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Case Report

Giant aneurysms of the common iliac arteries in convalescent Kawasaki disease initially presenting as aseptic meningitis: a case report

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Abstract: Background: Kawasaki disease (KD) is a systemic vasculitis that most commonly causes coronary artery aneurysms (CAA). The presentation of incomplete KD is atypical and can easily be misdiagnosed, especially in infants. Accurate diagnosis is important because incomplete KD has a higher risk for developing CAA. Systemic artery aneurysm (SAA) rarely occurs in incomplete KD. Case presentation: We present a 3-year-old female with a case of incomplete KD, complicated by giant aneurysms of the common iliac arteries in convalescence. The case was first diagnosed as aseptic meningitis at 8 months of age, and incomplete KD was confirmed when dilation of the coronary arteries was detected by echocardiogram. Intravenous immunoglobulin and aspirin treatment were used with clinical improvement. A coronary artery lesion recovered 3 years later. However, because of intermittent abdominal pain, an abdominal ultrasound was done, and it showed dilated common iliac arteries. Then cardiac catheterization and peripheral angiography demonstrated that the bilateral common iliac arteries were dilated as mirror giant aneurysms, with coronary artery regression. Conclusion: SAA is very rare and more serious in convalescent KD, even when CAA is in regression. Screening for SAA should be considered in incomplete KD, especially KD with CAA in the acute phase.

Keywords: Kawasaki disease, coronary artery aneurysm, systemic artery aneurysm

Background

Kawasaki disease (KD) is an acute vasculitis characterized by fever, conjunctivitis, mucositis, swelling of the hands and feet, truncal rash, and cervical lymphadenopathy. It is more common in children less than 5 years of age. Coronary artery aneurysm (CAA) is the most common complication of KD, which accounts for 15% to 25% of cases in untreated patients [1-3]. Because KD is a systemic vasculitis, aneurysms of other medium-sized arteries (noncoronary) have also been reported, including the axillary, subclavian, brachiocephalic, iliac, and femoral arteries [1, 4, 5]. The incidence of iliac artery involvement in KD is between 0.6% and 2.5% [6]. The presentation of incomplete KD is atypical and easily misdiagnosed, especially in infants. Accurate diagnosis is important because incomplete KD has a higher risk for developing CAA. It is rare that systemic artery aneurysm (SAA) occurs in convalescent incomplete KD. Here, we report an incomplete KD case of a 3-year-old girl who had mirror giant aneurysms of the common iliac arteries in convalescence. Interestingly, this incomplete KD case appeared as aseptic meningitis early in the illness at 8 months of age.

Case presentation

One ethnic Han Chinese female patient presented with one-week history of fever and a 2-day history of bulging fontanelles, at 8 months of age. Peripheral blood routine: the white blood cell (WBCs) count was 29.1×10^9/L, the ratio of neutrophils was 61%, and the C reaction protein (CRP) was 21 mg/L. The examination of the cerebrospinal fluid (CSF) was compatible with meningitis: the WBC count was 35/mm^3 with polymorphonuclear predominance, the protein

Table 1. Acute phase reactants laboratory evolution during hospitalization

Day of illness	D2	D4	D7	D10	D13	D19	D22
Hemoglobin (g/L)	100	105	106	92	85	87	92
White blood cells ($\times 10^9$/L)	29.1	20.2	21.6	18.4	17.5	17.6	12.3
Platelets ($\times 10^9$/L)	478	500	570	598	697	700	701
C-reactive protein (mg/L)	21	45	49	71	80	102	7
Erythrocyte sedimentation rate (mm/h)	/	77	/	/	106	/	68

Incomplete KD was confirmed on Day 19th (D19) and intravenous immunoglobulin (IVIG) 2 g/kg and oral aspirin 50 mg/kg/day were started. Fever, WBCs and CRP were improved on D22 (Day 22th).

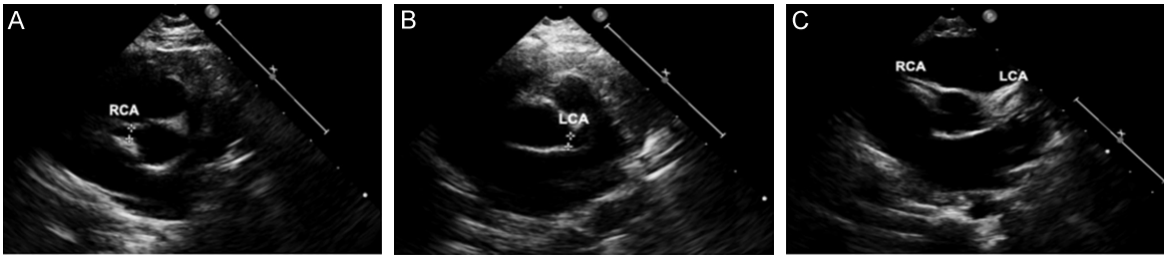

Figure 1. Echocardiogram showed coronary artery dilation. Persistent left and right coronary arteries dilatation which was subsequently improved at week 8. LCA: left coronary artery; RCA: right coronary artery.

was 46 mg/dL, and the glucose was 3.2 mmol/L. The CSF gram stain and culture were sterile. The blood culture was negative. The chest radiography and echocardiogram were normal. Then an intravenous ceftriaxone was started for suspected meningitis. There were no rash, conjunctival injection, changes in the lips or oral cavity, or edema/erythema of the hands. The patient remained febrile for the first 2 weeks, with no response to antibiotics which included ceftriaxone, meropenem, and vancomycin. The second CSF examination showed great improvement 2 weeks after the last antibiotic injection: The WBC count was 16/mm^3, the protein was 29 mg/dL, the glucose was 3.2 mmol/L. However, acute phase reactants were persistently elevated until day 19 of the illness (**Table 1**). Then the second echocardiogram showed right and left coronary artery dilatation (4.6 mm and 4.2 mm in diameter). So, incomplete KD was confirmed. Intravenous immunoglobulin (IVIG) 2 g/kg and oral aspirin 50 mg/kg/day were started, with significant clinical and analytic improvement. The patient was discharged to her home on low-dose aspirin (5 mg/kg/day), diagnosed with incomplete KD.

A follow-up echocardiogram at week 4 of therapy showed persistent coronary artery dilatation (3.8 mm) which was subsequently improved at week 8 (**Figure 1**). Aspirin was discontinued after the complete resolution of the coronary involvement, as demonstrated by an echocardiogram, after 3 months of treatment. However, 3 years later, the patient complained of intermittent abdominal pain for 2 days, and an abdominal ultrasound showed dilated common iliac arteries. The echocardiogram and electrocardiogram were normal. Whether she had additional noncoronary arterial lesions complicated with KD was unknown. Cardiac catheterization and peripheral angiography included an abdominal aortography were demonstrated. There was no significant expansion of the left and right coronary arteries (2.7 mm and 2.1 mm in diameter). The aortic arch, pulmonary artery, and the abdominal aorta were not expanded (**Figure 2**). However, the bilateral common iliac arteries were dilated as in mirror giant aneurysm. The right iliac aneurysm was 16.3×32.3 mm, and the left iliac aneurysm was 12.0×26.1 mm (**Figure 2**).

Discussion and conclusions

KD is a systemic vasculitis that can cause coronary artery aneurysms (CAA) in up to 25% of patients if left untreated [1-3]. Our patient exhibited the presence of more than five days of fever and CAA, indicating she should be diagnosed with incomplete KD [7]. Incomplete KD does not meet all established criteria for classi-

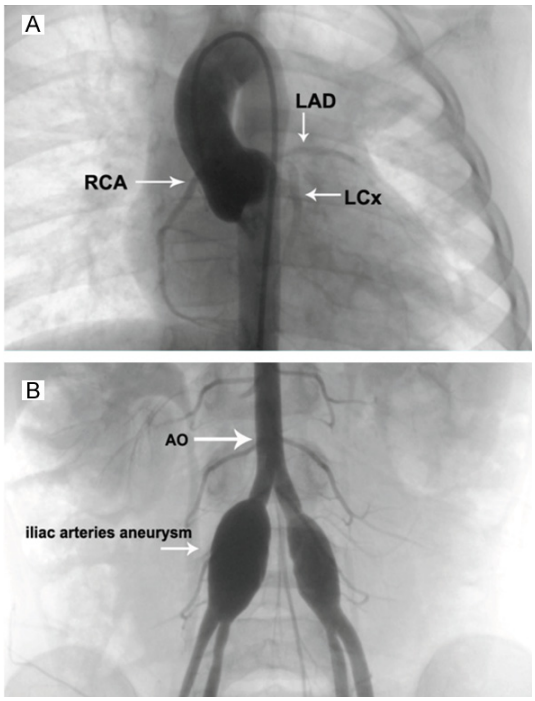

Figure 2. Mirror giant aneurysms of the common iliac arteries. Cardiac catheterization and peripheral angiography included abdominal aortography and demonstrated no significant expansion of the left and right coronary arteries. However, the bilateral common iliac arteries were dilated as mirror giant aneurysm. LCx: left circumflex coronary artery; LAD: left anterior descending coronary artery; RCA: Right coronary artery; AO: aorta.

cal KD, so it is more likely to go untreated [8, 9]. Incomplete KD complicated by aseptic meningitis, or, exhibiting as aseptic meningitis is not very rare [10-12]. It remains unclear whether incomplete KD has a greater propensity to develop CAA or if this complication results from a delayed diagnosis. Sometimes the presence of CAA may be the only definite means of diagnosing incomplete KD [10, 13].

The pathology of CAA has been studied in KD. The acute inflammatory reaction results in systemic vasculitis characterized by a weakening of the internal and external elastic lamina [14]. So, aneurysms may occur if the blood pressure exceeds what the newly weakened vessels can contain [15]. SAA is rare and occurs commonly in the subclavian, brachial, axillary, iliac, or femoral vessels in KD [4, 5]. It is not known precisely when the iliac artery aneurysm first started in our case. The delay in IVIG treatment in our case may be the cause of CAA and the iliac artery aneurysm in the acute illness. In convalescence, our case exhibited only an iliac artery aneurysm but no coronary artery lesions. Multiple mirror aneurysms has been reported in KD [14]. However, the mirror giant iliac artery aneurysm detected in our case of convalescent incomplete KD is the first reported in the literature. As illustrated in our case, SAA is very rare and more serious in convalescent KD, even when CAA is in regression. Theoretical, SAA is more easily to develop in incomplete KD consistent with CAA. Screening for SAA should be considered in incomplete KD, especially KD with CAA in the acute phase. Vessel ultrasound may be useful in detecting the signs of SAA in incomplete KD. Body MRA, cardiac catheterization and peripheral angiography can confirm an SAA diagnosis [4]. This case report may encourage the imaging of the systemic arteries as part of the follow up for incomplete KD, especially with CAA.

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Disclosure of conflict of interest

None.

Abbreviations

KD, Kawasaki disease; CAA, Coronary artery aneurysm; SAA, Systemic artery aneurysm; WBCs, White blood cells; CRP, C reaction protein; CSF, Cerebrospinal fluid; IVIG, Intravenous immunoglobulin.

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