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A Child with incomplete Kawasaki disease and erythema marginatum

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Abstract

Incomplete or atypical Kawasaki disease (KD) may delay or obscure the diagnosis of this disease. In the absence of full classical presentations, laboratory signs of KD and also criteria of coronary arteries involvement can help toward correct diagnosis and starting appropriate therapy. Erythema marginatum is a rare skin rash that predominantly seen in acute rheumatic fever. We report a 6-year-old child with fever and unilateral cervical lymphadenopathy and transient erythema marginatum, with diagnosis of incomplete KD, treatment with intravenous immunoglobulin (IVIG) started and had dramatic response to it.

Introduction

Kawasaki disease (KD) is an acute vasculitis that predominantly affects pediatric patients, chiefly infants. The illness is characterized by fever for five days or more and four or more of the following features: 1) bilateral conjunctivitis 2) changes in extremities 3) exanthems 4) cervical lymphadenopathy and 5) changes in lips and oral cavity. For the diagnosis of incomplete KD, patients should have fever for 5 days or more and less than three clinical features without alternative explanation for the diagnosis (2). Rashes in KD are polymorphous varying from macular to maculopapular and morbilliform, however it is never vesicular (3). Erythema marginatum is a rare skin rash and is a major criterion for the diagnosing of acute rheumatic fever (4). In the literature review erythema marginatum as a cutaneous manifestation of KD has been reported only in one case (5). We report here a 6-year-old child with incomplete KD, as having unexplained fever for more than five days and unilateral cervical lymphadenopathy and transient erythema multiform whom with diagnosis of incomplete KD, had dramatic response to therapy with intravenous immunoglobulin (IVIG) and high dose aspirin.

Key point

Erythema marginatum could be an early cutaneous manifestation of Kawasaki disease, particularly in incomplete form

Case Presentation

A 6-year-old boy admitted to our hospital because of fever (up to 40°C axillary) and left sided neck swelling from three days ago. At that time, child also had anorexia and night sweating. In physical examination a tender, erythematous, mobile neck mass with about 3×5 cm in size palpated in left neck site. The mass was painful, since child could not bend his neck to opposite site. Neck sonography revealed cellulitis and inflammatory lymph node in the left neck. Past medical history was unremarkable. In the second day of admission, the skin rashes as annular macules with pale center and pink to faintly red irregular–serpiginous border with different sizes, appeared at first in trunk then in last days progressed to face and extremities. These rashes were non-pruritic and non-tender (Figure 1A and 1B). Initial laboratory evaluation showed leukocytosis, thrombocytosis, hypoalbuminemia and evidence of systemic inflammation as marked elevation of erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) levels (Table 1).

Abdominal sonography showed absence of any organomegaly or hydrops of gall bladder. In cardiac evaluation electrocardiogram was normal and echocardiogram did not show

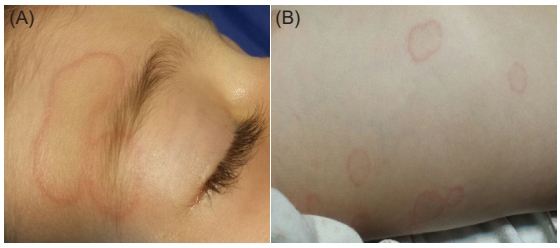

Figure 1. (A) The presence of erythema marginatum above right eyebrow of the presented patients. **(B)** The presence of erythema marginatum on the abdomen of the presented patients.

Table 1. Laboratory data of the presented patient

Variable	Value (unit)
WBC	22000 (/µL)
Neutrophils	55 (%)
Lymphocytes	40 (%)
Platelet count	671 000 (/µL)
Hemoglobin	10.2 (g/dL)
ESR	102 (mm/h)
CRP	36 (mg/L)
Albumin	2.3 (g/dL)
Sodium	135 (mEq/L)
AST	30 (U/L)
ALT	20 (U/L)

WBC: White blood cell count; ESR: Erythrocyte sedimentation rate; CRP: C-reactive protein; AST: Aspartate aminotransferase; ALT: Alanine aminotransferase

any coronary arteries involvement. Annular marginally erythematous skin rashes which at first appeared in trunk gradually progressed to involve face and extremities. The rashes accentuated with warmth, such as application of warm towels or a hot bath. Despite antibiotic therapy (intravenous clindamycin and ceftriaxone), the child persisted with high and remittent fever and irritability.

In 5th day of admission diagnosis of incomplete and atypical KD was suggested, according to the persistence of fever at least 8 days and two classic criteria (unilateral cervical lymphadenopathy and skin rashes as erythema marginatum) and laboratory findings as increased levels of ESR and CRP, leukocytosis, thrombocytosis, anemia and hypoalbuminemia and also unresponsive to antibiotics with exclusion of other possible diseases. Therefore the antibiotics discontinued and intravenous immunoglobulin (2 g/kg) and high dose aspirin started. Fortunately in the first day after immunoglobulin administration, clinical condition of the patient improved resolution of irritability. Skin rashes progressively faded while on the 7th day of admission rashes disappeared. Additionally, neck mass progressively reduced in size since in 5th day after immunoglobulin administration its size was unremarkable.

On the 11th day of admission the repeated cardiac evaluation with EKG and echocardiogram were normal. The fever also subsided and the patient discharged. In outpatient follow up 8 weeks after discharge ESR and CRP levels and CBC counts and echocardiographic evaluation of coronary arteries all were normal, therefore the low-dose aspirin discontinued. Cardiology follow up with echocardiographic evaluation of coronary arteries in the next six months were normal.

Discussion

KD is an acute febrile systemic vasculitis that was first described by Kawasaki et al in 1974 (6). Diagnostic criteria of KD are fever which persisting at least five days and the presence of at least four principal features such as changes in extremities (erythema and edema of hands and feet), polymorphous exanthema, bilateral bulbar conjunctival injection without exudates, erythema of lips and oral cavity, and cervical lymphadenopathy (>1.5 cm in diameter) usually unilateral (2). Some patients with suspected KD do not fulfill the diagnostic criteria, and thereby the diagnosis is made based on coronary artery abnormalities, laboratory findings and exclusion of other diseases. These cases would be so-called incomplete KD (7). Cutaneous eruptions are present in 80%-90% of patients early in the disease course. These eruptions include polymorphous erythematous, usually diffuse eruptions that may be macular, scarlatiniform, erythema multiforme–like, morbilliform or characterized by scaling plaques. Uncommonly these eruptions present as an urticarial exanthema, erythematous or rarely as a micropustular eruption, but it is rarely vesicular or bolus. They have been described as non-pruritic, which may or may not be tender. In almost all cases, eruptions occur on the trunk and extremities, often with perineal accentuation (3). Erythema marginatum is a diagnostic criterion of rheumatic fever, seen in less than 10% of cases. The annular eruption is macular or slightly raised, with a pink or red border and a paler center. Lesions are asymptomatic, transient and migratory (4).

Our presenting patient had incomplete KD, because he had unexplained fever for more than 5 days in addition to only two signs (unilateral cervical lymphadenopathy and skin rash as the erythema marginatum). Also patient was unresponsive to antibiotic therapy, but he has a dramatic response to IVIG therapy with improvement and well-being, which supports our diagnosis. Our case is interesting because association of erythema marginatum with KD is very rare and only one case reported in literature. The patient was a 7-year-old boy with classic KD and erythema marginatum; also he had bilateral pleural effusion (5). Erythema marginatum is characteristic of rheumatic fever and occurs in less than 5% of patients. Fortunately in our case coronary arteries were normal. While the injury to the coronary arteries could be detected in later stages of KD, therefore early treatment with IVIG could cure illness and prevent coronary arteries involvement.

Conclusion

Our case showed that erythema marginatum could be an early cutaneous manifestation of KD, particularly in incomplete form. Early treatment with IVIG could cure patient and prevents possible coronary arteries involvement in later stages of KD.

Conflicts of interest

There is no conflict of interest in this study.

Ethical considerations

Ethical issues including plagiarism, double publication, and redundancy have been completely observed by the author. The patient gave the consent to publish as a case report.

Author's contribution

MR is the single author of the manuscript

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