

Case Reports

Atypical Dermatological Presentation of Multisystem Inflammatory Syndrome in a Child with Polyarticular JIA: A Case Report

Kazi Iman¹, Laboni Akter², Kamrul Laila³, Mohammad Imnul Islam⁴,
Shahana Akhter Rahman⁴

Abstract

Multisystem inflammatory syndrome in children (MIS-C) is an inflammatory process that occurs after infection with SARS-CoV-2 virus. In this syndrome children usually present with fever, rash, abdominal pain, diarrhea or vomiting, hypotension and shock. Children may need urgent care if heart, blood vessels or other vital organs are involved. Poly-articular Juvenile Idiopathic arthritis is a subset of JIA defined by presence of arthritis of five or more joints within 6 months of disease onset along with other criteria of JIA. In this case report we report mucocutaneous manifestation of MIS-C in a child with poly-articular rheumatoid factor (RF) negative JIA case.

Key words: MIS-C, JIA, children.

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Introduction

Multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19 has many similarities to Kawasaki disease (KD), macrophage activation syndrome (MAS), and toxic shock syndrome.^{1,2} There are currently about 1000 cases of MIS-C documented worldwide.³ To help clinicians diagnosing the condition, the CDC and the WHO have released their own MIS-C diagnostic criteria. MIS-C patients may present with fever, rash, hypotension or shock, diarrhea and vomiting, abdominal pain, myocardial dysfunction. Along with these clinical findings, inflammatory markers like ESR, CRP or pro-calcitonin are very high and usually there are evidences of coagulopathy. MIS-C is an immune mediated secondary response to corona virus, evidenced by presence of COVID antibodies in the majority of cases.⁴⁻¹¹ Treatment option is intravenous immunoglobulin along with or without systemic steroids.

Juvenile idiopathic arthritis is defined as presence of arthritis in children below 16 years and the disease duration is at least six weeks after exclusion of other identifiable causes¹² According to The International

League of Associations for Rheumatology (ILAR) RF negative poly articular JIA is one of the sub type of JIA.¹² In this subtype arthritis occurs in more than four joints and both small and large joints are usually involved. According to ACR guideline treatments are NSAIDs along with DMARDs. This case was reported to increase the awareness of the symptoms of MIS-C in patients' with rheumatic disorders, with a focus on the difficulties to differentiate them.

Case report:

"K" a 10-year old boy, 1st issue of consanguineous parents hailing from Habiganj, was admitted in the Paediatric Rheumatology Division of Bangabandhu Sheikh Mujib Medical University (BSMMU) on 7th November 2021, with the complaints of swelling of right forearm and both ankle and rash on sole of both feet for 7 days. He had history of contact with COVID-19 positive case. He was a diagnosed case of RF negative poly-articular JIA since 2019. With adequate treatment he was maintaining clinical remission on medication for last one year. On examination we found him ill looking, febrile, vitally stable. Skin survey showed extensive erythematous cutaneous rash over both the soles (Fig-1). Locomotor system examination revealed arthritis over three joints (right forearm, both ankles). Tenderness grade was 2 over 4. Movement of all the affected joints were restricted. His gait was antalgic. Other systemic examinations were normal.

1. Assistant Professor, Dr. M R Khan Shishu Hospital & ICH

2. Junior Consultant, Dhaka Medical College & Hospital

3. Assistant Professor, Department of Pediatrics, BSMMU

4. Professor, Department of Pediatrics, BSMMU

Correspondence: Dr. Kazi Iman, Assistant Professor, Dr. M R Khan Shishu Hospital & ICH. Cell no: 01711317493, E-mail: kaziiman28@gmail.com

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Fig-1: Patient “K”



Fig-2: Erythema on sole

Relevant investigations were done which are showed in Table-I

Table-I:
Investigation Profile of the Patient:

Test name	Result	Normal range
Haemoglobin	9.7g/dl	15±2 gm/dl
ESR	100 mm/1 st hr	0-10 mm/1 st hr
WBC	39 X 10 ⁹ /L	7.0 ± 3.0 X 10 ⁹ /L
Neutrophils	81 %	40-80 %
Lymphocytes	14 %	20-40 %
Platelets	560 X 10 ⁹ /L	150-450 X 10 ⁹ /L
hCRP	>200.00 mg/l	<5 mg/l
Blood culture	No growth	
Urine culture	No growth	
SGPT	15 U/L	10-49 U/L
Creatinine	0.49 mg/dl	0.5-1.11 mg/dl
D-Dimer	>4.5 µg/ml	0.00-0.50 µg/ml
Troponin-I	>0.002 µg/ml	0.0452 µg/ml
CK-MB	0.18 ng/ml	0-5 ng/ml
Pro-calcitonin	0.11 µg/mL	<0.05 µg/ML
LDH	50 U/L	10-49 U/L
PT	13.9 sec	12-16.5 sec
INR	1.17	
APTT	28.8 sec	24-36.0 sec
IL-6	198.4 pg/ml	<7.5 pg/ml
Ferritin	324.3 ng/ml	7-140 ng/ml
COVID-19 Ig Antibody (ELISA)	3.9	<1.1

For cardiac evaluation we did ECG which was normal and colour doppler echo cardiograph showed normal cardiac anatomy with good bi-ventricular function.

On the basis of history, clinical examination findings and lab evidences, the boy was diagnosed as a case of MIS-C with RF-negative poly-articular JIA. “K” was treated with IVIG (2g/kg over 24 hour) followed by oral prednisolone and other supportive treatment. After complete recovery patient was discharged with the advice to follow up after two weeks. We followed up the patient and his clinical and laboratory parameters were normal.

Discussion:

The clinical and laboratory profile of MIS-C may be confused with different inflammatory diseases leading to diagnose the case.¹³ Initially we diagnosed this case as flare of poly-articular JIA disease, but there was high suspicion of MIS-C based on the COVID-19 pandemic situation of the country, history and clinical findings. We strictly followed WHO diagnostic criteria of MIS-C.¹⁴ In MIS-C fever and gastrointestinal symptoms are more common. Our patient had fever but no gastrointestinal manifestation. There were extensive erythematous lesions on both the soles which is one of clinical manifestations of MIS-C.¹⁵ Luisa et.al. reported arthritis in a case of MIS-C which is consistent with our case report.¹⁶ Many children with MIS-C present as KD like illness. In the Department of Paediatrics (Paediatric Rheumatology Division), Bangabandhu Sheikh Mujib Medical University, more than 64 cases of MIS-C were admitted from July 2020 to March 2022. Majority of the cases presented with KD like illness followed by severe MIS-C.¹⁷ Coronary artery and myocardial involvements are

more common in KD like illness. Though our patient “K” presented with cutaneous lesions over the soles along with other features of MIS-C, features were not typical of KD like illness and his echo-cardiograph findings were also normal.

Few published case series showed elevated IL-6 levels in MIS-C patients.¹⁸⁻²⁰ We also found high level of IL-6 in our case. Other inflammatory markers including ESR, CRP, Ferritin, pro-calcitonin and D-dimer were also high.

Many studies showed effective treatment of MIS-C with IVIG with or without steroids.²⁰ We treated our case with IVIG along with I/V Methylprednisolone followed by oral prednisolone. Our patient had a favorable response and improved without any squealy with this treatment.

Conclusion:

MIS-C is a recently identified entity of multi-system inflammatory illness that affects children as a post COVID-19 complication. Like any other comorbid conditions, children suffering from any rheumatologic disease are more susceptible to develop MIS-C following COVID-19 infection. Muco-cutaneous manifestation is common in young children. Presentation of MIS-C is typically diffuse and non-specific. Early and high index of suspicion and management can reduce mortality and morbidity.

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