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MINI-FOCUS ISSUE: IMAGING

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Acute Adult-Onset Kawasaki Disease Complicated by Coronary Artery Aneurysms, Thrombosis, and ST-Segment Elevation Myocardial Infarction

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ABSTRACT

A 21-year-old man who had an initial misdiagnosis of chest wall cellulitis and sepsis presented to the emergency department with chest pain. Electrocardiogram demonstrated ST-segment elevation in the inferior leads. Cardiac catheterization identified diffuse aneurysmal dilation and thrombosis of the distal right coronary artery. Clinical signs were consistent with acute Kawasaki disease. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:276–9)
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A 21-year-old man of Japanese and French descent presented to the emergency department (ED) with acute chest pain radiating to his left shoulder. His blood pressure was 137/82 mm Hg, heart rate was 61 beats/min, and temperature was 37.1°C. He was in moderate distress because of the pain and general malaise. His clinical examination findings included nonsuppurative conjunctivitis, dry and cracked lips with an erythematous tongue, mild swelling and erythema of his palms, and a faint maculopapular rash on his chest.

LEARNING OBJECTIVES

- To recognize the signs and symptoms of acute KD in adults, including persistent fever and rash.
- To recognize adult-onset KD as part of the differential diagnosis of chest pain with ST-segment elevation in a young adult.
- To initiate prompt therapy for KD to prevent further coronary artery damage and potentially fatal complications.

PAST MEDICAL HISTORY

The patient had no known past medical condition and no history of smoking. His illness started with chest discomfort, rash on his chest, and fever in May 2013, 20 days before the ED visit. He was evaluated by his primary care provider, was given a diagnosis of chest wall cellulitis, and was started on antibiotics. His clinical picture progressed with continued fever, worsened rash, and generalized malaise with dizziness. As a result of hypotension (90/50 mm Hg) and tachycardia (100 beats/min) at the follow-up visit

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4 days after the initiation of antibiotics, he was admitted to the hospital with the presumed diagnosis of sepsis. During the 8 days of hospitalization, he was given broad-spectrum antibiotics and supportive measures for hypotension, but he did not require admission to the intensive care unit. Leukocytosis (white blood cell count, 37,000/μl), elevated C-reactive protein (222 mg/l), sterile pyuria, and hypoalbuminemia developed. His repeated blood cultures remained sterile. An echocardiogram showed mildly decreased left ventricular function (left ventricular ejection fraction [LVEF] 51% by the biplane Simpson method) without any apparent wall motion abnormality, mild right ventricular dilation with normal function, no mitral regurgitation, mild tricuspid regurgitation, and a small pericardial effusion. Computed tomography (CT) of his chest and abdomen failed to reveal any infectious focus and did not show any coronary artery dilation 13 days before his ED presentation. Cervical lymphadenopathy, erythema of his palms, and mild conjunctivitis without discharge developed. Multiple viral studies (human immunodeficiency virus, hepatitis panel, monospot, cytomegalovirus) returned negative results. A repeat echocardiogram 3 days after the initial echocardiogram continued to show a mildly decreased LVEF (50% to 55%) without wall motion abnormality. His fever and rash improved gradually, and he was discharged after 8 days of hospitalization. A week after discharge, he presented to the ED with new onset chest pain.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses of acute chest pain in a young adult man includes angina, ST-segment elevation myocardial infarction (STEMI), pericarditis, myocarditis, aortic dissection, pulmonary embolism, pneumonia, and musculoskeletal causes.

INVESTIGATIONS

An electrocardiogram demonstrated ST-segment elevation in the inferior leads, consistent with acute myocardial infarction (**Figure 1A**). Cardiac enzymes were elevated, point-of-care troponin 0.59 ng/ml in ED, troponin-I 4.01 ng/ml (peak 15.79 ng/ml) and creatine kinase-myocardial band 5.7 ng/ml (peak 160.5 ng/ml). The patient underwent emergency cardiac catheterization, which demonstrated diffuse dilation of both coronary artery systems (Videos 1 and 2), with multiple ectasias throughout the entire left anterior descending (LAD) coronary artery, circumflex artery, and right coronary artery (RCA) (**Figures 1B and 1C**), measuring between 8 and 11 mm in diameter. A clot was identified in the distal RCA, with high-grade obstruction and a filling defect in the distal RCA and posterior descending artery (Video 1, **Figure 1C**). No wall motion abnormality was noted on left ventriculography. An echocardiogram confirmed dilated proximal segments of the LAD (**Figure 1D**) and the RCA, but no significant wall motion abnormality was noted.

MANAGEMENT

Following cardiac catheterization he was admitted to the intensive care unit and was started on thrombolytic therapy with eptifibatide. Pediatric cardiology and infectious disease consultations helped with the diagnosis of primary Kawasaki disease (KD) on the basis of the clinical signs and diffuse dilation of the coronary arteries. His past medical history was again carefully reviewed to exclude a previous history of KD. In retrospect, KD was not suspected during the initial encounter because coronary artery dilation was not present on the CT scan. He was started on intravenous immunoglobulin (IVIG) therapy and anticoagulation with enoxaparin because of the giant coronary aneurysms. His fever and chest pain resolved, his clinical symptoms improved, and his electrocardiogram normalized within 24 h. His follow-up echocardiogram showed resolution of the previously noted left ventricular systolic dysfunction (repeat LVEF, 64%), likely in response to resolution of mild myocarditis associated with KD. He was discharged from the hospital in 4 days with warfarin and aspirin.

DISCUSSION

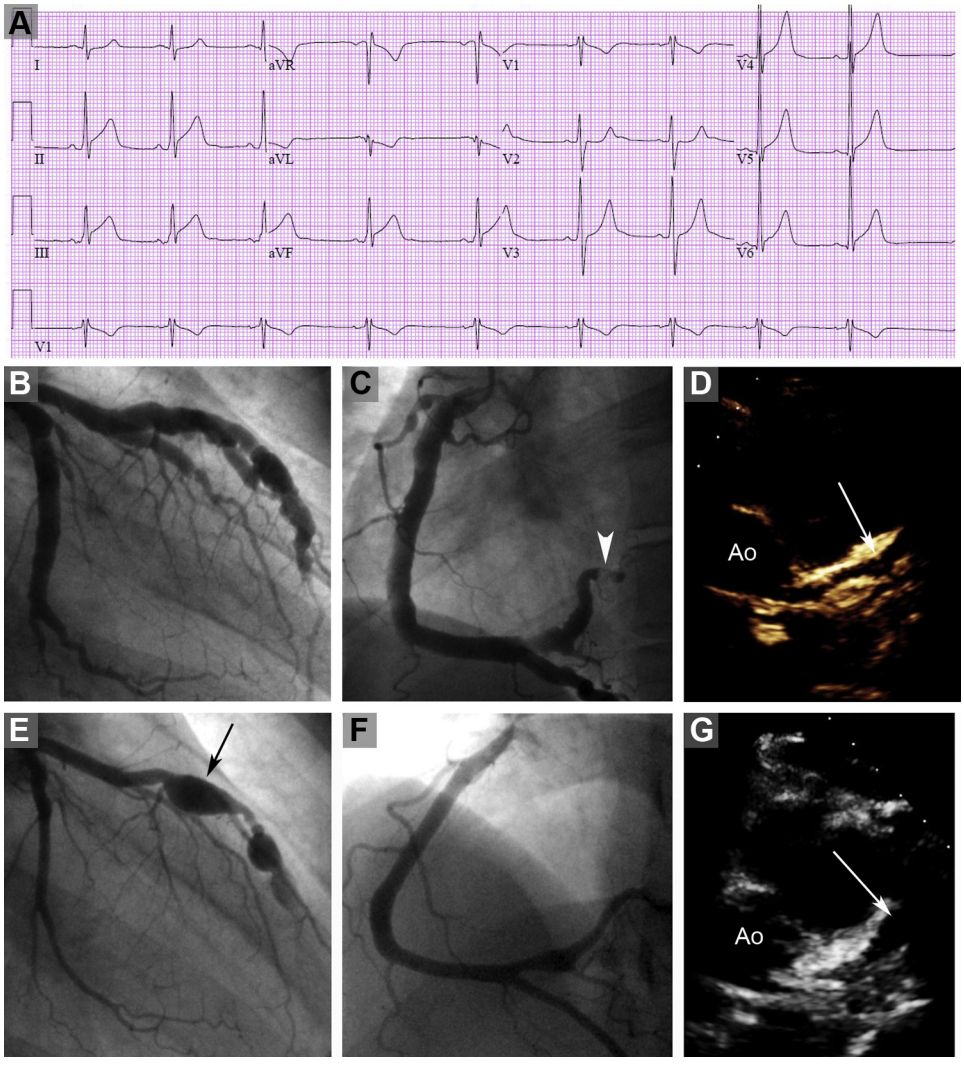
KD is the leading cause of acquired heart disease in children in the United States and many other industrialized countries (1), and it rarely occurs in adults (2). The diagnosis is based on the presence of at least 5 days of fever and 4 of the following 5 clinical features: polymorphic exanthema, erythema or edema of hands and feet, nonsuppurative conjunctivitis, erythematous lips and tongue, or cervical lymphadenopathy (2). The lack of a confirmatory test in a disease with an unknown origin poses continued diagnostic and therapeutic challenges (1,2). In the rare occasion of adult-onset KD (3,4), the disease is frequently diagnosed late during the course of illness, thus increasing the risk for coronary complications with subsequent morbidity (5,6).

Coronary artery complications typically occur in 20% to 25% of untreated cases (1), and these complications usually develop during the second week of

ABBREVIATIONS AND ACRONYMS

CT = computed tomography
ED = emergency department
IVIG = intravenous immunoglobulin
KD = Kawasaki disease
LAD = left anterior descending coronary artery
LVEF = left ventricular ejection fraction
RCA = right coronary artery
STEMI = ST-segment elevation myocardial infarction

FIGURE 1 ST-Segment Elevation Myocardial Infarction Secondary to Coronary Artery Dilation and Thrombosis in a 21-Year-Old Man With Acute Primary Adult-Onset Kawasaki Disease



(A) Electrocardiogram shows ST-segment elevation in inferior leads at the time of presentation with chest pain. **(B)** Angiogram of the left main coronary artery on the day of presentation with chest pain demonstrates diffuse aneurysmal dilation of the entire vessel, including the left anterior descending coronary artery and circumflex branches. **(C)** Angiogram of the right coronary artery demonstrates diffuse dilation with a filling defect **(arrowhead)** suggesting coronary thrombosis consistent with the clinical findings of inferior ischemia. **(D)** Echocardiogram during admission demonstrates a dilated segment in the left anterior descending coronary artery **(arrow). (E)** Angiogram of the left main coronary artery 4 months after the onset of Kawasaki disease demonstrates residual large aneurysms **(arrow)** in the course of the left anterior descending coronary artery. **(F)** Angiogram of the right coronary artery 4 months after the onset of Kawasaki disease demonstrates no residual aneurysms and no filling defect. **(G)** Echocardiogram 4 months after the onset of Kawasaki disease demonstrates persistent aneurysmal dilation of the midsegment of the left anterior descending coronary artery **(arrow)**, consistent with angiography. Ao = aorta.

illness (7). Proper anti-inflammatory treatment with IVIG given within 10 days of fever onset decreases the risk of coronary artery dilation or aneurysm from 25% to 5%. Risk factors for aneurysm formation include prolonged fever, unfavorable response to IVIG therapy, and young age (<6 months) or old age (>10 years) at presentation (1,2). The size of the coronary aneurysm is the most important prognostic factor for persistent coronary artery disease (8). Giant (>8-mm diameter) coronary aneurysms have a poor prognosis because they usually do not resolve (8,9). It is well known that childhood KD complicated by

aneurysms can lead to adult onset coronary artery problems, such as thrombosis and myocardial ischemia (9). However, adult-onset KD manifesting as STEMI secondary to coronary thrombosis has not been described before.

In this case, although the patient was barely beyond his teenage years, given the unusual age at presentation, KD was not suspected until the recognition of coronary artery aneurysms complicated by STEMI. The differential diagnosis of coronary aneurysm in a young adult includes acute primary KD, a history of KD, other vasculitic disease such as Takayashu arteritis and juvenile idiopathic arthritis, infectious disease such as mycotic aneurysm, drug use, and trauma. Given the typical clinical signs of acute KD, the patient received the KD diagnosis and was treated for primary adult-onset KD.

FOLLOW-UP

At 4 months after the onset of KD, follow-up cardiac catheterization with angiograms demonstrated persistent mild dilation of the RCA (Video 3) and aneurysmal dilation of the LAD (Video 4), requiring continued anticoagulation (**Figures 1E and 1F**). As part of the follow-up strategy, the patient was monitored by annual echocardiograms for left ventricular function and cardiac exercise stress test for signs of ischemia. The echocardiogram showed persistent dilation of the proximal LAD (**Figure 1G**). A year after the initial presentation, a CT angiogram revealed persistent fusiform aneurysms in the proximal and middle segments of the LAD (9 mm) and a small aneurysm in the distal RCA (6 mm).

CONCLUSIONS

Primary adult-onset KD is an uncommon disease in adults, and this rarity may lead to a delay in appropriate diagnosis and treatment. Our case, which is the first documented coronary aneurysm in a young adult as a result of primary KD complicated by thrombosis and STEMI, demonstrates the importance of diagnosing KD in adults. The severity of complications in patients with untreated KD warrants a high degree of suspicion in young adults presenting with persistent fever, rash, and nonsuppurative conjunctivitis. Early diagnosis is paramount, allowing timely initiation of specific therapy and possible prevention of potentially fatal coronary complications.

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The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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KEY WORDS coronary aneurysm, Kawasaki disease, myocardial infarction

APPENDIX For supplemental videos, please see the online version of this paper.