Case Report

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Kawasaki Disease in an Adult Woman from Iran

Saeid Rezaei-Jouzdani[1], Bahram Pakzad[2]

1- Department of Internal Medicine, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran
2- Associate Professor, Department of Internal Medicine, School of Medicine, Isfahan University of Medical Sciences, Isfahan, Iran

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Corresponding author:
Saeid Rezaei-Jouzdani

Email:
s.rezaei.jouzdani@gmail.com

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ABSTRACT

Kawasaki disease is a rheumatologic disorder that is due to small- to medium-sized vessels necrotizing vasculitis with unknown origin. The disease affects children predominantly, but rarely occurs in adults. We report a case of 23-year-old woman from Iran who had got high grade fever, bilateral non-purulent conjunctivitis, myalgia, palms and soles exanthema, and desquamation. After 5 days of empirical antibiotic therapy without any organisms identified for source of infection, the diagnosis was established. No treatment with intravenous gammaglobulin and aspirin was administered and her echocardiogram showed normal coronary arteries. The age of our patient is an unusual aspect of the disease presentation in Iran. The diagnosis of Kawasaki in adults should be considered in patients with prolonged fever not responding to antibiotics with clinical features of Kawasaki disease.

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Introduction

Kawasaki disease is an acute necrotizing vasculitis of small- to medium-sized vessels affecting predominantly children younger than 5 years old; but it occurs rarely in adults (1). Despite its rarity and broad differential diagnosis, it should be considered in adults with clinical features of the disease as previous studies from other regions reported typical and atypical manifestations of Kawasaki disease in adults (2). In this case study, we report an adult Iranian woman who lives in Iran with acute Kawasaki disease presented with typical clinical features; the diagnosis was set lately.

Case Report

A 23-year-old White Iranian woman who

lives in a countryside of Chaharmahal was referred to Alzahrah hospital, a main referring medical center in Isfahan City in the center of Iran because of prolonged fever and arthralgia began 1 week before admission in March 2017. She was healthy before and did not have any rheumatic or familial disease in her family or contact with any others with similar features. Her drug history and substance abuse was unremarkable before the presentation.

She reported a 1 week of fever with arthralgia and general malaise. During several outpatient visits, oral and parenteral antibiotics such as coamoxiclavunate, ampicillin, and azithromycin were prescribed for her; but the fever did not abate and general condition deteriorated. On physical examination, the patient's temperature was 39.7 °C. She had bilateral non-purulent conjunctivitis and strawberry tongue (Figures 1 and 2).

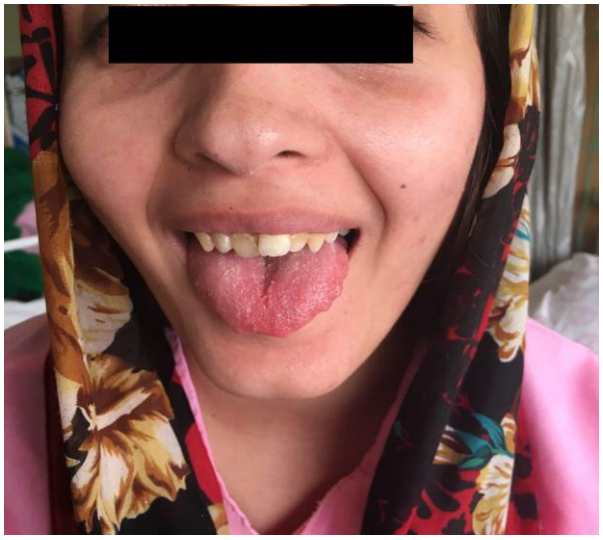

Figure 1. Strawberry tongue after 1 week from onset of presentation

She had hyperemia and desquamation on both palms, but only hyperemia on soles. After 10 days from onset of fever, desquamation of soles began (Figures 3 and 4). The patient did not have any lymphadenopathy in the first day of admission; but after 5 days, bilateral cervical lymphadenopathy in 1.5 × 1.5 cm size was palpable.

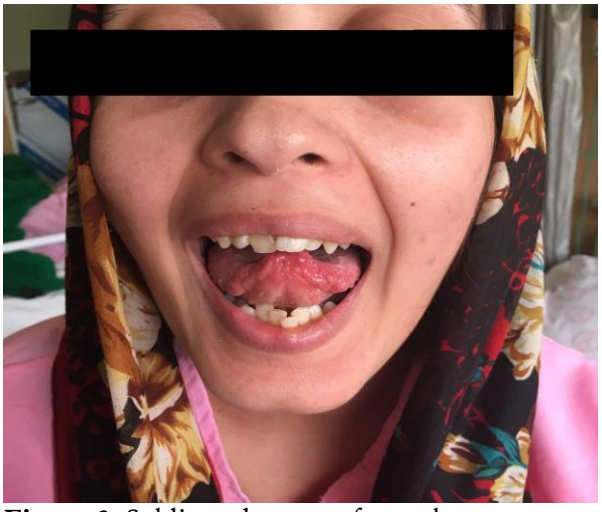

Figure 2. Sublingual aspect of strawberry tongue

There was not any positive findings in her physical exams, and chest imaging and abdominal sonography was unremarkable. During hospital admission, she complained of general malaise and arthralgia.

Figure 3. Lamellar scaling and desquamation of lower limbs

The diagnosis of viral infections was suspected and laboratory tests including cell blood count, procalcitonine, Wright, liver function tests, and viral serology for parvovirus, Epstein-Bar virus, hepatitis A, B and C, and human immunodeficiency virus (HIV) were performed. All viral serology tests were reported normal.

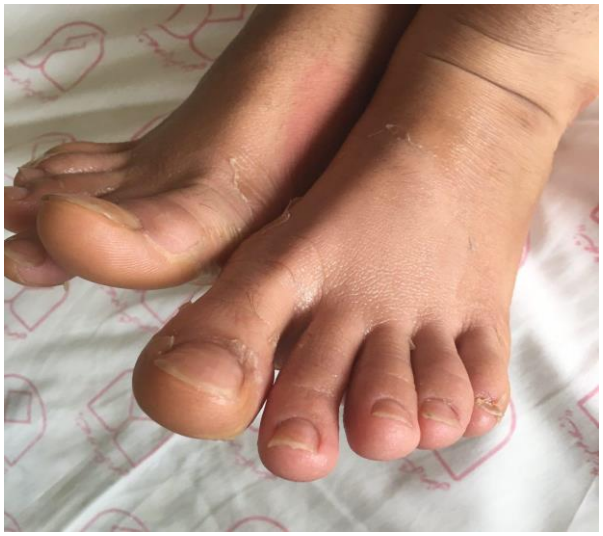

Figure 4. Dorsum of the feet with lamellar scaling

An echocardiogram was performed and was found to be normal without any abnormality in the coronary arteries. Bone marrow biopsy was normal and cervical lymphadenopathy abated in 1 week.

As procalcitonine was low (0.1 µg/l), erythrocyte sedimentation rate (ESR) was elevated (87 mm/hour), fever did not abate with meropenem, and all blood, urine, and lumbar puncture analysis and cultures were negative, the antibiotic was discontinued after 5 days from admission to our hospital. All other medications discontinued at the day 14, when fever abated, and ESR declined; the diagnosis of Kawasaki was set. The patient discharged from hospital with scheduled follow-up visits and echocardiogram in 2 weeks apart. The patient did not receive aspirin and immunoglobulin because of delay of the diagnosis; but follow-up echocardiograms showed no coronary aneurysm.

Discussion

Kontopoulou et al. had described Kawasaki disease in adults with geographic distribution (2). According to their review, only 100 cases of adult Kawasaki disease had been described until 2015, and never reported in the literature from Iran, yet. There is no specific test for its diagnosis; and due to its rarity, diagnostic criteria in adults has not been established. The diagnostic criteria for Kawasaki disease was reported since 1967 for children (3). It is defined complete when 4 of the 5 principal features are fulfilled despite of at least 5 days of fever and is defined incomplete when clinical features do not fulfill the criteria but based on coronary abnormalities, the diagnosis is established (4) (Table 1).

Table 1. Diagnostic criteria for Kawasaki disease

Fever of 5 days and presence of 4 principal features:
Changes on the extremities: in the acute phase, erythema of palms and soles, and edema of hands and feet; in the subacute phase, desquamation of fingers and toes
Polymorphous exanthema
Bilateral bulbar conjunctival injection
Changes of the lips and oral mucosa: erythematous and cracked lips, strawberry tongue, and oral and pharyngeal hyperemia
Cervical lymphadenopathy (diameter of more than 1.5 cm)

Complete Kawasaki disease is and unusual disease in adults (1). In a report of 10 cases by Gomard-Mennesson et al., the clinical and laboratory features of complete and incomplete Kawasaki disease in adults were described (5); similar to this report, our patient fulfilled the criteria for the complete or typical features. In the differential diagnosis, we considered other diseases such as bacterial and viral infections and other rheumatologic and immunologic disorders; however, negativity of serologic tests and course of the disease ruled out these possibilities. The accuracy of diagnosis is lower in adults, according to a recent report, leading to coronary complications in long-term delay in diagnosis, and absence of gammaglobulin treatment (1). Despite the lack of proper treatment in most adults, Kawasaki disease coronary aneurysms is less frequent (8 percent) than children (25-30 percent) due to unknown reason (6).

The recommended treatment for Kawasaki disease in adults is intravenous gammaglobulin administration before 10^{th} day from onset of fever, without any beneficial effect after resolution of the fever (3). The

diagnosis was established in our patient as in most of the reported cases (7) after resolution of the fever. This is the first report of Kawasaki disease in adults in Iran and among Persian population. It is imperative to consider the possibility of the disease in this region.

The aim of this study was to report a rare case of Kawasaki disease in adulthood from Iran. The importance of early diagnosis according to the clinical manifestation of the disease is to initiate appropriate treatment earlier to prevent major complications. Because of the broad differential diagnosis for this disease in adults, and absence of a specific test, it should be kept in mind to make early diagnosis and proper treatment.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

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