyanosis accesses at the time of suckling.

Clinical examination revealed a tachypneic infant with a respiratory rate of 22cpm, a saturation of 98%, apyretic, without signs of respiratory struggle. Auscultation of the lung fields found sibilant rales.

The chest radiograph showed a filiform stenosing narrowing of the middle third-lower third junction of the trachea, with doubts of an aortic button on the right.

However, it did not target any pulmonary or cardiac parenchymal abnormality. **(Figure 01).**

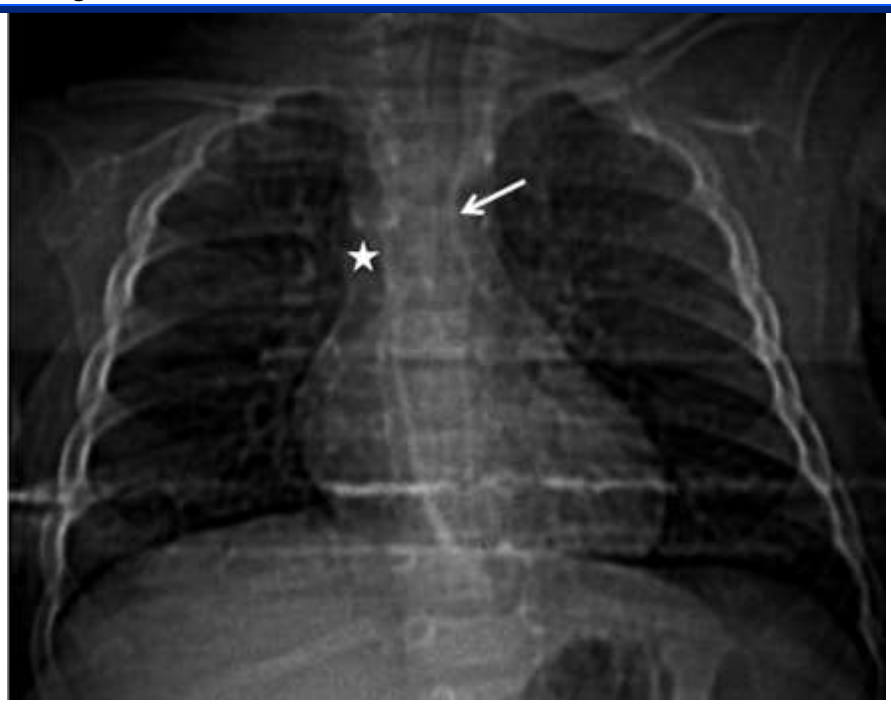

Figure 01: Chest X-ray showing a filiform narrowing of the middle third-lower third junction of the trachea (arrow), with doubt on a right aortic button (star).

A cervico-thoracic angioscanner showed a double aortic arch joined by a thin ligament associated with a retroesophageal diverticulum encircling and stenosing the tracheo-esophageal axis by 50% **(Figure 02-03).**

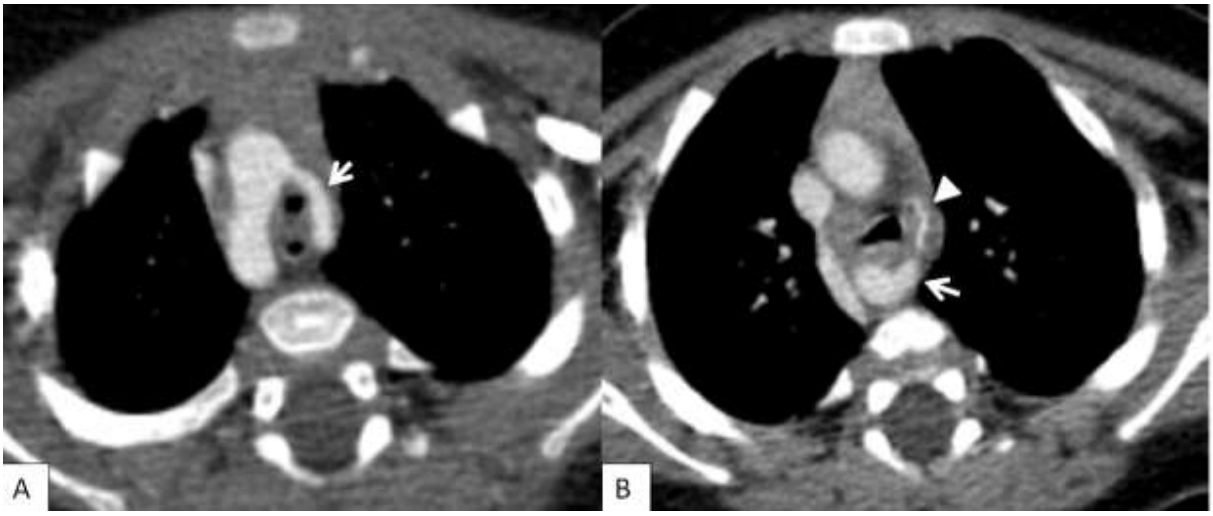

Figure 02: Angio Thoracic CT in axial section:

(Image.A)Double aortic arch with retro-esophageal aortic diverticulum (arrow), continued by a thin ligament going towards the right aortic arch (arrowhead) encircling the tracheoesophageal axis (Image.B).

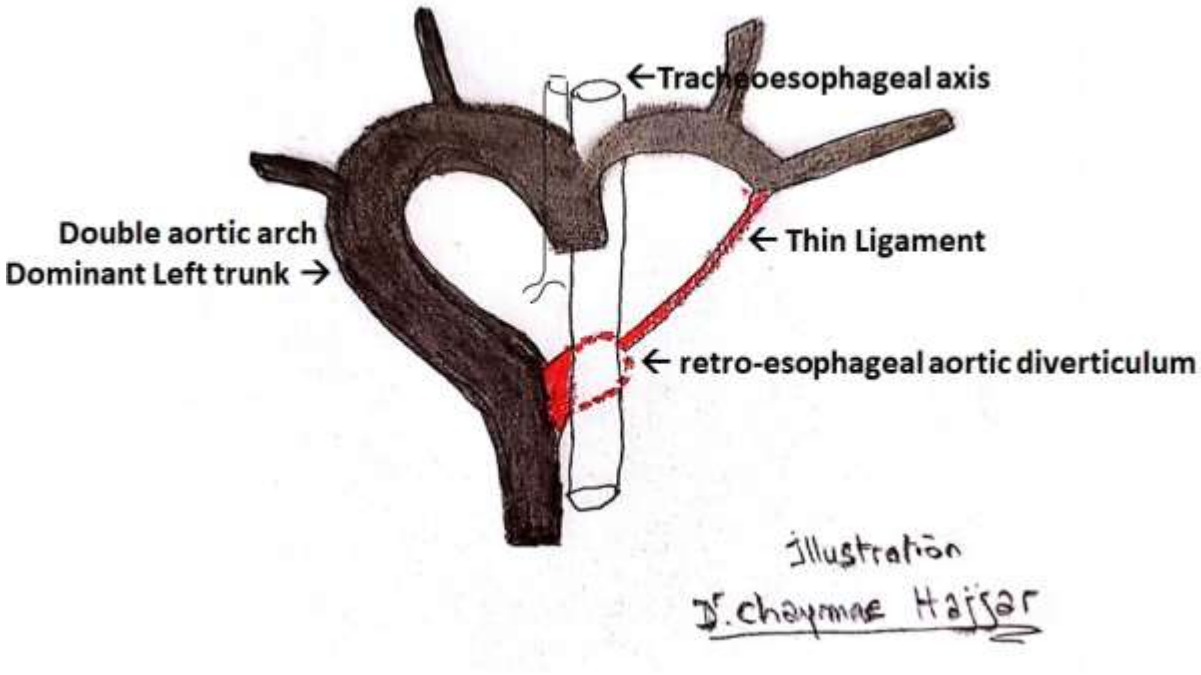

Figure 03: Illustrative diagram of our patient: Double aortic arch with dominant left trunk, associated with a retro-esophageal aortic diverticulum, continued by a thin ligament continuing to the right aortic arch encircling the tracheo-esophageal axis.

Paraclinical evaluation was completed 1 month after this acute episode by trans thoracic echography which did not reveal any other associated cardiac abnormalities.

The surgical indication was discussed with the pediatric and cardiovascular surgeons and the risks were shared with the family. However, the family refused any intervention because of their age and the complexity of the surgical procedure.

Discussion :

The double aortic arch is the most common form of symptomatic aortic vascular rings. It results from the persistence of the two fourth arches and consequently the formation of right and left aortic crosses on either side of the tracheo-esophageal axis before joining backwards to form the descending thoracic aorta. This is the most frequent form of complete annulus [1].

Usually, both arches are patent. Each provides the origin of the primary carotid artery and the homolateral subclavian artery. They have a variable caliber, thus determining three types of double aortic arch, which are: a dominant right aortic arch with a small left arch that can go up to atresia (80%), (as in the case of our patient); a dominant left aortic arch and a small right arch (10%), and balanced aortic arches (10%). [2]

There are several variants, according to several criteria, due to the independence of the embryological development of the different structures. The position of the subclavian artery and the descending aorta corresponds to an independent embryological origin, unrelated to this pathology. However, depending on the dominance of the arch, the ductus arteriosus may be located on the left or right, with the descending aorta usually deviating to the side of the ductus arteriosus. In the balanced arrangement, it can be bilateral [3].

The double aortic arch often goes unnoticed. However, it is complicated by respiratory symptoms (chronic cough, stridor, respiratory distress) and digestive symptoms (dysphagia). The importance of the signs depends on the space between the two aortic arches, depending on whether the compression forms a tight or loose ring. Dyspnea is the main revealing mode in infants and young children,

usually before 3 years of age, while dysphagia sometimes appears later in adulthood. The age of discovery of the disease was early in our patient (05 months), which is consistent with the results of the literature. [4]

Dyspnea is not a pathognomonic condition of a cardiac disorder. However, the recurrent and early character of dyspnea, which does not subside with appropriate treatment, should lead us to think of an embryonic vascular malformation, in particular a double aortic arch. This is the case of our patient who presented repetitive attacks of cyanosis and stridor, more marked at the time of suckling.

This congenital vascular anomaly can be isolated or complex, hence the interest in performing a **transthoracic echocardiography** in search of an associated cardiac malformation such as Fallot tetralogy, interventricular communication, persistence of the ductus arteriosus, open septal pulmonary atresia, coarctation of the aorta. [5]

Imaging plays an important role in the diagnosis of double aortic arch.

Chest radiography is the first-line examination. It allows one to orient the etiological diagnosis toward a vascular compression. However, a right aortic arch usually mimics a right upper mediastinal mass [6].

Fibroscopy allows one to demonstrate a right anterior compression of the trachea by the right aortic arch; to specify also its degree, and to eliminate differential diagnoses. [7]

The oeso-gastro-duodenal transit sometimes allows, depending on the location and orientation of the impression, to specify the type of anomaly. In practice, barium transit should be one of the first examinations performed in a child suspected of having an encircling anomaly of the aortic arch [8].

The thoracic angioscanner is the reference examination. Today, thanks to the new technology, an angioscanner with reconstructions gives excellent images in tridimensions, and offers a high specificity and sensitivity. It specifies the detailed and typical anatomy of the double arch as was the case in our patient: dimensions and position of each arch, dominance of one in relation to the other, relationship between the vascular ring and the trachea, degree of tracheal and esophageal compression, position of the supraaortic trunks on the arches [9].

Trans thoracic ultrasound offers a complete and comprehensive morphological evaluation of associated congenital heart disease.

Magnetic resonance imaging MRI is an alternative to CT. It is neither irradiating nor invasive, however, it is limited by respiratory and cardiac artifacts and by the less good study of the esophagus and trachea; and requires in children a heavy sedation. [10]

Treatment is exclusively surgical if the esotracheal compression syndrome is severe and life-threatening.

Conclusions

In front of dyspnea associated with a laryngeal stridor that does not regress in a child younger than 06 months, it is necessary to keep in mind malformative pathologies of the aortic arches, even if rare, such as a double aortic arch. Angiography and cardiac echography are of great diagnostic interest as well as in the choice of the therapeutic approach. Surgical treatment is indicated if the symptomatology is life threatening.

CONFLICT OF INTEREST

None.

Consent to publish:

Written informed consent was obtained from the parents. They consented to the submission of the case report to the journal.

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