CASE REPORT

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Single arterial trunk arising from the aortic arch with aortic coarctation. Report of a case. Embryological considerations

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Abstract

Abnormalities of the aortic arch occur mainly due to an abnormal development of the fourth aortic arch in the first 12 weeks of fetal life. These abnormalities may be asymptomatic for a long period, or can develop clinically when vascular rings surround the trachea and esophagus and cause tracheal and/or esophageal obstruction. We present the case of a full-term newborn baby, male. The pregnancy had no medical surveillance from a family physician or an obstetrical. After five days of age, the infant died due to severe malformations, incompatible with life, respectively due to single arterial trunk and aorta coarctation. In the literature, there have been cited several other cases similar to our case; however, this one remains an extremely rare anomaly. We believe that the abnormality is due to a regression on an abnormal site of the fourth left aortic arch, but due to the very small number of these anomalies reported so far, we cannot say with certainty if there is an embryologic reason for these abnormalities.

Keywords: aorta, congenital malformations, vascular embryogenesis.

Introduction

Abnormalities of the aortic arch occur mainly due to an abnormal development of the fourth aortic arch in the fourth in the first 12 weeks of fetal life. These abnormalities may be asymptomatic for a long time, or can develop clinically when vascular rings surround the trachea and esophagus and cause tracheal and/or esophageal obstruction [1]. Stridors, recurrent respiratory infections, shortness of breath, difficulty swallowing and gastro-esophageal reflux are symptoms resulting from these obstacles. The most common types of vascular compression anomalies are double aortic arch (DAA), right aortic arch and pulmonary ring [2]. Diagnostic tests may include chest X-ray, barium esophagography, angiography and, more recently, echocardiography, CT and MRI [3]. Surgical decompression is often necessary and normally improves immediate severe symptoms [4, 5]. Long-term results of surgical treatment are being evaluated but not knowing the exact long-term effects of this treatment on clinical symptoms and on pulmonary function [6].

In the literature, there have been cited several other cases similar to our case. Therefore, in 1991, Pearl reported a similar case with a single arterial trunk arising from the aorta and aortic coarctation [7]. Similar cases have been cited also by McDowell *et al.* in 1980 and Roguin *et al.* in 1982 [8, 9]; however, this type of anomaly remains a very rare one.

Materials, Methods and Results

This is the case of a male infant, born at term, without medical surveillance, in parous mother, alcohol drinker, coming from a broken home but without other associated pathologies. The infant died five days after birth due to severe malformations incompatible with life, respectively the single arterial trunk was arising from the aorta and aorta coarctation. There were no intrapartum infections reported, trauma or drug consumption during embryogenesis.

At birth, the child presented bilateral vesicular sounds, without rates, AV=120 beats/min, rhythmic, no pulmonary sounds. At birth, the child presented circular cord; therefore, a cesarean birth was performed. On the fourth day after birth, his general condition worsened declaring the death after resuscitation. At necropsy, there were found that the wall of the aorta were normal, the inner membrane had an yellowish color, it was smooth, with a single arterial trunk that presented a bifurcation with a significant narrowing of the aortic lumen at approximately 0.8 cm from its base (Figure 1); of this bifurcation, two brachiocephalic trunks branched and each one divided into a subclavial artery and a common carotid artery (Figure 2).

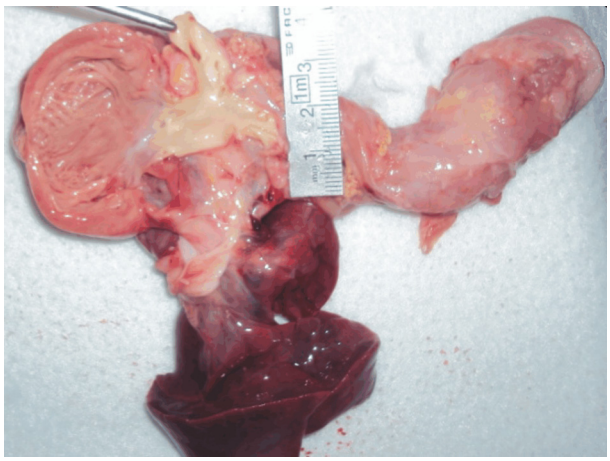

Figure 1 – ***Image of the aorta at necropsy.***

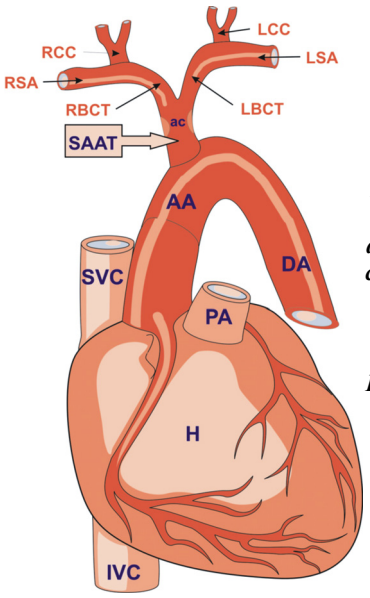

Figure 2 – ***H: Heart; IVC: Inferior vena cava; SVC: Superior vena cava; PA: Pulmonary artery; AA: Aortic arch; DA: Descending aorta; SAAT: Single aortic arterial trunk; ac: Aortic coarctation; LBCT: Left brachiocephalic trunk; RBCT: Right brachio-cephalic trunk; RSA: Right subclavian artery; LSA: Left subclavian artery; RCC: Right common carotid artery; LCC: Left common carotid artery (original drawing).***

Conclusion of the anatomic pathology department was that death intervened because of the acute cardio-respiratory failure due to severe malformations of the cardiovascular system.

Discussion

A wide spectrum of congenital anomalies may occur during branching of the aortic artery, brachiocephalic arteries and ductus arteriosus. Most of them are asymptomatic, discovered only by chance and very few showing significant cardiovascular or respiratory symptoms requiring treatment. To understand these malformations is imperative to know embryogenesis of the aorta (Figure 3).

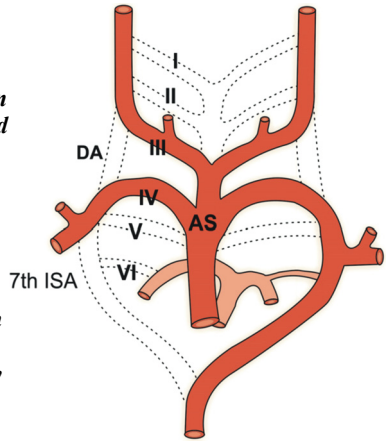

Figure 3 – ***Drawing illustrating the six pairs of aortic arches (I–VI) formed between the aortic sac (AS) and paired dorsal aortae (DA). Parts of the dorsal aortae and aortic arches that normally have disappeared at seven weeks are shown with broken lines. ISA: Intersegmental artery (adapted from Kellenberger [10]).***

From this point of view, most of aortic arch malformations can be explained by assuming the double arch system described by Edwards [11]. According to this theory, aortic malformations come from the failure of regression, or regression on an abnormal site of the fourth aortic arch. Probable regression sites of left and right arches are:

Left aortic arch

Left aortic arch is a cardiovascular anomaly occurring with a rate of 1.2%, caused by regression of the fourth left aortic arch between the right carotid artery and right subclavian artery, resulting in the formation of a left aortic arch with abnormal origin of the right subclavian artery as the last branch [12]. Such an abnormal right subclavian artery (lusoria artery) descends again behind the esophagus and makes a symptomatic incomplete vascular ring. Depending on the regression of the distal part of dorsal aorta root from the ductus arteriosus, two situations may be encountered: ductus arteriosus is open, the blood supply is made by the right pulmonary artery, and if it is off, subclavian artery is supplied by collateral vessels, particularly by the vertebral artery. In the presence of an arterial duct on the right side, between the right pulmonary artery and right subclavian artery, the abnormal subclavian artery, may have originated from a retro-esophageal aortic diverticulum (Kommerell diverticulum), which form together with the duct ligament from the right side, a complete vascular ring [13]. If the descending aorta is found with the left aortic arch, the distal portion of the arch passes behind the esophagus. This is named with the term of "circumflex aorta". Depending on the distal or proximal regression of right subclavian artery, brachiocephalic arteries may branch normally or may branch on the right subclavian artery [10, 14].

Right aortic arch

Most often, the right aortic arch is associated with cardiovascular malformations as persistence of the ductus arteriosus, Fallot tetralogy, pulmonary atresia or ventricular septum defects resulting by regression of the fourth distal left aortic arch by the right subclavian artery. In this case, brachiocephalic vessels will originate – mirror image – from left aortic arch [15]. The first arterial branch passes to the left of the left innominated artery, producing the left common carotid artery and left subclavian artery, followed by right common carotid artery and right subclavian artery [16, 17]. If the fourth left aortic arch regresses distal to the left subclavian artery, an abnormal right aortic arch will result with left subclavian artery as the last branch, aberrant subclavian artery may originate in this case from an aortic diverticulum, and passing behind the esophagus, it may form with the ligament duct on the left side one complete vascular ring. The blood supply for subclavial artery can be made by left arterial duct, if the duct is open or by vascular collateral especially by vertebral arteries, if the duct is closed. Persistence of the left descending aorta with a right aortic arch leads to the formation of a circumflex arterial arch. In the presence of an isolated left subclavian artery, the left hand pulse can be reduced [18, 19]. Presence of a retroesophageal right aortic arch where the aorta passes over the main right bronchus, goes to the left and behind the esophagus, and then down the left side of it, is a rare case. It can compress the trachea and esophagus and can cause severe symptoms [20]. There have been reported cases of right aortic arch with isolation of the left common carotid artery. This malformation was explained by the hypothesis of Edwards, with an interruption of the aortic arch distal to the left common carotid artery, between the ductus arteriosus and left subclavian arteries. This situation would imply a lack of migration proximal to subclavian artery so that the ductus will emerge into the left dorsal aortic root in the proximity of aberrant left subclavian artery. Usually, this malformation is associated

with other cardiac malformations such as atrial septal defect [21] or tetralogy of Fallot [22].

Persistent left fifth aortic arch

Persistent left fifth aortic arch is an anomaly represented by a double-lumen aortic arch [23]. Fifth aortic arch is under the real aortic arch and stretches from the opposite innominated subclavian artery up to the subclavian artery with branches at both ends which communicates with the aortic lumen. Herrera *et al.* [24] and Gerlis *et al.* [25] presented cases in which there is a communication between the pulmonary and systemic circulation. Such a case was also described by Freedom *et al.* [26]: associated with tricuspid and pulmonary atresia and coarctation of the aorta. In these cases, fifth aortic arch lies between the ascending aorta and derivatives of the sixth aortic arch. Fifth aortic arch persistence is often associated with complex cardiac malformations.

Subclavian artery as the first branch of the aortic arch

Subclavian artery as the first branch of the aortic arch is an extremely rare vascular anomaly, in which the first branch that emerges from the ascending aorta is the subclavian artery on the collateral side of the arch. Common carotid arteries can branch sequentially from the arc or may have originated from a common trunk. The arch can be either on the right or on the left. The anomaly can be explained by the persistence of the fifth branchial arch associated with regression of the fourth ipsilateral branchial arch and dorsal aorta segment of the arches III and IV. Once the seventh intersegmental cephalic artery migrated, the next subclavian artery will emerge from the fifth aortic arch and not from the fourth aortic arch [27, 28].

Double aortic arch

Double aortic arch occurs by persistence of both IV aortic arches; this double arch surrounds the trachea and esophagus. Each aortic arch descends along the bronchus roots and rejoins forming the descending aorta. With a left descending aorta, the right arch will pass behind the esophagus and left arch in front of the trachea. The size of the aortic arches is usually asymmetric; right aortic arch is usually bigger than the left one and positioned higher than this [29]. A part of the left aortic arch can present atresia and persist as a fibrous band [30]. Common carotid and subclavian arteries may come separately from the respective aortic arches. A ductus arteriosus may be present on both sides only on one side. Such malformations are not usually associated with cardiac malformations and form a tight ring indicating stridor, dyspnea and recurrent respiratory infections [31, 32]. But, there have been reports that showed association of this anomaly with other cardiac abnormalities such as mitral stenosis or persistent ductus arteriosus [33] or atresia of proximal portion of the right common carotid artery [34]. Surgical separation of the smaller or atresic aortic arch and of the ductus ligament is often required [36].

Interrupted aortic arch

Interrupted aortic arch occurs in 1.4% of cases with cardiovascular malformations and results when both fourth aortic arches regress so that the ascending aorta is not in continuity with the descending aorta and descending aorta is supplied by the ductus arteriosus or collateral arteries. Regression site may vary for each aortic arch resulting in different anatomical patterns. Celoria and Patton [37] have classified this anomaly in three types, depending on the site of interruption: type A (from the isthmus artery beyond the subclavian aortic), type B (between left common carotid and left subclavian artery) and type C (between the two common carotid arteries). In each type, the right subclavian artery may originate normally from the innominated artery and can have an aberrant origin as the last aortic branch or can be isolated or connected by the pulmonary artery through a ductus arteriosus.

Type B is the most common form, followed by types C and A. This anomaly is often associated with cardiac abnormalities such as patent ductus arteriosus, transposition of big vessels and aortopulmonary fistulas. A large majority of these patients have DiGeorge syndrome (deletion of chromosome 22q11.2) [38, 39].

A newborn baby with interrupted aortic arch after birth will present congestive heart failure and respiratory distress when the ductus arteriosus closes. Currently, surgical correction is attempted during the first weeks of life [40].

Isolated brachiocephalic arteries

Isolated brachiocephalic arteries occurs by regression of a single IVth aortic arch in two portions resulting isolation of one or more brachiocephalic arteries in the opposite part of the aortic arch in a manner they will not follow the aorta but link to the ipsilateral pulmonary artery through the ductus arteriosus [41]. Isolated arteries are supplied by vertebral artery or arteries on the neck and thorax side. If the ductus arteriosus is open and pulmonary arterial pressure is normal, there is a left to right shunt in the pulmonary artery. Clinically, it may be a vertebrobasilar insufficiency or inadequate vascular supply of the ipsilateral arm with a low pulse. Isolation of the left subclavian artery is more common than isolation of the right subclavian artery and is often associated with tetralogy of Fallot. Rare cases have been reported on both isolation of innominated left artery and left common carotid artery.

Cervical aortic arch

Cervical aortic arch is a rare malformation, given by the extension of the aortic arch top above the sternum [42, 43]. It is believed that this anomaly is the result of persistent III aortic arch and regression of IIIrd and IVth aortic arches. Ipsilateral internal and external carotid arteries may originate directly from the aortic arch. Descending aorta often remains on the contralateral side with a retroesophagian segment arch and an aberrant subclavian artery, arising from an aortic diverticulum. In this case, cervical arches form a vascular ring with a contralateral ligament duct. It was reported a case of right cervical aortic arch associated with brachiocephalic left subaortic vein, whose mechanism could not be explained [44, 45].

Double lumen aortic arch

Double lumen aortic arch occurs by persistence of Vth aortic arch with the IVth ipsilateral aortic arch, resulting brachiocephalic arteries [46]. Persistence of Vth aortic arch spans below the real aortic arch connecting the ascending aorta opposite to the innominated artery with aortic arch on the distal side opposite to the subclavian artery.

Conclusions

Abnormalities of the aortic arch occur mainly due to an abnormal development of the fourth left aortic arch in the first 12 weeks of fetal life and less to the fourth right aortic arch. This case reports a very rare malformation of the aortic arch, and similar cases are also very rare even in literature. We believe that the abnormality is due to a regression on an abnormal site of the fourth left aortic arch, but due to the very small number of these anomalies reported so far, we cannot say with certainty if there is an embryological reason for these abnormalities. This anomaly could not be associated with some determining factors or a genetic predisposition, the cause for this anomaly remains uncertain.

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