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Fatal late cardiovascular sequelae of previously unrecognized Kawasaki disease in 12-year-old child

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Abstract

Background Kawasaki disease (KD), previously termed mucocutaneous lymph node syndrome, is a childhood vasculitis affecting medium-sized arteries and is the leading cause of acquired heart disease in children. It primarily affects children under five years of age. If left untreated, KD can lead to serious cardiovascular complications, particularly coronary artery aneurysms (CAA) and thrombosis. Incomplete KD presents with fewer clinical criteria, making it more difficult to diagnose. Importantly, long-term sequelae such as CAA may remain clinically silent for years. This case highlights the critical need for awareness that even minimal or transient symptoms can be the only warning sign of life-threatening complications in adolescents with a remote history of incomplete or unrecognized KD.

Case presentation We describe a fatal case of a 12-year-old boy with a history of presumed myocarditis at age five, which retrospectively fulfilled criteria for incomplete KD but remained undiagnosed. From age five to twelve, he was asymptomatic except for occasional, brief chest tightness. At twelve, he presented with mild chest pain followed by rapid clinical deterioration, cardiac arrest, and death. Post-mortem imaging and autopsy revealed a thrombosed giant aneurysm of the left anterior descending coronary artery, consistent with chronic coronary disease.

Conclusion This case illustrates the potentially fatal long-term cardiovascular sequelae of unrecognized and untreated incomplete KD. Early recognition and treatment with IVIG are critical to reduce coronary complications. Healthcare providers must maintain clinical vigilance for patients with a history of KD. Even subtle or transient symptoms in patients with a history of KD should prompt immediate evaluation to prevent fatal outcomes.

Keywords Kawasaki disease, Incomplete Kawasaki disease, Coronary aneurysm, Myocardial infarction, Pediatric cardiac arrest, Case report

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Introduction/background

Kawasaki disease (KD), formerly called mucocutaneous lymph node syndrome, is one of the most common vasculitides of childhood. The syndrome is characterized by vasculitis of the medium-sized arteries. The acute illness is self-limited and includes high fever, non-exudative conjunctivitis, inflammation of the oral mucosa, rash, cervical adenopathy and findings in the extremities (edema of hands and feet) [1]. Kawasaki disease mainly affects infants and children under five years of age. The incidence is higher in Japan (>200/100,000) than in Western countries (5–22/100,000) and males are more commonly affected (1.5:1) [2].

The cause of KD is unknown, although it is suspected to be triggered by an unidentified infectious pathogen in genetically predisposed children. KD might not be a normal immune response to an unusual environmental stimulus, but rather a genetically determined unusual and uncontrolled immune response to a common stimulus [2, 3]. KD is the leading cause of acquired heart disease among children and unrecognized and untreated children are at risk for serious cardiovascular sequelae, particularly abnormalities of the coronary arteries (CA). The article presents a fatal case of late cardiovascular complication of previously unrecognized KD in a 12-year-old boy. He remained virtually asymptomatic from the age of five, when he experienced myocarditis of unknown origin, most likely representing an unrecognized case of incomplete Kawasaki disease.

Case presentation

We present the case report of a 12-year-old Ukrainian boy with a history of myocarditis of unknown etiology at the age of 5.

The patient was admitted at the age of 5 to a local pediatric infectious disease department for 1 week of fever, cervical lymphadenopathy and pharyngitis. At the time of admission, he was on oral amoxicillin/clavulanic acid for 5 days. On admission, his labs showed leukocytosis 20, ESR 26, CRP 20 mg/L, ALT 89 U/L (1,48 ukat/L), AST 39 U/L (0,65 ukat/L). After the admission, the serology for viruses was negative (EBV, CMV, adenovirus, HHV6). Throat swab was positive for Streptococcus pneumoniae. The antibiotic was changed do 3rd- generation cephalosporin (ceftazidime). For persisting fever and epigastric pain, CXR and abdominal CT scan were performed, both without pathological findings. Due to an increase of Troponin-I 114 ng/mL (normal range 0–0,04 ng/mL) and CK-MB 187 U/L (normal range 5–25 U/L) and suspicion of acute myocarditis, he was transferred to the pediatric cardiology department. The initial echocardiogram, performed by a pediatric cardiologist, showed mildly decreased left ventricular function (EF 52%) without structural defects. The echo report did not mention examination of coronary arteries. One dose of intravenous immunoglobulin (IVIG) was given (1 g/kg) and methylprednisolone (4 mg/day) was started, tapered off within 2 weeks. Follow-up echocardiogram showed improvement in left ventricular function (EF 65%). The values of Troponin-I 0,326ng/L a CK-MB 22U/L normalized. He was dismissed with no medication and the condition was concluded as myocarditis of unclear etiology. He was followed by a cardiologist, but several echocardiograms showing normal cardiac function were performed by a radiologist; assessment of the coronary arteries was not mentioned in any of the reports. The follow-up was completed at the age of 8 years. In the next period, the patient was in good condition. He actively played football with a good tolerance for physical activity. He occasionally described feeling a brief pressure on the chest that spontaneously subsided within minutes. Since discharge from cardiology follow-up, he had not been monitored by a cardiologist.

The patient was recently admitted at the age of 12 years to our pediatric department with a short episode (several hours) of mild chest pain localized in the lower left sternal border with no recent history of acute respiratory tract infection or fever. On admission, there were no clinical signs of respiratory or circulatory compromise. Changes in repolarization were evident on the ECG (Fig. 1), with only a mild elevation of high sensitive troponin T = 51 ng/L (< 14 ng/L), NT pro BNP 110 ng/L (< 14 ng/L) and normal levels of CK of 1.76 μkat/L (1.1–4.8 μkat/L). An interstitial pattern was visualized on the X-ray of the lungs. The chest pain resolved spontaneously within admission to the ward.

Shortly after admission, the patient developed progressive tachypnea and increased work of breathing. Our differential diagnoses included pulmonary embolism, myocarditis, primary pulmonary pathology, aortic dissection, acute coronary syndrome and multisystem inflammatory syndrome in children (MIS-C). CT angiography was indicated.

A brief improvement in his clinical status was followed by a sudden deterioration, circulatory and respiratory collapse, and the need for intubation with rapid progression to cardiac arrest. The initial rhythm was pulseless ventricular tachycardia. After the first shock, pulseless electrical activity refractory to treatment and ultimately resulted in death. At the time of ongoing resuscitation, the result of CT angiography was not available. Due to the unavailability of extracorporeal resuscitation in our institution, resuscitation was terminated after 40 min. Nasopharyngeal and tracheal Covid-19 Multiplex RT-PCR Kit (DIANA Biotechnologies, Czech Republic) was negative, as patient had no recent signs of respiratory infection.

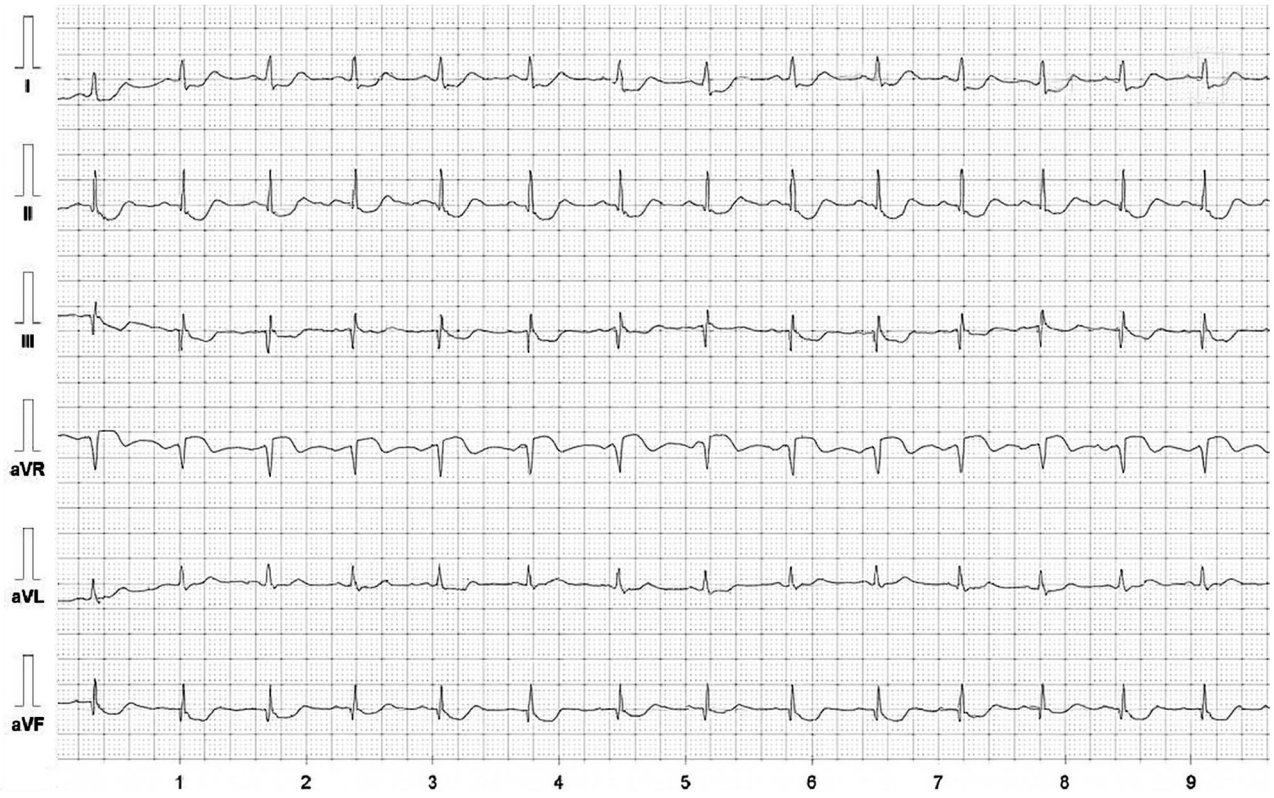

Fig. 1 EKG strip - ST segment depression in I, II, III, aVF, V5 and V6 ECG leads

CT angiography subsequently revealed diffuse pulmonary congestion with thickened interstitium and severe dilatation of the proximal left anterior descending artery (LAD) with thrombotic occlusion (Fig. 2).

Autopsy of lungs and heart

Lungs

Both lungs were heavy and edematous (right 740 g, left 660 g), with frothy, blood-tinged fluid on the cut surface. Microscopically, marked pulmonary edema and congestion with areas of intra-alveolar hemorrhage. No hyaline membranes, no interstitial fibrosis. Bronchioles contained focal blood, pulmonary vessels were patent without thrombi.

Conclusion Findings consistent with acute cardiogenic pulmonary edema due to left-sided heart failure. No features of diffuse alveolar damage or COVID-19-related pneumonia.

Heart

Aneurysm of the left ventricular wall (40 mm in length, 20 × 15 mm in diameter) filled with organized, adherent thrombus (Fig. 3). Several well-demarcated foci of myocardial softening (6–10 mm) in the anterior septum and left ventricular wall, consistent with old infarcts.

Coronary arteries (LAD, circumflex, RCA) showed marked intimal thickening, focal calcification, and severe luminal narrowing.

Microscopy Chronic inflammatory infiltrate with fibrous intimal thickening and focal calcifications in coronary arteries; loss of elastic fibers. Myocardium with cardiomyocyte hypertrophy and areas of replacement fibrosis. (Figures 4 and 5)

Conclusion Chronic coronary disease with advanced vascular changes, myocardial ischemic injury with old infarcts and aneurysm formation, complicated by coronary thrombosis.

Discussion

KD is an inflammatory disorder of young children, associated with vasculitis of CA with subsequent aneurysm formation in up to one-third of untreated patients [4].

Typical KD is diagnosed when fever lasting more than 4–5 days is associated with ≥ 4 principal clinical criteria (bilateral non-exudative conjunctivitis, changes of lips and oral mucosa, cervical lymphadenopathy, polymorphous exanthema, changes of the extremities and perineal region). Incomplete KD, mostly seen in children younger than 12 months, is suggested if a fever lasting for more than 5 days is associated with < 4 (2–3/5) principal

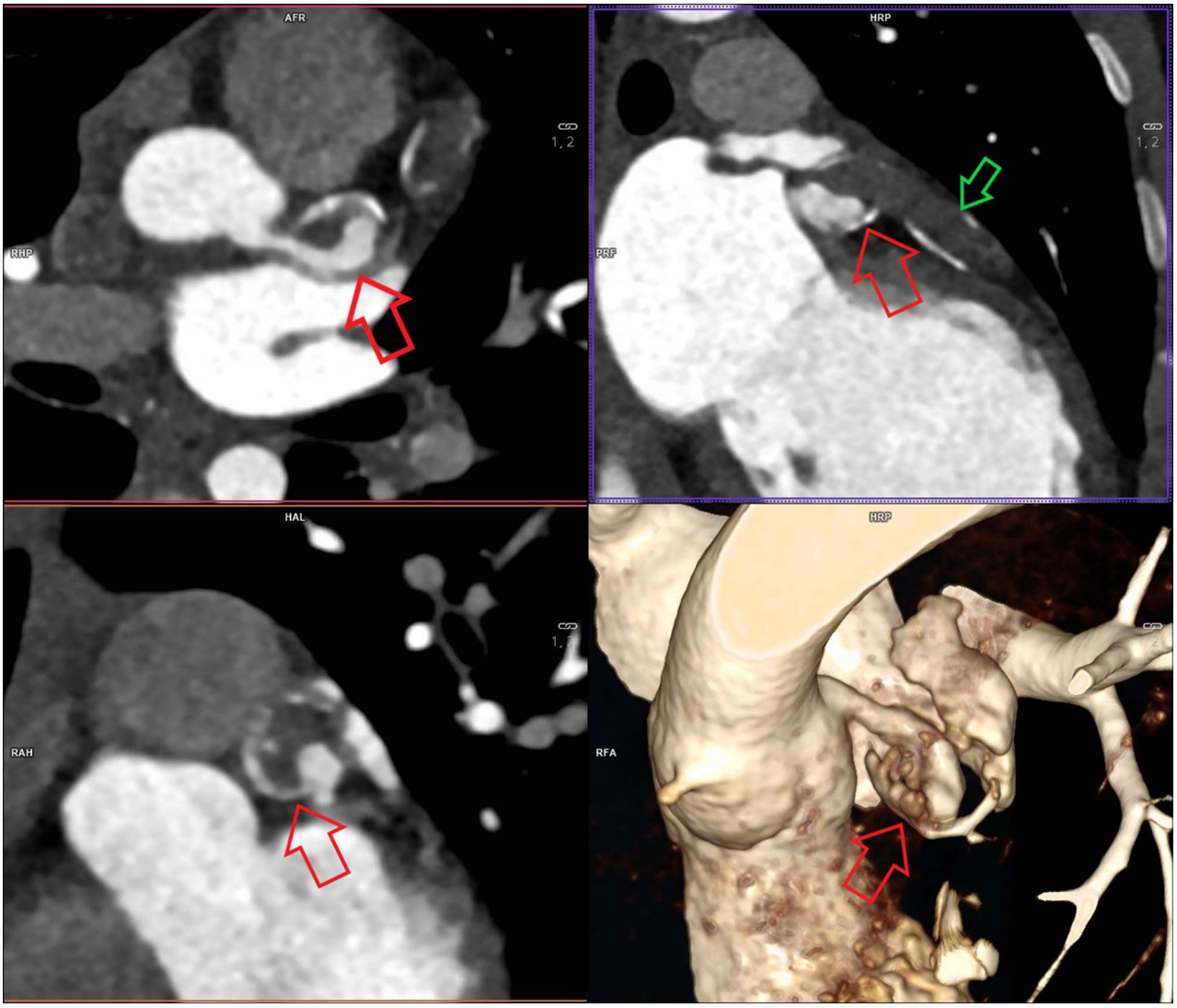

Fig. 2 CT scan image - Aneurysm (red arrow) of the proximal third of LAD with mural thrombosis, which causes occlusion of the further course of the dilated artery (green arrow)

clinical features, compatible laboratory or echocardiographic findings and other febrile illnesses have been excluded [5, 6]. We assume that the myocarditis seen in our patient at the age of 5 was most likely an incomplete form of KD. According to the parents and available medical records, our patient presented with fever, pharyngitis and neck lymphadenopathy at that time. No coronary pathology had been diagnosed on echocardiography scans and the patient had been treated for myocarditis (insufficient immunosuppressive therapy had been initiated). In our opinion, the coronary artery aneurysm (CAA) in our patient developed due to inflammatory involvement at the age of 5.

CAA may regress, remain stable or enlarge over time. Fortunately, up to half of CAA regress spontaneously within the first 2 years of disease onset. Predicting the risk of developing CAA remains challenging [4, 7]. CA complications are the primary cause of morbidity and mortality related to KD and have become the leading form of acquired heart disease in children in developed countries [4, 7]. A standardized imaging protocol supervised by an experienced pediatric echocardiographer, including a specific sequence of imaging views to clearly delineate all segments of the CA, is necessary for an accurate and complete assessment [8]. Serial echocardiographic studies in acute KD show that CA dilatation may be visible early in the illness, but maximal development is usually in the second and third week of the acute illness [9]. The initial CA diameter is a factor determining the risk of CA [7]. The proximal LAD and the proximal right CA are the most frequent locations of CAA, and the posterior descending artery is the least common [5]. This corresponds to the findings in our patient.

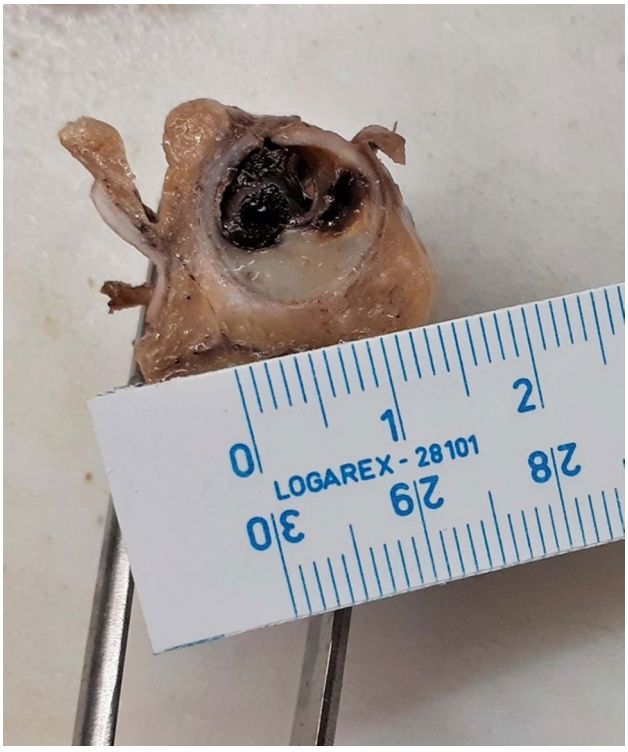

Fig. 3 Mural thrombus within an aneurysmal segment of the LAD

Patients with CAA might develop thrombosis or stenotic lesions of CA leading to myocardial ischemia, infarction, or death [10, 11]. The risk of aneurysm thrombosis is greatest in the first 2 years after the acute episode but persists lifelong. Despite the 3-year follow-up in the cardiac center, no CA pathology had been diagnosed at the time of the acute illness or during the follow-up period in our patient. The presence of an aneurysm of the LAD artery had most likely been overlooked. No chronic antithrombotic or anticoagulant therapy was initiated. Our patient had presented no episodes of severe chest pain, shortness of breath or other severe clinical symptoms despite the regular intensive physical activity (club soccer player). Only occasionally he reported a short feeling of tightness in the chest, which always resolved spontaneously within minutes, and which did not limit his physical activity. These episodes were most likely manifestations of myocardial ischemia associated with an undiagnosed coronary artery aneurysm secondary to Kawasaki disease. Recognition of such ischemic symptoms at that time might have prompted appropriate medical evaluation and management, potentially altering the subsequent course of the disease [10].

The main goal of treatment in the acute phase of KD is to suppress systemic inflammation to minimize the risk of CAA development. The highest risk reduction of CAA development is achieved when the treatment is initiated as early as possible within the first 7 days of illness and no later than 10 days after disease onset. The rate of CA aneurysm development increases significantly after days 8–9. Numerous international treatment guidelines have been published [5, 12–14].

Aspirin and IVIG are two drugs that are conventionally used to treat KD. The early single transfusion of IVIG (2 g/kg) is currently the most effective anti-inflammatory treatment of KD, substantially reducing the prevalence of persistent CA lesions [12–14]. Historically, high dosages of aspirin were used during the acute phase of KD for anti-inflammatory effects, but there is no evidence of a benefit with high versus low dose aspirin when considering coronary vascular damage. Aspirin treatment does not reduce the risk of coronary aneurysms but risk of CA thrombosis [15, 16]. The duration of antiplatelet therapy is determined by the extent of CA involvement. In the absence of CA involvement, aspirin is administered for a total of 6–8 weeks, in the transient involvement of CA up to complete regression and in the presence of CAA for life.

Children diagnosed with incomplete KD are at an increased risk of cardiac complications. The lack of recognition and subsequent absence of appropriate anti-inflammatory and antithrombotic treatments significantly elevate the likelihood of thrombotic events, as illustrated in the case of our patient. Aneurysm size is the strongest predictor of major cardiac events. Our patient developed a giant aneurysm and met the criteria for combined anticoagulation and antiplatelet therapy. However, this treatment was not initiated as the diagnosis was not recognized [5, 10].

Inflammatory vasculopathy resembling acute KD affecting children who had prior infection with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) emerged recently. The term MIS-C associated with COVID-19 has been used to describe the condition. As MIS-C shares clinical features with acute KD, this condition must be included in differential diagnosis of acute KD [17–19]. As our patient did not present with signs and symptoms corresponding to MIS-C, and PCR was negative from nasopharyngeal and tracheal sample. There were also no signs of inflammation corresponding to MIS-C or Covid-19 infection in the histology of the lungs. Recent evidence shows that the outcome regarding coronary arteries in patients with MIS-C is generally excellent [20].

The development of critical myocardial ischemia and malignant arrhythmia are among the most serious complications of long-term CA disease. When compared with the incidence of KD, only a limited number of cases described in the literature suffered from sudden death, underwent autoptic and histologic examination as this condition is rarely fatal [21–23].

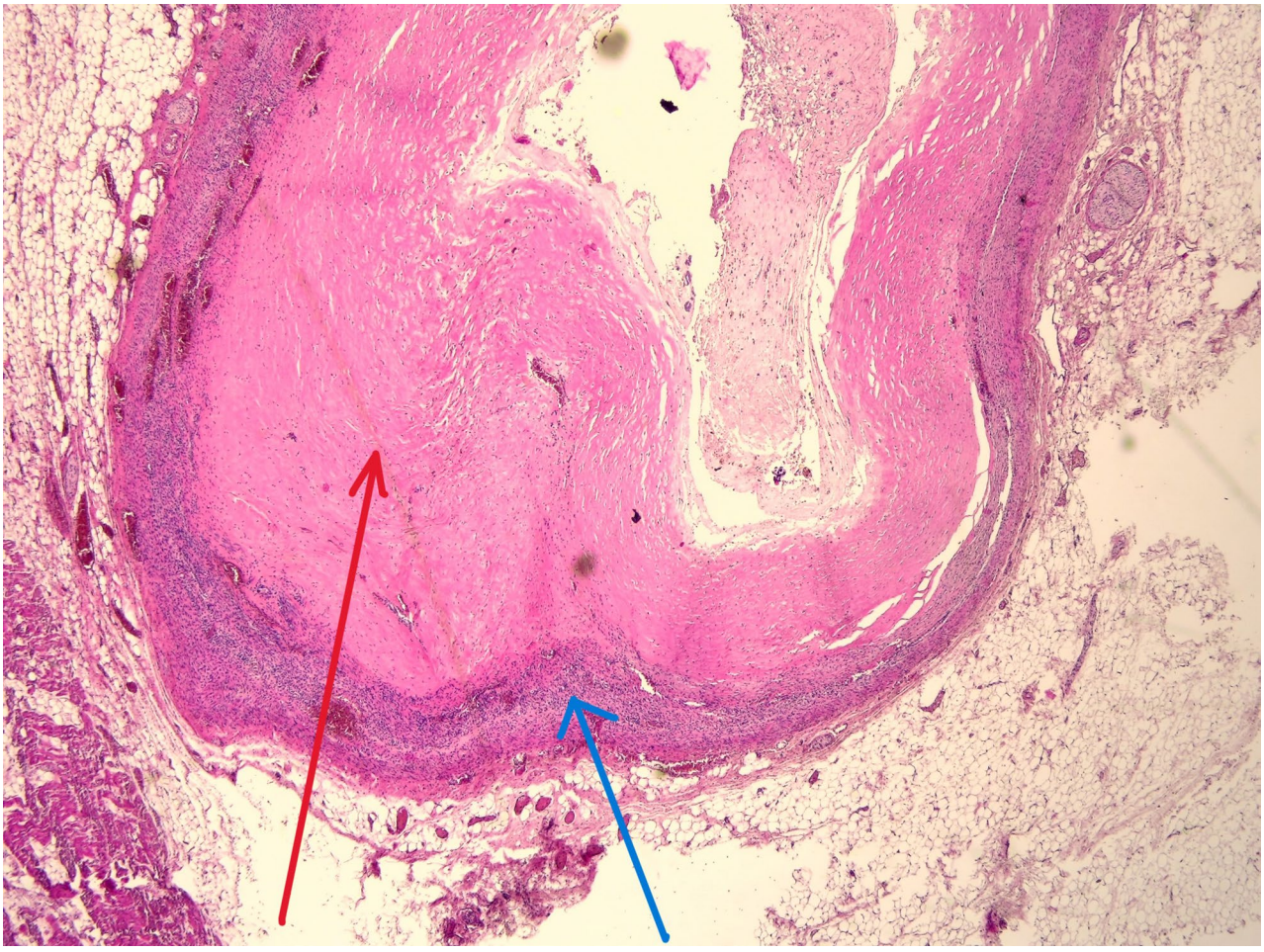

Fig. 4 Microscopic image - CA with chronic inflammatory infiltration of the vessel wall (blue arrow). Marked thickening of the tunica intima with secondary regressive changes (red arrow)

Pediatric services are largely unfamiliar with the detection and management of myocardial ischemia due to its rarity in children. Patients with a previous history of KD presenting with acute chest pain should be urgently transported to the tertiary pediatric cardiac surgical center as clinical features cannot reliably determine the underlying etiology. Patients with STEMI (ST elevation myocardial infarction) require urgent catheter re-vascularization or surgical revascularization. Thrombolysis is initiated in cases of CAA occluded by a thrombus [10, 11].

The diagnostic methods of choice for non-STEMI (non-ST- elevation myocardial infarction), as demonstrated in our patient's case, is ECG, serial high sensitive troponin levels, echocardiography and coronary imaging with CT or coronary angiography.

Due to possible development of significant coronary collaterals over time, serial ECG and hsTnT levels may be unremarkable, even with significant myocardial ischemia [10, 11]. In our patient, the clinical circumstances did not permit repeat measurements of troponin levels. The initial hsTnT concentration fell within the intermediate, or 'grey-zone,' range; however, the ECG demonstrated clear ischemic ST–T changes pathognomonic of an acute coronary syndrome. These findings should have prompted the immediate initiation of intravenous heparin and antiplatelet therapy while awaiting advanced imaging. Prompt and accurate interpretation of the CT angiography was therefore crucial, as it might have enabled the timely initiation of systemic thrombolysis and potentially reduced the risk of a fatal outcome.

Conclusion

KD is the leading cause of acquired coronary disease in children, and patients with CAA remain at lifelong risk of myocardial infarction, arrhythmia, and sudden death. Early recognition and prompt initiation of appropriate therapy are essential to prevent long-term complications. Emergency physicians should be aware, that clinical symptoms of patients with late CA complications may be subtle, and patients with a previous history of KD or previous unexplained myocarditis presenting with suspected

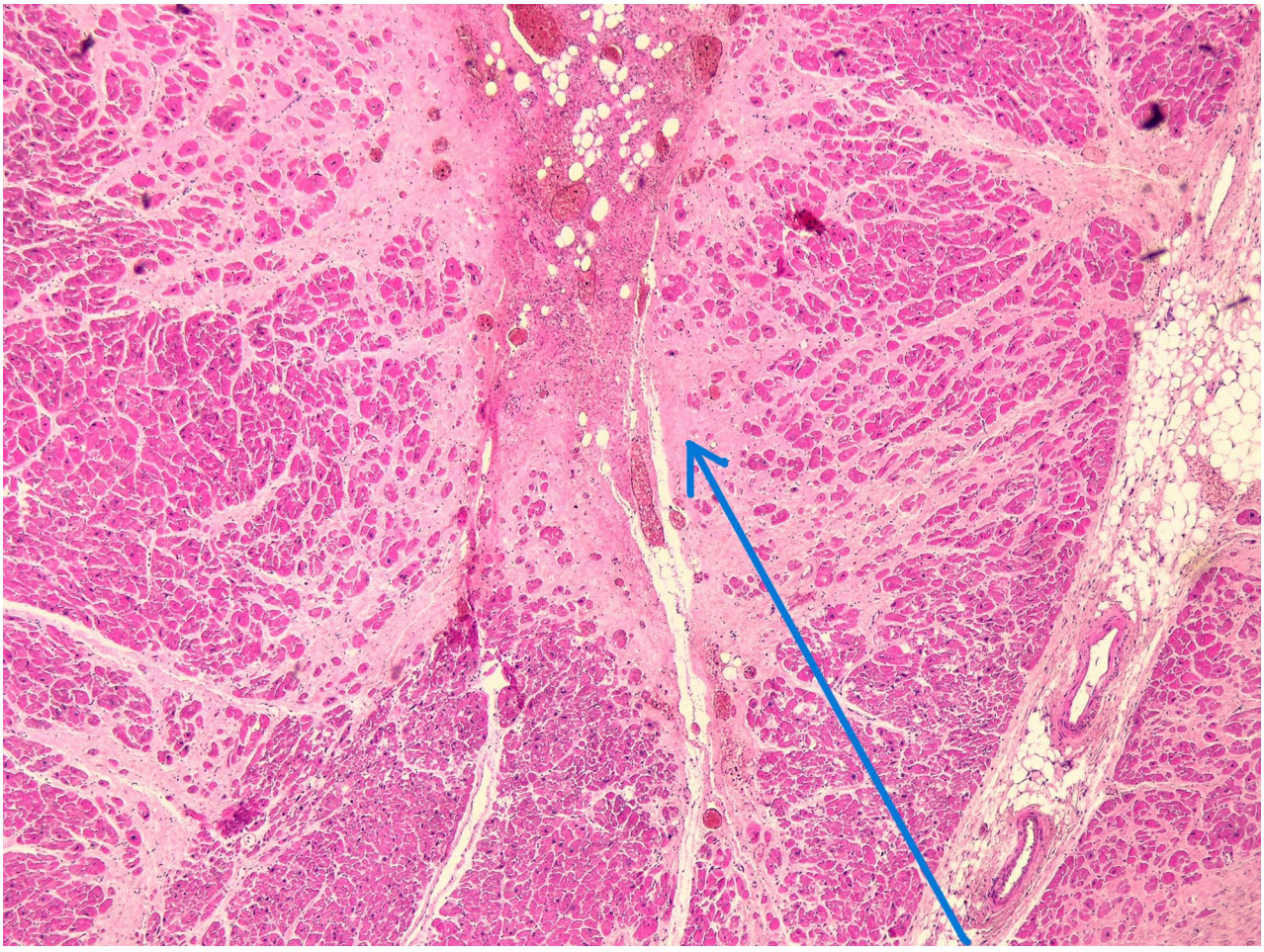

Fig. 5 Microscopic image - Multiple areas of myocardial fibrosis in the wall of the left ventricle with compensatory hypertrophy of cardiomyocytes (blue arrows)

cardiac emergency require timely referral to a pediatric congenital cardiac center.

Abbreviations

CA	Coronary artery
CAA	Coronary artery aneurysm
COVID-19	Coronavirus Disease 2019
CT	Computed Tomography
ECG	Electrocardiogram
EF	Ejection fraction
IVIG	Intravenous immunoglobulin
KD	Kawasaki disease
LAD	Left anterior descending (artery)
MIS-C	Multisystem Inflammatory Syndrome in Children
RT-PCR	Reverse transcription polymerase chain reaction
SARS-CoV-2	Severe acute respiratory syndrome coronavirus 2
STEMI	ST-elevation myocardial infarction
non-STEMI	non-ST- elevation myocardial infarction

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Author contributions
FJ, FT - original draft preparation; writing – review and editing, HM, KA, SJ, BJ, HL contributed to clinical evaluation, imaging, and post-mortem analysis. All authors critically revised the manuscript and approved the final version.

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Data availability
All relevant data are included within the manuscript. Additional information is available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
This case report was conducted in accordance with the Declaration of Helsinki. Ethical approval was not required for this single-patient case report. Written informed consent for participation was obtained from the patient's parents.

Consent for publication
Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images.

Competing interests
The authors declare no competing interests.

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