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CASE REPORT

ANALOGY IN THE INTERRUPTION OF AORTIC BLOOD FLOW: COARCTATION OF THE AORTA OR INTERRUPTION OF THE AORTIC ARCH

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Abstract

Introduction. Interruption of the aortic arch is commonly associated with genetic conditions such as Turner and DiGeorge syndromes. This cardiovascular abnormality is associated with significant hemodynamic imbalances, leading to increased vascular resistance and progressive structural complications, finally developing into severe heart failure with an elevated risk of mortality. The most common presentation resembles aortic coarctation; other presentations include aortic arch hypoplasia or interruption, which are more complex congenital cardiovascular diseases.
Case presentation. The case of an 18-year-old male patient with no significant medical history is described. The patient came to the internal medicine department with chest pain, without radiation, lasting approximately 15 minutes, making it impossible for him to walk and gradually decreasing without self-medication. The next day he presented a new episode of chest pain of greater intensity accompanied

Résumé

Analogie dans l'interruption du flux sanguin aortique: coarctation de l'aorte ou interruption de la voie aortique

Introduction. L'interruption de l'arc aortique est généralement associée à des maladies génétiques telles que les syndromes de Turner et de DiGeorge. Cette anomalie cardiovasculaire est associée à des déséquilibres hémodynamiques importants, entraînant une augmentation de la résistance vasculaire et des complications structurelles progressives, évoluant finalement vers une insuffisance cardiaque sévère avec un risque élevé de mortalité. La présentation la plus courante ressemble à une coarctation aortique, cependant, d'autres présentations incluent une hypoplasie ou une interruption de l'arc aortique, qui sont des maladies cardiovasculaires congénitales plus complexes.
Rapport du cas. Le cas d'un patient de sexe masculin de 18 ans, sans antécédents médicaux significatifs, est

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by pulsating, holocranial headache, as well as palpitations. The cardiology evaluation identified dyspnea on mild effort, pulses present but decreased in the pelvic limbs, blood pressure in the thoracic limbs 143/87 mmHg, pelvic limbs 122/83 mmHg, holosystolic crescendo-decrescendo murmur in the aortic area, with interscapular irradiation.

Conclusions. Usually more common in men, interruption of the aortic arch causes retrograde heart effects and represents a severe form of aortic coarctation with left ventricular involvement. This ultimately leads to multisystem overload and heart dilatation.

Keywords: interrupted aortic arch, coarctation of the aorta, heart failure.

List of abbreviations:
NYHA II – New York Heart Association class II

décrit. Le patient s'est présenté au service de médecine interne avec une douleur thoracique sans radiation, d'une durée d'environ 15 minutes, l'empêchant de marcher et diminuant progressivement sans automédication. Le lendemain, il a présenté un nouvel épisode avec douleur thoracique d'intensité plus élevée accompagnée de céphalées holocrâniennes pulsatiles, ainsi que de palpitations. L'évaluation cardiologique a identifié une dyspnée aux petits efforts; les pouls sont présents avec diminution dans les membres pelviens, la pression artérielle dans les membres thoraciques 143/87 mmHg, membres pelviens 122/83 mmHg, un souffle holosystolique crescendo-decrescendo dans le foyer aortique avec irradiation interscapulaire.

Conclusions. Généralement plus fréquente chez les hommes, l'interruption de l'arc aortique provoque des effets cardiaques rétrogrades et fonctionne comme une forme grave de coarctation aortique avec atteinte du ventricule gauche. Cela conduit finalement à une surcharge multisystémique et à une dilatation cardiaque.

Mots-clés: arc aortique interrompu, coarctation de l'aorte, insuffisance cardiaque.

Introduction

Interruption of the aortic arch is a congenital condition that affects approximately 1.5% of patients with congenital heart disease[1]. Coarctation of the aorta is common in patients with Turner syndrome, and up to 68% of patients with DiGeorge syndrome have type A interruption of the aortic arch, between the ascending and descending aorta[1]. This is considered the most severe form of coarctation, and mostly occurs with the closure of the ductus arteriosus during the first two weeks of life. Generally, blood flow typically depends on communicating ducts to ensure distribution to areas distal to the interruption[2,3]. Approximately 73% of cases of interruption of the aortic arch are associated with interventricular communication resulting from a poor posterior alignment of the conal septum[4]. This is considered a branchial arch defect, a definition that includes interruption of the aortic arch, coarctation of the aorta and right aortic arch, and potential left ventricular outflow obstruction. In addition to ventricle defects, more complex anomalies may occur, such as right ventricle double outlets, transposition of the great vessels, single ventricles, and aortic valve atresia[5,6]. Classification is primarily based on the site of interruption. For example, type A, with interruption distal to the left subclavian artery, represents 13% of cases; type B, with interruption between the left carotid and left subclavian arteries, represents 84% of cases; and type C, where the interruption is between the innominate artery and the left carotid artery, occurs in 3% of cases[7]. Subclassifications are determined according to the origin of the subclavian artery. Type 1 defines a subclavian artery of normal origin; type 2 indicates an aberrant right subclavian, distal to the left; and type 3 indicates an isolated right subclavian artery with patent right ductus arteriosus. Aortic arch interruptions B and C occur from anomalies in the formation of the fourth aortic arch. However, regardless of the presentation or classification, intervention is necessary; the lack of treatment leads to a variety of complications, including stroke, heart failure, infective endocarditis, and aortic dissection. Within the surgical approach, complications may occur after repair, such as the formation of pannus and/or stenosis of the surgical site, which requires intervention by cardiac catheterization[6,8].

Case presentation

An 18-year-old male patient, with no relevant medical history, presented with a sudden onset of intense precordial pain without radiation, which made it impossible to walk. The pain lasted approximately 15 min before gradually subsiding without self-medication. The next day, he presented with a new episode of precordial pain of greater intensity with a pulsating, holocranial headache and palpitations. The cardiological evaluation identified a murmur without a specified focus and decreased pulses in the pelvic

limbs. A chest angiography was requested, which revealed collateral circulation involving the para-intervertebral, internal mammary, and bilateral axillary arteries, with clear evidence of post-ductal coarctation in the proximal descending aorta (Figure 1). The utilization of radiopaque iodine confirmed a probable post-ductal aortic coarctation. Treatment was started with telmisartan (40 mg every 12 h). However, the persistence of elevated arterial pressure suggested resistance to pharmacological therapy.

An echocardiogram was performed, showing critical post-ductal aortic coarctation. A bicuspid aortic valve with mild insufficiency and mild aortic root dilation was also observed. The left ventricular ejection fraction was 60%, with normal segmental kinetics at rest. The pulmonary artery systolic pressure was 17 mmHg, with no associated shunt.

At the next evaluation, a transthoracic echocardiogram revealed a bicuspid aortic valve and mild regurgitation. Juxtaductal aortic coarctation with minimal flow was observed, and the patient remained hemodynamically stable. On physical examination, cardiac auscultation revealed rhythmic heart sounds with a holosystolic murmur predominantly heard at the accessory aortic area (Levine 3). A pulmonary murmur (Levine 2) was noted, with a rough, protosystolic timbre, without the presence of a thrill. A mesosystolic interscapular murmur with radiation and increased tone and intensity was detected (Levine 3). Upon questioning, the patient reported dyspnea on mild effort (NYHA II); the pulses were present but decreased in the pelvic limbs. The blood pressure in the thoracic limbs was 143/87 mmHg, and 122/83 mmHg in the pelvic limbs. The electrocardiogram showed sinus rhythm with a heart rate of 71 bpm and an axis of –30°; the PR interval was 0.16 seconds, with a QTc of 360 ms; left ventricular hypertrophy and incomplete left bundle branch block, with no evidence of ischemia or necrosis. The suspicion was of probable postductal aortic coarctation (Bakey type III A).

The cardiothoracic surgery service assessed the patient. Following consultation, the surgical management was initiated, during which the diagnosis of aortic arch interruption was confirmed. A #16 Dacron graft was placed in the thoracic aorta, and coartectomy was performed without complications, leading to a final diagnosis of aortic arch interruption. No aortic murmur was detected on physical examination after surgery. The patient was discharged with no symptoms.



Figure 1. CT angiography using 3D reconstruction reveals that the descending aorta shows a segment of aortic arch interruption, with mild dilation distal to the interrupted segment.

Discussion

Interruption of the aortic arch is considered a defect of the branchial arch. It is the result of congenital heart disease, with a low incidence of approximately 1.5%[2]. The risk of mortality is the highest in the first years of life; if a patient reaches adolescence without intervention, it is typically due to dependence on a collateral duct allowing distributed blood flow to distal tissues, bypassing the interruption[3,5]. This dependence can lead to later vascular abnormalities at the origin site of the duct. The embryological origin of congenital heart disease can lead to both ventricular defects, such as left ventricular outlet obstruction, right ventricle double outlets, or a single ventricle, as well as valve defects, such as aortic valvular atresia[3,9]. The classification currently considers three parameters of the segment presentation (transverse arch, juxta ductal, and isthmic), the absence or presence of lesions (bicuspid aortic valve, septal defects, subaortic stenosis, left ventricular hypoplasia, and mitral valve anomaly), and age (adult, infantile, or neonatal)[5,6]. Our patient was diagnosed with double congenital heart disease of the juxtaductal aortic arch interruption type, with the presence of a bicuspid aortic valve and an adult age at presentation[9,10]. The physical examination identifies weak or absent pulses, and the blood pressure in the lower extremities is usually significantly lower than in the upper extremities[3,8]. Chest X-ray can show findings

suggestive of aortic coarctation; electrocardiogram shows an enlargement of cavities. Echocardiography is the test of choice for the diagnosis of this pathology, as the length of the interrupted space and the branching pattern can be confirmed[11].

Conclusions

Interruption of the aortic arch tends to have a higher incidence in men and behaves as a severe form of aortic coarctation with left ventricular involvement, leading to retrograde effects on the heart. This eventually results in heart dilatation and subsequent multisystem overload. The typical survival is 34 years of age. The main causes of mortality are bacterial endocarditis, intracranial hemorrhage, cardiogenic shock, and aortic rupture. This pathology has a low incidence and is poorly represented in the literature, with only a few multicenter studies. The optimal surgical management has not been fully established, as the age of presentation, affected segment, and concomitance with other congenital heart defects must be considered, including the pressure gradient. This pressure varies in patients with aberration and collateral vessels, complicating the surgical options.

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Compliance with Ethics Requirements:

"The authors declare no conflict of interest regarding this article"

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