CASE REPORT

Double aortic arch. Case report

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Double aortic arch is a rare pathology but it is the most frequent variant of a complete vascular ring. It occurs when the ascending aorta bifurcates into two arches, one posterior right and the other anterior left. Both arches surround the trachea and esophagus and converging into descending aorta. For all the above, respiratory or digestive symptoms could appear. Management is basically surgical. We report the case of a pediatric patient with double aortic arch successfully treated by surgery.

Key words: **Aorta; Congenital heart disease; Double aortic arch; Vascular ring.**

El doble arco aórtico es una patología rara pero es la variante más frecuente de anillo vascular completo, ocurre cuando la aorta ascendente se bifurca en 2 arcos, uno derecho posterior y otro izquierdo anterior, ambos arcos rodean la tráquea y el esófago y posteriormente confluyen en la aorta descendente, por lo mismo dan síntomas respiratorios o digestivos y su manejo es quirúrgico. Nosotros reportamos el caso de una paciente pediátrica con doble arco aórtico que es manejada quirúrgicamente de manera exitosa.

Palabras clave: **Anillo vascular; Aorta; Cardiopatía congenita; Doble arco aórtico.**

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Double aortic arch (DAA) is a relatively rare congenital cardiovascular malformation. It is the most common of the congenital vascular rings causing tracheo-esophageal compression. It is an anomaly of the aortic arch in which the 2 aortic arches are surrounding the trachea as well as the esophagus giving rise to a complete vascular ring that causes compression over the trachea, esophagus or both of them. Symptoms are related to such compression. High suspicion should be taken in patients with respiratory or digestive symptoms such as dysphagia without a specific cause [1].

DDA was firstly reported by Hommel in 1737. Gross performed the first successful surgical intervention in 1945 at the Boston Children's Hospital, laying the surgical principles foundation for vascular ring section. Operation is indicated in symptomatic patients [2].

We present here one clinical case about one pediatric patient having DAA underwent successful operation.

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A 6-years old female pediatric patient with diagnosis of DAA was admitted at our institution. With no important pathological history but a systolic murmur detected at one and

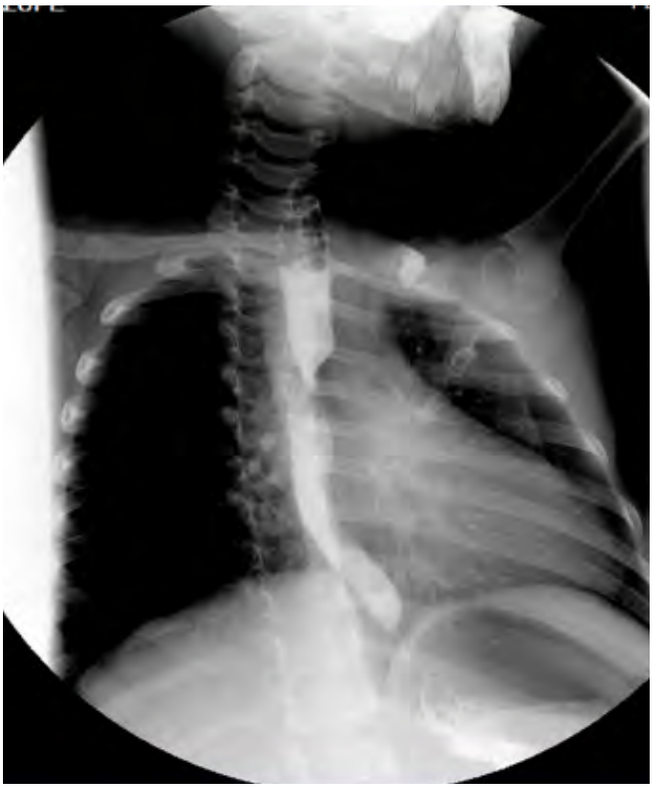

Figure 1. Esophagogram demostrating eaophageal external compression

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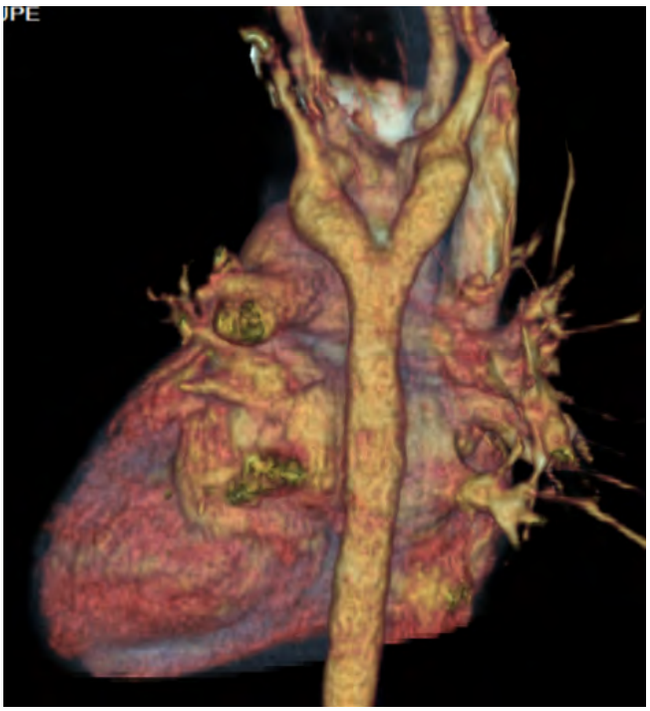

Figure 2. CT-angiography. The union of the left and right arches that compose the descending aorta can be seen.

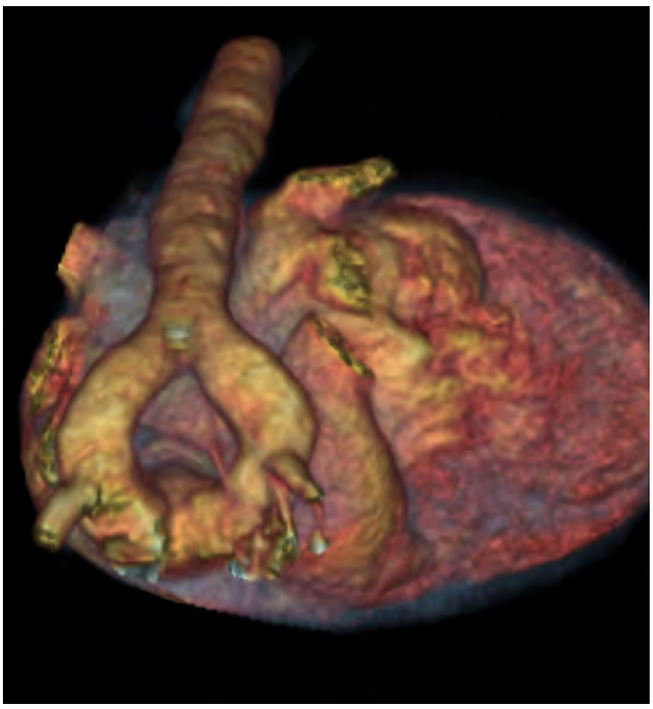

Figure 3. CT-angiography. The double aortic arch forming the complete vascular ring can be clearly observed here.

a half year of life, an echocardiographic study showed DAA. The left one was inferior and anterior where the left carotid artery and subclavian emerge; the right is superior and posterior where the brachiocephalic trunk emerges; both arcs of the same diameter, LVEF of 60%, PSAP of 24 mmHg, any structural alteration of the heart was ruled out. An esophagogram was also performed. An extrinsic compression of the esophagus was more than evident at the level of the carina with a 50% of luminal reduction (**Fig. 1**). CT-angiography let have an accurate and detailed anatomical description of the DAA and of the compression on adjacent structures. It demostrated both arch of similar diameter size. The left one giving rise to the left carotid and subclavian while the right one having the origin of the brachiocephalic trunk (**Fig 2**) (**Fig. 3**).

Operation was performed through left posterior-lateral thoracotomy via third intercostal space. After dissecting all anatomic strictures around, a plain identification of the descending aorta, ligamentous arteriosus, left aortic arch, supraaortic vessels took place. After a whole anatomic identification, aortic cross-clamping of the left aortic arch below the subclavian artery. After that, surveillance of the oximetry, blood arterial pressure, capillary refill time was closely carried out all along 10 minutes. No shred of any variation was obtained at the lower limbs. Based upon all this info, section of the left aortic arch below the left subclavian artery was performed (Fig. 4). No complications at the operating room, getting extubated to the ICU. Postoperarive course was uneventful. Length of stay in ICU was for 2 days, and in-hospital discharge was at 6º day after operation.

COMMENT

DAA is a very rare congenital malformation due to four primitive right and left arches. Vascular rings make up a small percentage of all congenital heart problems, less than 1%. The problem arises when having an aorta that is made up of two vessels instead of one. The two parts to the aorta have smaller arteries branching off of them. As a result, the two branches go around and press down on the trachea and esophagus. Most often, a right arch is the main one in 70% of all times. More rarely, both arches are symmetric. [1]. This is just the case we present here having both patent as well as symmetric arches.

Early diagnosis is difficult because of its normal or nearly normal clinical status with almost asymptomatic patients. When present, symptoms are secondary to tracheal and/or esophageal external compression from the vascular strictures as part of the vascular ring. Stridor, dysphagia, dyspnea and recurrent respiratory tract infections are some of the most frequent clinical manifestations. In our presented here, mild dysphagia made us strongly suspect the diagnosis. Thus, an intentional search by means of the esophagogram and ANGIO-TC confirmed our initial suspicion. In 20% of series, this pathology is associated with other cardiac defects [3]. In our case, an echocardiographic study demonstrated that it was just a single vascular ring with no other alteration.

Surgical treatment consists of cut-and-sew, surgical division of the smaller diameter size arch, below the subclavian artery in confluence with the descending aorta [4]. We

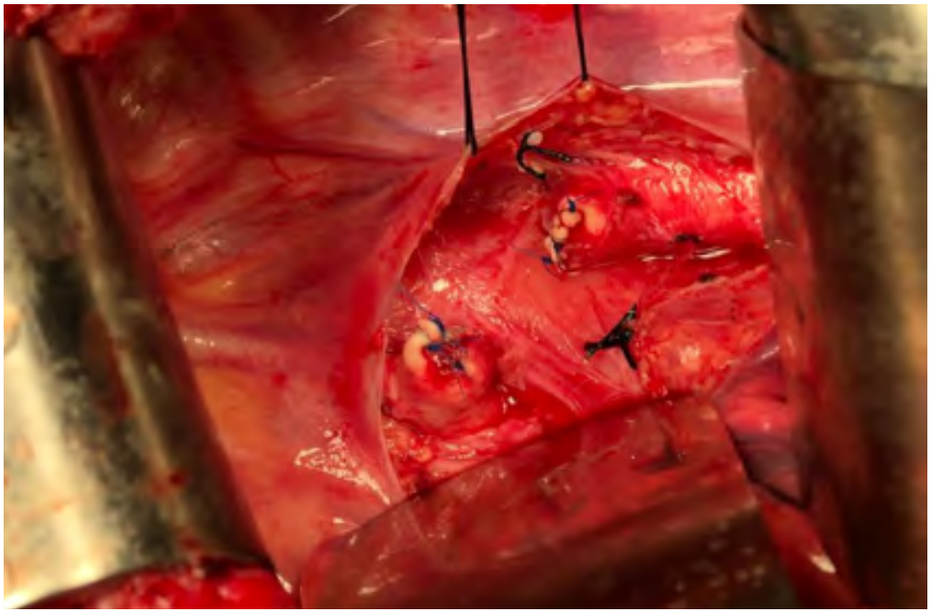

Figure 4. Operative view. Sectioned left aortic arch and both ends already sutured.

approached the vascular ring through a left posterior-lateral thoracotomy. In such a way, after analyzing both arches were very similar in size and diameter, we made up our mind to section over the left one, previously sectioned the ligamentous arteriosus.

Taking in mind that surgical complications could be a very lengthy list of, including phrenic nerve paralysis, vocal chords paralysis, bleeding, pneumonia, pneumothorax, chylothorax, residual dysphagia and dyspnea [2]. Nonetheless, our patient was free of complications at all.

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