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Case Report

Multisystem inflammatory syndrome in an adult with the unusual constellation of symptoms and sign acting as red herrings

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Abstract

A multisystem inflammatory syndrome is a rare emerging entity, resembling Kawasaki disease after covid 2019 viral illness(1). Here, we describe a case of MIS -A in a 21year old girl who presented with fever, mucocutaneous rash and multisystem involvement, which was treated with intravenous immunoglobulins and methylprednisolone, resulting in uneventful recovery with prompt treatment and care at Jaffna Teaching hospital.

Keywords

Covid 19, Kawasaki disease, MIS-A

Background

The multisystem inflammatory syndrome (MIS) associated with covid 19 is rare. A life-threatening condition was diagnosed in April, and a similar illness was reported in June 2020 in adults (1). Immunopathology of MIS is not identified but, it is characterized by a hyper immune response involving extrapulmonary organs (2). Most of the patients had presented with negative PCR and positive serology at the time of diagnosis. The MIS in adults has been described in patients aged 21years or above. The clinical features are like as Kawasaki disease (KD), macrophage activation syndrome (MAS), and cytokine release syndrome(3). Steroids and IV immunoglobulins are two primary treatment options in the severe clinical spectrum of covid related MIS-A (4)

Case report

A 21-year-old girl from Jaffna, presented with fever and sore throat for two days and myalgia, arthralgia and loose stool for one-day with no other systemic symptoms. She denied a contact history of Covid or similar febrile illnesses. On examination, she was febrile and pale. She was hypotensive (80/50mmhg) and tachycardiac (120/min) on admission.

She had generalised erythematous blanching rash and erythema multiforme like an eruption involving the palms and soles. She had cervical lymphadenopathy and lip swelling without strawberry tongue.

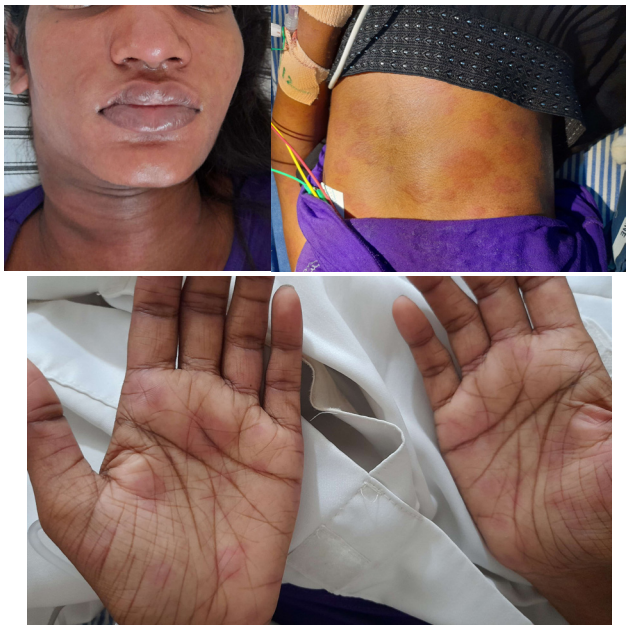

Figure1: erythematous blanching rash and eruption

Investigations showed neutrophilic leucocytosis with lymphopenia and thrombocytopenia (WBC -133000, lymphocytes -430, HB 9.8g/dl, MCV 81 fl, Platelets -140000), raised inflammatory markers (CRP-153mg/l, ESR -45mm in 1st hour), elevated transaminases (ALT-44 U/l, AST-68U/l) with hypoalbuminemia (15g/dl) and high INR (1.72).

The level of d-dimer (3.63mg/l) fibrinogen (4.0 g/l) and ferritin were elevated with high normal level of LDH (240 U/l). Cardiac screening including ECG, Troponin I and 2D echocardiography were normal. Nasopharyngeal swab for covid rapid antigen and RT-

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PCR was negative and serum for SARS – COV-2 IgG antibody was positive. Serology for Mycoplasma and Typhus was negative.

Discussion

The multisystem inflammatory syndrome is an emerging serious condition that is linked to COVID 19 disease. Clinicians may face challenges in diagnosing and treating MIS, as it mimics toxic shock syndrome, Kawasaki disease, haemophagocytic lymphohistiocytosis(4). Unlike the acute cytokine storm seen in the early covid 19 illness, the temporal relation between covid infection and MIS-A remains unclear.

Center for disease control and prevention defined MIS-A as a condition, where a patient aged ≥21 years admitted with acute febrile illness who meets three clinical criteria including at least one must be a primary clinical criterion and without an alternative diagnosis for the illness (e.g., bacterial sepsis, exacerbation of a chronic medical condition). Severe cardiac illness *or* Rash and non-purulent conjunctivitis are primary criteria.

Secondary clinical criteria include neurologic signs and symptoms, Shock, or hypotension not attributable to medical therapy, abdominal pain, vomiting, or diarrhoea, and thrombocytopenia (platelet count <150,000/ microliter) supported by Laboratory evidence of a presence of inflammation and SARS-CoV-2 infection (5).

Our patient filled the criterion by fever for more than three days with rash, non-purulent conjunctivitis, diarrhoea, thrombocytopenia, elevated CRP and ESR, high fibrinogen level with positive serology for SARS CoV-2. The American College of Rheumatology recommends IVIG to be considered as first-line immunomodulatory therapy, and steroids can be used as adjuvant therapy in shock. IVIG alters regulatory T cells number and function, which helps to control the inflammatory process (5). Our patient was given IVIG 2g/kg in a single dose followed by IV methylprednisolone 30mg/kg for 3days. Then she was started on oral prednisolone that was gradually tailed off and patient had showed marked improvement and achieved complete remission.

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