Kawasaki Disease: Two Episodes of Recurrent Disease in a Greenlandic Inuit Boy

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Kawasaki disease is a vasculitis of medium-sized arteries presenting with mucocutaneous manifestations and lymphadenopathy (1, 2). Kawasaki's syndrome typically occurs in childhood, and is commonly confused with infectious exanthemas (3). This case report outlines the characteristics of the condition.

Patients typically present with fever and erythema of the lips and oral mucosa, rash, and changes in the extremities, including erythema of the palms and soles, bilateral non-exudative conjunctivitis and cervical lymphadenopathy. These classical features normally develop after a short course of respiratory or gastrointestinal symptoms (1).

Normally, Kawasaki disease is self-limiting (3). Often the fever and mucocutaneous manifestations resolve within 10–12 days without treatment (4). The major problem of the disease is the occurrence of cardiovascular complications, which may cause significant morbidity and mortality (5). Patients can develop aneurysms of the coronary arteries, reduced contractility of the myocardium, heart failure, cardiac arrhythmias, and occlusion of peripheral arteries (5).

Recurrence of Kawasaki disease with new episodes of fever, mucocutaneous manifestations and lymphadenopathy is rare (6, 7). We report here an Inuit boy in Greenland who experienced two episodes of Kawasaki disease.

Case report

A 10-month-old Greenlandic Inuit boy was admitted to the local hospital. He had been coughing and spiking fevers for a few days. Due to pulmonary crepitations he was started on oral antibiotics. The fever persisted, and he developed the classical erythema of the lips and oral mucosa (Fig. 1), rash, palm and sole erythema, bilateral non-exudative conjunctivitis (Figs 2 and 3) and cervical lymphadenopathy. He was treated with intravenous (IV) immunoglobulins and recovered. Echocardiography was performed and slight dilatation of the coronary arteries was seen. He started treatment with acetylsalicylic acid. After 2 months he was re-admitted with fever, general malaise, and had recurrence of all the classic symptoms of Kawasaki disease; erythema of the lips and oral mucosa, rash, changes

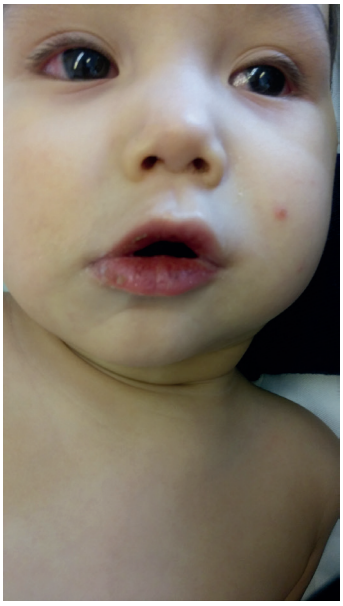

Fig. 1. Erythema of the lips and oral mucosa.

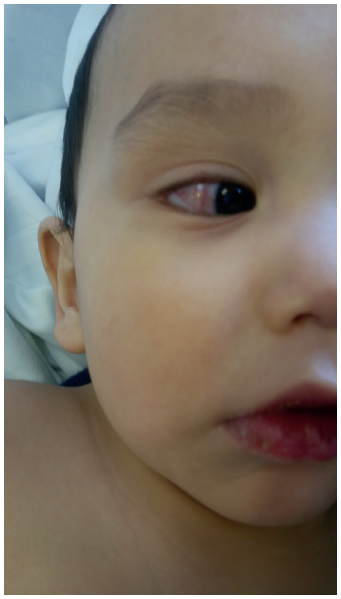

Fig. 2. Non-exudative conjunctivitis.

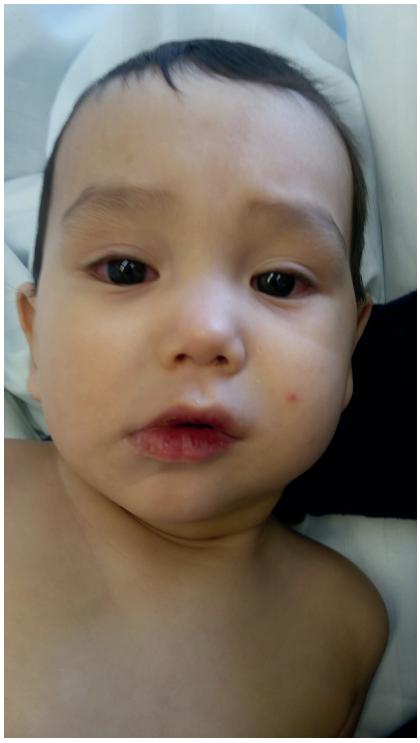

Fig. 3. Erythema of the lips and oral mucosa, slight rash, and bilateral non-exudative conjunctivitis.

in the extremities, bilateral non-exudative conjunctivitis and cervical lymphadenopathy. The same day he was treated with IV immunoglobulins, and shortly afterwards, all symptoms had disappeared. As recurrence of Kawasaki disease is rare, he was tested for immunological defects and other genetic diseases, but none were found. After 5 months, a new episode with fever and the classic features of Kawasaki disease occurred. Once again treatment with IV immunoglobulins was given, and he recovered quickly. As this was the second episode of recurrent Kawasaki disease, he again underwent investigation for immunological diseases, but all tests again were negative. A repeat echocardiography showed normal myocardial function, and no progression of the coronary artery dilatation. At follow-up one year after the initial episodes of Kawasaki disease, no further episodes had occurred.

Discussion

Fever is common during childhood due to infectious diseases. However, fever due to systemic inflammation is also one of the main features of Kawasaki disease (1). Kawasaki disease should be considered in children who have unexplained fever for more than 5 days (8).

Kawasaki disease can be diagnosed by the presence of typical clinical manifestations. A polymorphous rash is seen in 70–90% of cases (4). Oral mucous membrane symptoms in 90%, symptoms in the extremities in 50–85%, ocular symptoms in more than 75%, and cervical lymphadenopathy in 25–70% (1). This means that not all symptoms and clinical manifestations occur in all patients with Kawasaki disease; symptoms do not always come in the same order, and are not always present at the same time. Due to the very high occurrence of cutaneous rash and oral mucous symptoms, dermatologists are frequently consulted (3, 4).

The rash often appears in the early phase of the disease, typically as erythema in the region of the perineum, and desquamation. This is followed by morbilliform, targetoid or macular skin lesions on the torso and extremities (3). Kawasaki disease may initiate a psoriasiform eruption in children not previously diagnosed with psoriasis. Vesicles and bullae are normally not seen with Kawasaki disease. Patients may have redness and crust formation at the site of the Bacille Calmette Guerin (BCG) vaccination (3, 4). This is, of course, only relevant in countries, such as Greenland, where BCG vaccination is part of the childhood immunization programme.

Changes on the extremities occur in the final phase of the disease. Symptoms include indurated oedema on the dorsal side of the hands and feet, and diffuse erythema of the palms and soles. Recovery from Kawasaki disease is associated in 68–98% of children with sheet-like desquamation of the periungual hand and feet regions. Furthermore, linear nail creases occur, also called Beau's lines (1).

Oral mucous membrane symptoms include red, cracked lips and a strawberry tongue. The strawberry tongue is caused by sloughing of filiform papillae and denuding of the inflamed glossal tissue. Often these symptoms become more evident as Kawasaki disease progresses. Discrete oral lesions, such as ulcers, vesicles, or tonsillar exudate, are suggestive of conditions other than Kawasaki disease (1).

Arthritis is not part of the diagnostic criteria of Kawasaki disease, but occurs with either oligoarticular or polyarticular involvement in 7–25% of cases (1).

Cardiovascular symptoms and complications of Kawasaki disease, although not part of the diagnostic criteria, may cause severe morbidity and mortality. At the time of diagnosis, 30% of patients have dilatation of the coronary arteries.

It is important to highlight that there is no single laboratory test that can confirm or deny the diagnosis of Kawasaki disease. Kawasaki disease causes systemic inflammation, and increases in C-reactive protein and erythrocyte sedimentation rates are frequently seen, as well leukocytosis and thrombocytosis (2).

The aetiology of Kawasaki disease is unknown (9). It is thought to be caused by an overreaction of the immune system following a mild infection (1). The disease is more common in Asian people, and the incidence of Kawasaki disease is 8–10 times higher in Japan compared with Northern America (6, 7, 9). The incidence of Kawasaki disease among Inuits is unknown.

Recurrence of Kawasaki disease is rare and occurs in only 1–4% of cases (6, 7, 10–12). An incomplete immune response is thought to be the primary reason for recurrence of Kawasaki disease (6, 11). In the current case, the patient experienced two episodes of recurrent disease, which is extremely rare. In all 3 episodes, the patient presented with a rash, oral mucocutaneous symptoms, bilateral conjunctivitis and cervical lymphadenopathy. Hence, the diagnosis of Kawasaki disease is very likely. Due to the frequent recurrences, the patient was tested for immunological disease and genetic defects, but none were found.

Treatment of Kawasaki disease consists of administration of intravenous immunoglobulins (5). Immunoglobulins reduce the incidence of cardiovascular complications and coronary aneurysms five-fold when given within the first 10 days of disease, compared with no administration of immunoglobulins (5). Hence, early diagnosis is important to allow for early treatment and reduction of cardiovascular complications.

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