Kawasaki Disease presentation by an uncommon presentation of torticollis: A case report

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Abstract

Kawasaki disease (KD) is an acute, febrile systemic inflammatory disorder of childhood. A limited group of KD patients does not fulfill the classic criteria of KD. This group are generally infants or older children. In some cases, delay in diagnosis atypical KD can lead to coronary artery complication. We report the case of a 7-year-old previously healthy girl presented by isolated cervical adenitis with no fever, conjunctivitis or edema who underwent treatment by suspicious to infection disease. After 10 days some other criteria was expressed and by suspicious to atypical KD, giant coronary aneurysms was diagnosed and coronary artery bypass graft surgery (CABG), aneurysmectomy and aneurysmorrhaphy was done. She had no problem in 2 years follow-up. A highly suspicious should be considered in children presenting with fever and unusual manifestations like lymphadenopathy, especially where empiric antibiotics were ineffective.

Keywords: Kawasaki disease, coronary artery aneurysm, coronary artery bypass graft surgery

Abbreviations

KD= Kawasaki disease

CABG= coronary artery bypass graft surgery

CT= computed tomography

IVIG= Intravenous immune globulin

LV= left ventricular

MR= mitral regurgitation

EF= ejection fraction

LIMA= Left internal mammary artery

LAD= left anterior descending artery

RCA= Right coronary artery

Introduction

Kawasaki disease (KD) is a systemic vasculitis involving all blood vessels, but principally medium-sized arteries and is a common vasculitis in childhood (1). It can lead to severe adverse events, morbidity and mortality if not suitably managed. Majority of patients have less than five years old and male to female ratio is about 1.3-1.6:1 (2, 3). It is a widespread disorder all over the world with higher predispose in Asian ethnic. Typically, KD can be diagnosed by fever persisting for at least 5 days, and the presence of at least four of the following principal features: (1) changes in the extremities; (2) polymorphous exanthema; (3) bilateral bulbar conjunctival injection without exudates; (4) changes in the lips and oral cavity; and (5) cervical lymphadenopathy (>1.5 cm diameter). Exclusion of other diseases with similar symptoms is also necessary (4). When involving the cardiovascular system, patients may present with myocarditis, pericarditis, coronary artery aneurysm, and aortic root dilatation (2).

Because of the systemic involvement, it has high variability in all organ systems, and early diagnosis in patients with atypical KD or in those with uncommon manifestations is difficult. With the wide-spread use of computed tomography (CT), there is an increasing number of reports of KD with retropharyngeal edema and enlarged cervical adenopathy (5, 6). Progressing into shock is uncommon, although some studies have reported patients with KD and shock syndrome (7, 8). In this report in addition to interesting echocardiographic and angiographic films and views, diagnostic criteria, cardiovascular involvement, risk factors also have been reviewed in KD patients.

Case presentation

A 7-year-old girl presented by isolated cervical adenitis with no fever, conjunctivitis or edema. She had no history of previous disease, and had appropriate development. She was treated by suspicious to infection disease with cephalexin. Because of no suitable response, cefotaxime and clindamycin was started. Pain and swelling over the right side of her neck with torticollis was remained. There was no history of trauma to head/neck. Due to prolonged fever (39-40 C), fatigue, malaise, ankle, hand and elbow pain and arthritis, she was admitted in our hospital

The child was immunized for her age as per the national vaccination program. After 7 days, she deteriorated and was admitted in hospital by torticollis, evidence of systemic inflammation and treatment failure. She was ill and her head was tilted to the right with chin rotation to the left. Her vital signs at time of admission were: heart rate: 85/min, respiratory rate: 25/min, blood pressure: 110/80 mmHg and temperature: 37.2°C. After 10 days, other classic signs of KD were revealed. Physical examination found erythematous and congested throat, normal breath sounds with palpable lymph nodes, no audible heart murmur, and no skin rash. Initial laboratory data demonstrated pyuria, white blood cell count of 23.4 $x10^9$/L, and an elevated CRP level of 103 mg/L and platelet count 620000/mm^3. She was put on intravenous antibiotic (cefotaxime and vancomycin). She had received steroid, because of lacking IVIG in pediatric infectious services and aspirin was started at an anti-inflammatory dose at 80 mg/Kg in four divided doses. 2-dimensional echocardiography in day 14 showed giant coronary aneurysms, left ventricular (LV) dyskinesia and mitral regurgitation (MR) beside 60 percent EF. Catheterization and selective coronary angiography at day 16 of disease showed giant aneurismal dilatation of both coronary arteries, clot formation in coronary arteries and LV dysfunction

(Figure1). Coronary artery bypass graft surgery (CABG; LIMA to LAD and venous graft to RCA), aneurysmectomy and aneurysmorrhaphy was done by cardiac surgeon at the second month of disease. Although LV function improved and she was not any complaint but warfarin therapy continued due to aneurismal LAD. Recent angiography was done also (Figure2). She is followed up until now (2 years, every 6 months) and has no sequel.

Discussion

Atypical initial presentation in KD is very important, because it can lead to serious complications, especially cardiovascular adverse events. Atypical or incomplete KD diagnosis is challenging. Uncommon manifestations of KD in children have been reported in previous case reports including pyuria, meningitis, ventricular arrhythmia, or shock (9, 10). Introducing such atypical presentations of KD will be beneficial in increasing physicians' attention in suspicious to KD and prevent delayed diagnosis. In the case, we mentioned here, a seven-year-old girl presented with initial isolated torticollis that subsequently other classic symptoms were added and finally giant aneurysm was diagnosed.

Torticollis in this seemed to be due to painful cervical lymphadenitis (11). The rate of cervical lymphadenopathy occurring as the initial presenting symptoms is only approximately 12% (12). Despite of uncommon manifestation, some cases are reported previously KD presented by torticollis. Dyer et al. reported a 6-year-old male who presented with torticollis. In their case, a series of investigations for elevated inflammatory markers revealed dilated coronary artery aneurysms on echocardiogram and treated with high-dose IVIG and low-dose aspirin (11). Runel-Belliard et al. reported a case of KD with arthritis in a 4-year-old girl whose initial presentation was a febrile torticollis (13). In another case, a nine-year-old boy presented with fever, lymphadenopathy, parotitis, and torticollis with initial diagnosis of infective parotitis and finally KD was diagnosed (14). In our case, she was underwent CABG because of LV dysfunction and after that, she was followed and was healthy until 2 years later.

Dilatation of the coronary arteries can occur in approximately 10-20% of KD patients. In half of them, the aneurysms can regress within 1 or 2 years, and no abnormalities are found by angiography in the coronary-arterial system. In the remaining, in contrast, the aneurysms can persist with obviously irregular lumens of the coronary arteries. In 3% of the patients initially having aneurysms, coronary-arterial obstruction progresses. The time-span between the onset of the disease and development of the coronary-arterial stenosis leading to CABG varies from several months to 20 years. The indications for CABG are to be determined not only by findings derived from angiography, but also on the basis of other clinical factors, such as severity of myocardial ischemia, history of myocardial infarction and ventricular performance (15-19). It is very difficult to determine the indications for catheter intervention and CABG in children, based on the diagnosis and localization of the affected coronary artery, using standard methods (20).

Conclusion

The main finding of this case was very atypical presentation of silent KD presented by torticollis. Patient had no manifestations of KD, the diagnosis was not based on criteria, and it was out of it. So, in isolated torticollis, considering KD diagnosis in children as atypical KD should be emphasized beside other differential diagnosis.

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Conflict of interest

None.

Authors' contributions

H.M.M analyzed and interpreted the patient data regarding the cardiovascular disease and managed patient. F.K helped in data gathering and diagnosis patient in echocardiographic evaluation. M.H.N helped in

management of patient, diagnosis and was a major contributor in writing the manuscript. M.S.S helped in diagnosis and management of patient. All authors read and approved the final manuscript.

Ethical approval

This study was performed in accordance with the Helsinki declaration. Data published anonymously.

Consent statement

A written informed consent was obtained from patient.

Data Availability

Data sharing is not applicable to this article, as no datasets were generated or analyzed during the current study.

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Figure legends

Figure1- Initial angiography

Figure2- Postoperative angiography. a) angiographic view, b) LIMA to LAD, c) venous graft to RCA.

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