

Deep neck inflammation as a presentation of Kawasaki disease

Abstract

Kawasaki disease is typically a self-limiting condition that is a common cause of pediatric vasculitis¹ and the leading cause of pediatric acquired heart disease in developed countries.² The disease most frequently occurs in children aged between 6 months and 5 years³ and is associated with coronary artery aneurysms in up to 25% of untreated cases.⁴ There is currently no laboratory test for diagnosing Kawasaki disease. Rather, diagnosis depends on recognizing established clinical criteria.⁵ Unfortunately, atypical manifestations of KD appear to be on the rise, decreasing the likelihood of timely diagnosis and appropriate treatment. Herein, we report an unusual case of Kawasaki disease: a 5-year-old boy who presented with fever and right side cervical lymphadenitis.

Keywords: Kawasaki disease, deep neck, pediatric vasculitis, c-reactive protein, sternocleidomastoid muscle

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Abbreviations: CRP, C-Reactive Protein; EBV, Epstein-Barr virus; ESR, Erythrocyte Sedimentation Rate

Introduction

Kawasaki disease is typically a self-limiting condition that is a common cause of pediatric vasculitis¹ and the leading cause of pediatric acquired heart disease in developed countries.² The disease most frequently occurs in children aged between 6 months and 5 years,³ and is associated with coronary artery aneurysms in up to 25% of untreated cases.⁴

There is currently no laboratory test for diagnosing Kawasaki disease. Rather, diagnosis depends on recognizing established clinical criteria.⁵ Unfortunately, atypical manifestations of KD appear to be on the rise,¹ decreasing the likelihood of timely diagnosis and appropriate treatment. Herein, we report an unusual case of Kawasaki disease: a 5-year-old boy who presented with fever and right side cervical lymphadenitis.

Case description

A 5-year-old boy presented with an 8-days history of fever and neck pain with swelling that is associated with restriction of neck movement. The patient was seen in a private clinic and diagnosed with acute viral pharyngitis and was started on paracetamol 15mg/kg without effect.

A physical examination revealed highly febrile child with T max of 39.1o C with tachycardia at rest. Neck examination revealed bilateral firm cervical lymphadenopathy with right side anterior cervical lymph node measuring 4x4 cm that is tender to palpation and crossing the sternocleidomastoid muscle. Thy lymph node was not fluctuating. Cardiovascular exam revealed a vibratory grade 3 ejection systolic murmur over the left upper sternal border, with no radiation and was best heard in supine position. There were no other abnormal physical examination findings.

Blood testing revealed the following: White Blood Cell 16,000 / mm³ with neutrophil predominance (89%), Hemoglobin of 9.5 g/dL, platelet count, 332,000/mm³, C-reactive protein (CRP) concentration, 334 mg/L, and ESR 105 mm/hr. A neck ultrasound revealed bilateral

hypochoic masses suggestive of cervical lymphadenitis with no evidence of abscess or collection. Acute cervical lymphadenitis was suspected, and treatment with intravenous clindamycin (40 mg/kg/day divided Q6H) was initiated.

5 days after admission the patient started to have red cracked lips with strawberry tongue. He was also noticed to have bilateral conjunctivitis sparing the limbus. No rash was reported at that time and no extremity changes. Blood tests repeated at that time revealed WBC counts of 10,000/mm³ and CRP concentrations of 280 mg/L, ESR concentration was 135 mm/hr. The patient's condition had not improved, and the neck masses remained painful.

Despite 5 days of intravenous antibiotic treatment and despite the improvement in the inflammatory markers, the patient continued to be febrile and the cervical lymphadenopathy revealed minimal improvement. Hence, a CT scan of the neck was performed which revealed bilateral cervical soft tissue fullness in keeping with enlarged lymph nodes. There was no abscess or collection. Ultrasound of the abdomen was obtained because of the patient's complaints of abdominal pain. The result was unremarkable. Still the possibility of Kawasaki disease was entertained at this stage. Therefore, the patient went for another round of investigations which revealed the followings. Repeated ESR and CRP were trending up, ferritin level was 1385 microg/L (high), AST was 60 IU/L (high), ALT was 38 IU/L (normal) and albumin "trending down from 36g/L to 23g/L. Urinalysis showed no pyuria. US abdomen was repeated to rule out hydrups of the gall bladder which was absent.

By the end of the fifth day of admission the patient was started on IVIG and high dose aspirin for the suspicion of atypical Kawasaki disease based on the following criteria: Fever for 8 days, cervical lymphadenopathy 4 x4 cm, oro-pharyngeal changes with red cracked lips and strawberry tongue, bilateral non purulent conjunctivitis sparing the limbus, high ESR/CRP, leukocytosis, anaemia, high liver enzymes, high ferritin and low albumin.

The patient became afebrile within 24h with complete resolution of the tachycardia; and reductions in the neck mass size and tenderness were also apparent. A cardiac ECHO was obtained to rule out coronary aneurysm and to evaluate the heart murmur which was normal. Antibiotic was stopped at this stage.

The patient was discharged on the ninth day after complete resolution of all of his symptoms. Laboratory investigations before discharge revealed the followings: WBC count, 8000/mm³; platelet count, 945,000/mm³; CRP concentration, 165 mg/L; erythrocyte sedimentation rate, 62 mm/hr., Epstein-Barr virus (EBV) antibody titers negative. Viral screening including influenza, para influenza and adenovirus were negative. The blood /urine and throat swab culture were negative.

3 days after the discharge the patient presented to the emergency department of picture of urethritis and was reassured and sent home.

2 weeks after discharge he presented to our outpatient department for follow up and he was found to be well. Laboratory investigations revealed thrombocytosis with a platelet count of 616,000/mm³, ESR 109 mm/hr and CRP 7 mg /L. Aspirin dose was shifted from high dose to low dose after completing 14 days.

Discussion

The clinical manifestations of Kawasaki disease can be diverse and the diagnosis is based on demonstrating characteristic clinical signs and excluding other febrile diseases. However, a number of patients with Kawasaki disease do not fulfill the diagnostic criteria at onset but manifest symptoms several days later. Other group of patients may not meet all the classic diagnostic criteria. Children with Kawasaki disease exhibiting fever and fewer than four of the other characteristic symptoms are labeled as atypical or incomplete Kawasaki disease. Such patients are believed to be at higher risk of developing coronary artery aneurysms due to delayed or lack of therapy. Indeed, recent case reports have highlighted atypical cases of Kawasaki disease involving patients presenting with symptoms such as severe shock⁶ and pancreatitis.⁷ A recent case series has also highlighted the existence of Kawasaki disease in adults, indicating that age is not a defining characteristic of the disease.⁸ It is important that clinicians are made aware of atypical cases to facilitate prompt diagnosis and appropriate treatment. If the diagnosis is not made and treatment is not instituted, the patient may develop coronary artery disease and even sudden death secondary to myocardial infarction or rupture of coronary aneurysm.

In our case, a 5-year-old boy presented with fever and lymphadenitis. Although fever and lymphadenopathy are well-known symptoms of Kawasaki disease, the lack of other initial symptoms consistent with Kawasaki disease led us to consider other diagnoses, such as group A beta-hemolytic streptococcal infection or EBV infection. Although cervical lymphadenopathy is a major criterion for Kawasaki disease it is the least common and is present in only about 50% of cases, while the other clinical criteria occur in about 90%.⁹

According to a study by April et al.¹⁰ Kawasaki disease patients with cervical lymphadenopathy were older than those without lymphadenopathy. Stamos et al. reported a large series of 450 Kawasaki disease patients within a 10-year period. In this series, marked cervical lymphadenopathy was noted in 11 cases, and the median age was 5 years old.¹¹ 10 of 11 patients in that study were initially diagnosed as having bacterial lymphadenitis, but with poor response to antibiotics and negative drainage culture. The cervical mass was usually large, erythematous and tender to palpation but without fluctuation. Fluid aspiration was usually not available, or

only a scanty amount was present. Similar results were also reported by Kao et al.¹² Furthermore, Pontell et al. reported retropharyngeal abscess and peritonsillar abscess as initial presentations of atypical Kawasaki Disease.¹³

This case emphasizes that Kawasaki disease should be considered in children presenting with prolonged fever and cervical lymphadenitis, especially when there is a poor response to empiric antibiotic therapy. Careful and repeated examinations are necessary because the classical features of Kawasaki disease may only become evident over time.

Acknowledgments

None.

Conflicts of interests

The authors disclose no conflicts of interest.

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