Anomalous Origin of the Right Coronary Artery with an Interarterial Course: Incidental Finding in a Pediatric Patient with Suspected Kawasaki Disease: Case Report

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ABSTRACT

Congenital anomalies of the coronary arteries are uncommon and mostly involve abnormalities in the origin and course of the coronary vessels. We present the case of an 8-year-old pediatric patient with prolonged febrile syndrome and mucocutaneous manifestations, in whom incomplete Kawasaki disease was clinically suspected. During the diagnostic workup and imaging evaluation, an anomalous origin of the right coronary artery from the left coronary sinus, with an interarterial course and significant proximal narrowing, was incidentally identified. The imaging findings, their anatomical relevance, and the associated clinical risks are described, highlighting the educational value of coronary CT angiography in accurately characterizing coronary anatomy and differentiating congenital anomalies from potential acquired dilatations in the context of systemic inflammatory disease.

Keywords: Coronary arteries, congenital anomalies, computed tomography, Kawasaki disease, sudden death.

Origen anómalo de la arteria coronaria derecha con trayecto interarterial: hallazgo incidental en un paciente pediátrico con sospecha de enfermedad de Kawasaki

RESUMEN

Las anomalías congénitas de las arterias coronarias son poco frecuentes y corresponden en su mayoría a alteraciones en el origen y trayecto de los vasos coronarios. Se presenta el caso de un paciente pediátrico de 8 años con síndrome febril prolongado y manifestaciones mucocutáneas, en quien se planteó la sospecha clínica de enfermedad de Kawasaki incompleta. Durante el proceso diagnóstico y la evaluación por imágenes se identificó, como hallazgo incidental, un origen anómalo de la arteria coronaria derecha a partir del seno coronario izquierdo, con trayecto interarterial y reducción significativa de su calibre proximal. Se describen los hallazgos imagenológicos, su relevancia anatómica y los riesgos clínicos asociados, destacando el valor educativo de la angiotomografía coronaria en la correcta caracterización de la anatomía coronaria y en la diferenciación entre anomalías congénitas y posibles dilataciones adquiridas en el contexto de enfermedad inflamatoria sistémica.

Palabras clave: arterias coronarias, anomalías congénitas, tomografía computarizada, enfermedad de Kawasaki, muerte súbita.

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INTRODUCTION

Kawasaki disease is an acute systemic vasculitis that predominantly affects young children and may involve the coronary arteries, leading to dilatation or aneurysm formation with a potential risk of thrombosis and myocardial ischemia[1,2]. In parallel, congenital anomalies of the coronary arteries are uncommon entities, with an estimated incidence of 0.5% to 1% in the general population, and may remain asymptomatic or present with angina, ventricular arrhythmias, or sudden death, particularly in young patients[3,4].

Among these anomalies, the anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course is considered high risk due to the potential for dynamic compression of the vessel between the aorta and the pulmonary artery during systole, which may lead to myocardial ischemia and severe arrhythmic events[4-6]. Accurate anatomic characterization of these variants is crucial for determining their clinical significance and guiding therapeutic management[4-6].

Available imaging modalities include echocardiography, coronary computed tomography angiography, cardiac magnetic resonance imaging, and coronary arteriography. Multidetector coronary computed tomography angiography provides high-resolution anatomical detail of the coronary origin, course, and extracoronary relationships, and is particularly useful when anatomical uncertainty remains after echocardiography or when a comprehensive evaluation of the coronary tree is required[6,7].

CASE DESCRIPTION

An 8-year-old male patient with no relevant past medical history presented to a primary care center with an 8-day history of persistent fever, malaise, dysphagia, abdominal pain, intolerance to oral intake, conjunctival hyperemia, strawberry tongue, and a rash involving the neck and chest. Given the persistence of the febrile syndrome and the presence of mucocutaneous manifestations, incomplete Kawasaki disease was clinically suspected, and the patient was referred to a higher-complexity center for specialized evaluation.

Upon admission to the referral center, laboratory studies showed elevated acute-phase reactants, without significant hematologic abnormalities. The differential diagnoses considered included scarlet fever, streptococcal pharyngitis, adenovirus infection, and other causes of prolonged febrile syndrome in pediatric patients. The patient had not received prior antibiotic therapy.

Treatment was initiated with intravenous immunoglobulin at the standard dose of 2 g/kg administered as a single infusion, in combination with acetylsalicylic acid. Subsequently, the patient showed clinical improvement with progressive resolution of fever, and hemodynamic monitoring was continued in an intermediate care unit.

Transthoracic echocardiography did not reveal structural heart disease or abnormalities of systolic or diastolic function. Coronary measurements reported a right coronary artery of 1.9 mm (Z-score: −0.7) and a left coronary artery of 2.9 mm (Z-score: +2.1). Due to anatomical uncertainty regarding the origin and proximal course of the right coronary artery on echocardiography, coronary computed tomography angiography was requested for better morphological characterization.

Coronary computed tomography angiography demonstrated an anomalous origin of the right coronary artery arising from the left coronary sinus, with an interarterial course between the aorta and the pulmonary artery trunk, associated with an approximately 50% reduction in luminal caliber at its proximal segment (Fig. 1). In addition, mild diffuse dilatation of the left main coronary artery and the proximal segment of the circumflex artery was observed, without evidence of aneurysms or stenosis in the mid and distal segments (Figs. 2 and 3).

The case was discussed in a multidisciplinary meeting involving Clinical Cardiology, Cardiovascular Imaging, and Hemodynamics. The team concluded that additional dynamic evaluation was required, either by diagnostic cardiac catheterization or by cardiac magnetic resonance imaging with three-dimensional reconstruction, in order to confirm the degree of extrinsic compression and assess its impact on myocardial perfusion in the territory

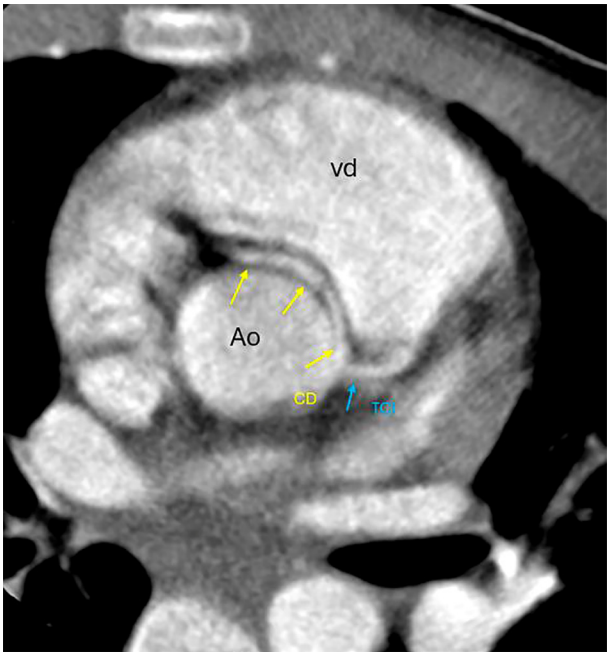

Figure 1. Coronary computed tomography angiography with an axial slice at the level of the great vessels, showing the right coronary artery (RCA) originating from the left coronary sinus, with an interarterial course that reduces its diameter by more than 50% compared with the following segment. No areas of stenosis or dilatation are observed in the mid, distal, or posterior descending artery (PDA) segments. RV: right ventricle; Ao: aorta; RCA: right coronary artery; LMCA: left main coronary artery; PDA: posterior descending artery.

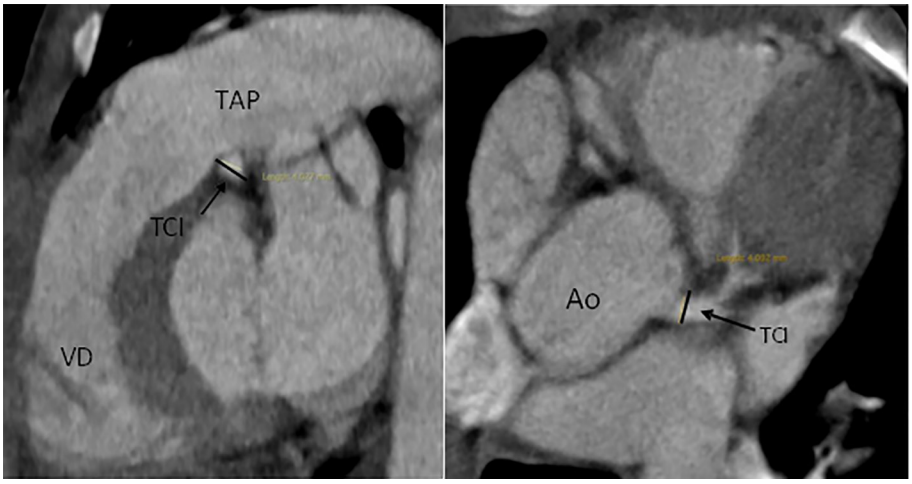

Figure 2. Axial measurement of the left main coronary artery (LMCA) showing a normal origin and course. Diffuse dilatation of the proximal and distal segments is observed. No areas of stenosis are identified. PT: pulmonary trunk

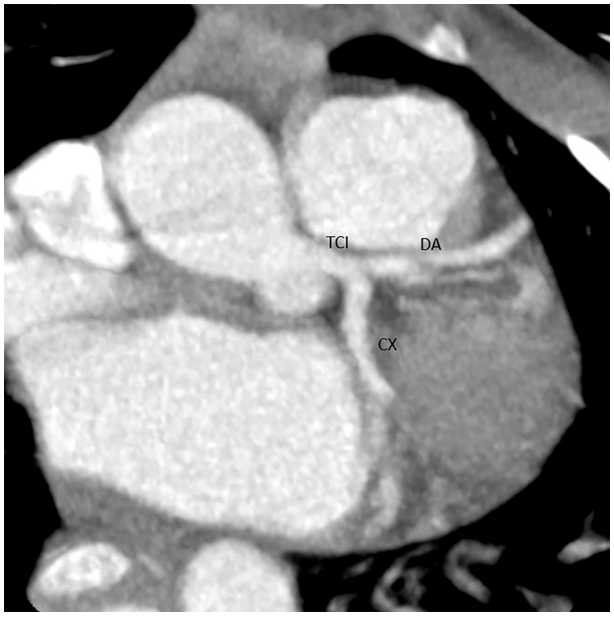

Figure 3. The circumflex artery shows a normal origin and course. The proximal segment presents mild diffuse dilatation. The first obtuse marginal branch shows no lesions, and the distal segment shows no lesions. The left anterior descending artery (LAD) shows a normal origin and course, with no evidence of atherosclerotic disease, stenosis, or dilatation in its three segments. The first and second diagonal branches show no lesions, and the first septal branch shows no lesions. CX: circumflex artery; LAD: left anterior descending artery.

supplied by the right coronary artery. The patient was referred to a specialized cardiovascular center for follow-up and potential therapeutic decision-making.

After referral to the higher-complexity cardiovascular center, the patient was evaluated by the Pediatric Cardiology service. The anomalous origin of the right coronary artery with an interarterial course was confirmed as a relevant anatomical finding, without clinical evidence of myocardial ischemia or acute hemodynamic compromise at the time of evaluation.

Given the patient's clinical stability and the absence of cardiovascular symptoms, the multidisciplinary team opted for an expectant management strategy, with close outpatient cardiology follow-up and recommendations to restrict intense physical activity until completion of the longitudinal evaluation. Serial echocardiographic follow-up was scheduled to monitor proximal coronary anatomy and ventricular function, along with periodic clinical reassessment to determine the need for additional diagnostic studies or therapeutic interventions.

DISCUSSION

The identification of congenital anomalies of the origin and course of the coronary arteries represents a diagnostic challenge, particularly in the context of systemic inflammatory diseases such as Kawasaki disease, in which acquired coronary dilatations may mimic or mask congenital anatomical variants[7,8]. Clinical series have noted that some congenital anomalies may be misinterpreted as coronary "dilatations" during echocardiographic evaluation of patients with suspected Kawasaki disease, highlighting the need for appropriate anatomical correlation when atypical findings are present[8].

The anomalous origin of the right coronary artery from the left coronary sinus with an interarterial course is considered a high-risk variant due to the potential for dynamic compression during systole, which may lead to myocardial ischemia and predispose young patients to malignant arrhythmias or sudden cardiac death[4–6].

In the present case, coronary computed tomography angiography enabled precise anatomical characterization of the coronary origin and course, as well as its relationships with the great vessels, providing critical information for

risk stratification and diagnostic/therapeutic planning[6,7]. From an educational perspective, this finding highlights the importance of including congenital variants in the differential diagnosis when inconclusive coronary findings are encountered in pediatric patients, and of reserving coronary computed tomography for scenarios involving anatomical uncertainty or the need for detailed mapping of the coronary tree[6,7].

Dynamic evaluation with cardiac magnetic resonance imaging or catheterization remains essential to determine the indication for surgical intervention in patients with evidence of significant compression and/or hemodynamic impact[4-6].

CONCLUSION

The present case highlights the diagnostic and imaging value of coronary computed tomography angiography in the incidental identification of a congenital anomaly of the origin of the right coronary artery with an interarterial course in a pediatric patient with clinical suspicion of incomplete Kawasaki disease. Accurate anatomical characterization of these variants is essential to establish their clinical relevance, guide therapeutic management, and prevent potential major complications, including myocardial ischemia and sudden death[4-6].

Conflicts of interest: The authors declare no conflicts of interest related to the content of this study.

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