Journal of Microbiology, Immunology and Infection (2014) 47, 152—157

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CASE REPORT

Unusual manifestations of Kawasaki disease with retropharyngeal edema and shock syndrome in a Taiwanese child

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Received 14 September 2011; received in revised form 27 December 2011; accepted 20 January 2012
Available online 12 April 2012

KEYWORDS
Kawasaki disease;
Retropharyngeal edema;
Shock

We report a 3-year-old girl with Kawasaki disease who presented with retropharyngeal edema and shock syndrome. This is the first reported case in Taiwan. The patient initially presented with fever, cough, and pyuria followed by rapidly progressive enlarged bilateral cervical lymphadenopathy. On the third day of the fever, computed tomography for airway compression sign found widening of the retropharyngeal space mimicking a retropharyngeal abscess. Later, an endotracheal tube was inserted for respiratory distress. A skin rash over her trunk was also noted. On the fifth day of the fever, the clinical course progressed to hypotension and shock syndrome. Because of more swelling of bilateral neck lymph nodes, computed tomography was arranged again and revealed partial resolution of the edematous changes in the retropharyngeal space. Edema of the hands and feet, bilateral bulbar conjunctivitis, and fissured lips were subsequently found. The diagnosis of Kawasaki disease was confirmed on the eighth day of fever. There was no evidence of bacterial infection. She was administered intravenous immunoglobulin (2 mg/kg) and high dose aspirin (100 mg/kg/day). One day later, the fever subsided, and her blood pressure gradually became stable. Heart echocardiography on the Day 13 revealed dilated left coronary artery and mitral regurgitation. Follow-up echocardiography six months later showed normal coronary arteries. To date, the patient has not experienced any complications. This case illustrates that retropharyngeal edema and shock syndrome can be present in the same clinical course of Kawasaki disease. Clinicians and those who work in intensive care units should be aware of unusual presentations of Kawasaki disease to decrease rates of cardiovascular complications.

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1684-1182/$36
doi:10.1016/j.jmii.2012.01.017

Introduction

Kawasaki disease (KD), also known as acute febrile mucocutaneous lymph node syndrome, was first described in the Japanese literature by Dr Tomisaku Kawasaki in 1967, and in the English literature in 1974.[1] This disease is a systemic vasculitis involving all blood vessels, but predominantly medium-sized arteries and is one of the most common vasculitides of childhood.[2] However, it may cause severe complications, morbidity, and even mortality if not appropriately treated. Eighty-five percent of affected patients are younger than 5 years old,[3] and it is more common in boys than in girls (male to female ratio, 1.36:1 to 1.62:1).[3–5] It occurs worldwide in all ethnic groups, and Asian populations have a higher risk than other ethnic groups.

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.[6] When involving the cardiovascular system, patients may present with myocarditis, pericarditis, coronary artery aneurysm, and aortic root dilatation.[3]

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.[7–11] Progressing into shock is uncommon, although some studies have reported patients with KD and shock syndrome.[12–14] In this paper, we review the literature and report the case of a Taiwanese child with KD who had retropharyngeal edema with bilateral cervical adenopathy resulting in airway compression, and also shock syndrome.

Case report

A 3-year-old girl presented with cough for 2 weeks and fever up to 40°C for 2 days. Physical examination found injected throat, coarse breath sounds with no palpable lymph nodes, no audible heart murmur, and no skin rash. Initial laboratory data revealed pyuria, white blood cell count of 39.6 x10^9/L, and an elevated CRP level of 197 mg/L. Empiric antibiotics with ampicillin and gentamicin were prescribed; however, she had bilateral progressive swelling of the lymph nodes in her neck on the following day. On the third day of the fever, in addition to her progressive neck swelling (Fig. 1A in comparison to Fig. 1B, her usual appearance), she had torticollis, respiratory distress, and a skin rash over her trunk. A lateral view X-ray of the neck found retropharyngeal space widening. CT was performed to localize the lesion causing airway compression, and showed retropharyngeal edema, a suspected retropharyngeal abscess, and enlarged lymph nodes (the left was largest at: 2.5 × 3 cm) along bilateral internal jugular veins, from the submandibular triangle to the level of cricoid cartilage (Fig. 2A, B). The empiric antibiotics were shifted to oxacillin with cefotaxime and metronidazole. Due to the suspicion of a retropharyngeal abscess, surgical exploration was performed with open drainage for tissue proof. Intubation with mechanical ventilation was used after the operation to relieve the airway compression and was kept for persisted enlarged lymph nodes of the neck.

On the fifth day of the fever, tachycardia and hypotension (77/36 mmHg), cooling of extremities, and oliguria (0.47 ml/kg/h) were noted. After fluid resuscitation with normal saline (20 ml/kg), inotropic agents were given for sustained hypotension and shock syndrome. Dopamine was used on the same day (up to 20 μg/kg/min), and epinephrine was added the next day (up to 0.1 μg/kg/min). Laboratory data revealed a white blood cell count of 12.9 × 10^9/L (band form: 11%), platelets 294 × 10^9/L, D-dimer 6.24 mg/L, lactate dehydrogenase 203 IU/L, fibrinogen 6.54 g/L, prothrombin time 10.5 seconds, partial

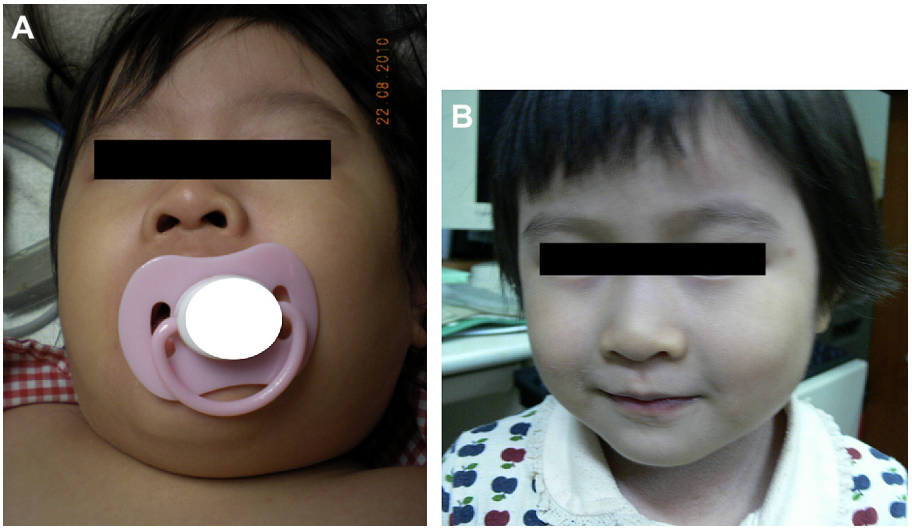

Figure 1. (A) On the third day of the illness, the patient had torticollis, respiratory distress and bilateral progressive swelling lymph nodes in her neck. (B) Patient's usual appearance before the illness.