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Recurrent Kawasaki disease presenting as acute airway obstruction

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ABSTRACT Kawasaki disease is a common paediatric vasculitide. It is usually diagnosed by its classical constellation of mucocutaneous signs. Recurrent Kawasaki disease is a rare phenomenon that occurs in approximately 3% of all patients diagnosed with Kawasaki disease. Its presentation is usually similar to the first episode of Kawasaki disease, and early diagnosis with prompt treatment is key in preventing associated cardiovascular morbidities. Recurrent Kawasaki disease is not well reported, and atypical presentations have not been previously reported in medical literature. Here, we report the case of a young girl with recurrent Kawasaki disease who presented atypically with acute airway obstruction secondary to retropharyngeal phlegmon.

Keywords: airway obstruction, atypical presentation, recurrent Kawasaki disease, retropharyngeal phlegmon
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INTRODUCTION

Kawasaki disease, or mucocutaneous lymph node syndrome, is a common childhood vasculitide. First observed and characterised in 1967 by Dr Tomisaku Kawasaki, it is prevalent in East Asia, with the highest incidence reported in Japan.[1,2] Recurrence is a rare phenomenon and occurs most commonly in children. In this case report, we present a case of recurrent Kawasaki disease in a four-year-old girl whose diagnosis was complicated by the presentation of acute airway obstruction.

CASE REPORT

In June 2008, a two-year-old Chinese girl presented with high fever for one week, associated with a nonproductive cough and poor feeding of the same duration. A full blood count was performed, revealing a slightly raised total white blood cell (WBC) count of 12.70 × 10^9/L (73% neutrophils, 18% lymphocytes, 9% monocytes), a normal haemoglobin level of 11.7 g/dL and a normal platelet count of 252,000/mm^3. Chest radiography showed mild bilateral bronchial wall thickening, but was otherwise unremarkable. An initial diagnosis of bronchiolitis was made and nebulised salbutamol was administered. However, her symptoms persisted. Two days later, she developed a generalised polymorphic rash associated with bilateral nonsuppurative conjunctivitis and palmar swelling. The injection site of the Bacillus Calmette-Guérin vaccine was noted to be erythematous and indurated. Cervical lymphadenopathy was also present. A repeat full blood count was performed, showing a markedly elevated total WBC count of 21.10 × 10^9/L (82% neutrophils, 2.74% lymphocytes, 0.63% monocytes). Both erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) level were elevated at 57 mm/50 mins and 314.0 mg/L, respectively. Other tests performed, such as renal function test, antistreptolysin O titre (ASOT), respiratory viruses antigens and aerobic blood culture, were either negative or normal.

In view of the clinical and biochemical findings, the diagnosis was changed to Kawasaki disease. Standard treatment with a single dose of 2 g/kg of intravenous immunoglobulin (IVIg) and high-dose aspirin was initiated. She responded positively to IVIg treatment and was discharged well ten days after admission. On cardiology outpatient follow-up, two-dimensional (2D) echocardiography showed mildly dilated right (RCA) and left coronary arteries (LCA), which mainly involved the left anterior descending artery and was associated with mild mitral regurgitation. The patient was otherwise clinically well and subsequently defaulted on follow-up.

Two years later, at four years and eight months of age, the patient presented with a massive left-sided neck swelling for five days, which was associated with respiratory distress and a high fever of Tmax 40°C. On examination, she was irritable and had an inspiratory stridor on crying. The patient also had a left torticollis secondary to the firm and tender left neck swelling, which measured 8 cm × 8 cm. The overlying skin was warm and erythematous. Apart from the massive cervical mass, there were no oral lesions or rash. On the day of admission, all blood investigations were normal, except for markedly raised ESR (114 mm/50 mins) and CRP (234.9 mg/L) level.

Cervical radiography revealed a large soft tissue swelling around the left submandibular region with a thickened epiglottis and a significantly widened prevertebral space (Fig. 1). Computed tomography of the neck was performed, showing multiple cervical lymphadenopathies and extensive retropharyngeal hypodensity suggestive of an underlying retropharyngeal phlegmon (Fig. 2). Tests for Epstein-Barr virus, cytomegalovirus and Bartonella infection were negative. Autoimmune workup for antinuclear antibodies and

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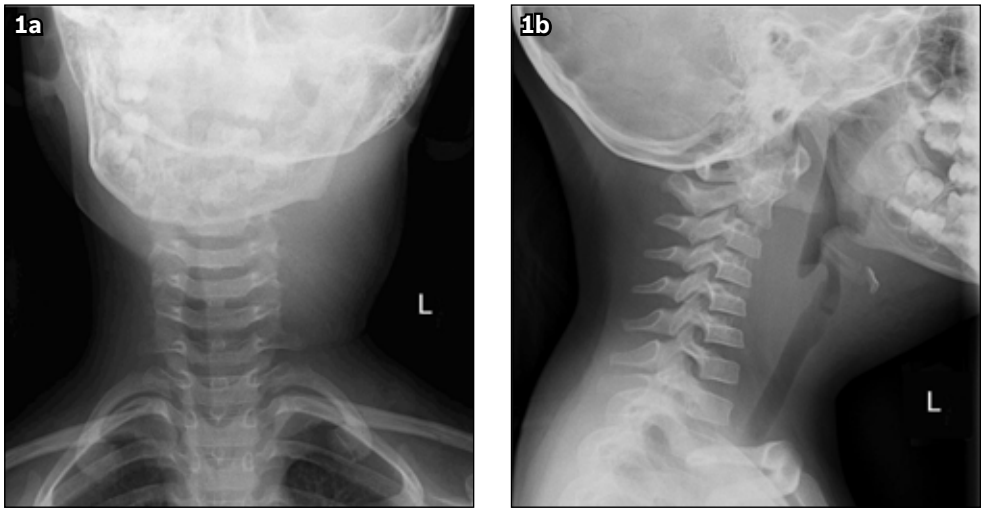

Fig. 1 Cervical spine radiography in the (a) anteroposterior view shows a large soft tissue swelling around the left submandibular region; and (b) lateral view shows a markedly widened prevertebral space extending from the C2 to C6 level, suggesting retropharyngeal soft tissue thickening.

antineutrophil cytoplasmic antibodies also yielded negative results. Additionally, urinalysis, urine culture, renal function test, liver function test, ASOT and blood cultures were all either negative or normal.

The patient's acute airway obstruction prompted otolaryngological involvement, and drainage of the retropharyngeal phlegmon was considered. However, her stridor began to resolve over the next day. In view of the clinical improvement, unusual presentation and unconfirmed aetiology, surgical intervention was held off and the patient was conservatively managed with close monitoring of any airway compromise. In the subsequent days of hospitalisation, her fever persisted and she started to develop bilateral conjunctivitis, red lips, oropharyngeal injection and a polymorphous rash. The clinical picture was similar to that of her first admission. As such, a diagnosis of recurrent Kawasaki disease was made and she was started on a single dose of 2 g/kg IVIg and high-dose aspirin, following which her symptoms improved significantly. The massive cervical lymphadenopathy and retropharyngeal phlegmon also completely resolved. On 2D echocardiography, both the RCA and LCA were found to be more severely dilated than during her first attack of Kawasaki disease. However, on subsequent follow-ups, the coronary aneurysms resolved and she remained well.

DISCUSSION

A clinical diagnosis of Kawasaki disease is made based on the presence of fever for more than five days and associated with at least four of the following: (a) polymorphic exanthem; (b) changes in the oropharynx such as injection, fissured lips and 'strawberry tongue'; (c) changes in the extremities such as swelling, erythema and desquamation; (d) bilateral nonsuppurative conjunctivitis; and (e) cervical lymphadenopathy > 1.5 cm.[3] The underlying pathological process in Kawasaki disease is systemic vasculitis, although the exact aetiology of this disease remains unclear.[4]

Recurrence is an interesting feature of Kawasaki disease. Rare but not unheard of, it is generally regarded as a new episode after at least three months from the initial episode.[5] The prevalence of recurrent Kawasaki disease is reported to be highest in Japan at 3%.[6] Maddox et al noted that the risk of recurrence was highest in children of Asian descent.[7] Several studies have attempted to elucidate the risk factors for recurrence, including cardiac sequelae at the initial onset, young age at first attack (1–2 years old) and use of IVIg at first attack.[8-10] Our patient had all of the features described. In another study, male gender and cardiac sequelae at the initial onset were identified as risk factors for sequelae at the second episode of Kawasaki disease.[9]

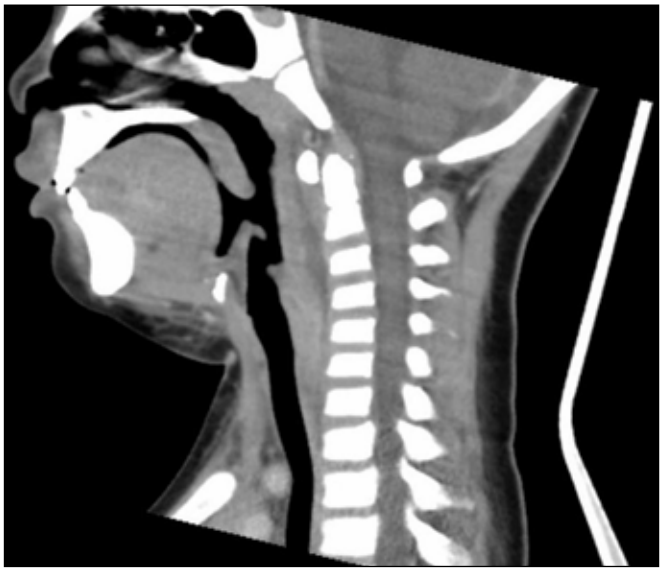

Fig. 2 Computed tomography of the neck shows area of hypodensity with soft tissue oedema seen from the level of C1 to C7, which is suggestive of an underlying retropharyngeal phlegmon.

The most feared complication of Kawasaki disease is cardiovascular sequelae. Coronary artery dilatation has been shown to happen in about 20%–30% of untreated patients.[4] The resultant thrombosis, myocardial infarction and/or dysrhythmia may cause death. Previous studies have shown that cardiac complications, particularly the formation of giant coronary aneurysms, are more likely to be observed among recurrent

cases.[7,11] Our patient already had cardiac sequelae during the first episode but suffered more severe cardiac involvement during the second attack. Apart from the higher risk of cardiovascular ill-effects during recurrence, incomplete lesions, atypical presentation, shorter period of fever and decreased response to IVIg are notable features of recurrent Kawasaki disease.[12]

In our patient, the second diagnosis was delayed, as she presented atypically with airway obstruction secondary to massive cervical lymphadenopathy and retropharyngeal phlegmon, instead of the characteristic skin and oral lesions seen in Kawasaki disease. Her acute airway obstruction prompted otolaryngological intervention and the proposal of surgical drainage. The latter was ultimately held off in view of the unusual clinical picture. Kawasaki disease presenting as massive cervical lymphadenopathy and retropharyngeal phlegmon without its characteristic skin and oral presentations is a rare phenomenon. Ganesh et al reported a case of Kawasaki disease presenting with retropharyngeal abscess in which drainage yielded low therapeutic success.[13] To the best of our knowledge, no such presentation has been reported in cases of recurrent Kawasaki disease. In our patient, massive cervical lymphadenopathy and retropharyngeal phlegmon resolved completely upon targeted treatment of her recurrent Kawasaki disease with a standard dose of IVIg. This proves that the decision to withhold surgical intervention was important, especially when the underlying pathological process had yet to be elucidated.

Although uncommon, recurrence of Kawasaki disease is definitely possible. When it does occur, the presentation may be atypical, such as with retropharyngeal abscess and massive unilateral lymphadenopathy, which will further add to the diagnostic challenge. However, in view of the increased risk and severity of cardiovascular morbidity and mortality associated with recurrent Kawasaki disease, prompt diagnosis and early treatment are of paramount importance. Surgeons should remain alert to atypical features of this disease and consider withholding surgical intervention in cases where the underlying aetiology has yet to be determined, as demonstrated in our patient.

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