Acta Scientific MEDICAL SCIENCES (ISSN: 2582-0931)

Volume 6 Issue 7 July 2022

Case Report

Multisystem Inflammatory Syndrome in Adults: Kawasaki Like Condition

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DOI: 10.31080/ASMS.2022.06.1282

Received: April 04, 2022

Published: June 20, 2022

Abstract

The 2019 novel coronavirus (2019-nCoV) or the severe acute respiratory syndrome corona virus 2 (SARS-CoV-2) is rapidly spreading causing a pandemic all over the world [1]. Mainly affecting the lungs, it can cause severe acute respiratory distress syndrome. It may also affect kidneys, heart, nervous system and other organs [2]. Researches have defined almost all forms of acute COVID19 infections. However, after Covid effects are yet to be discovered. Clinical series of Kawasaki-like multisystem inflammatory syndrome (MIS), occurring after viral clearance, have been described in children (MIS-C) [3] and less commonly among adults (MIS-A) [4].

Keywords: SARS-CoV-2; COVID-19; Kawasaki

Case Report

After ethical and family approval we report a forty-eight year old white male with no medical history, underwent surgery for acute hemorrhoids. Shortly after, the patient reported fatigue, arthralgia and mild fever. No local surgical complications were found. The following day, the patient was admitted to intensive care unit in Mahdia teaching Hosptal (Tunisia) for multiple organ failure. Physical examination showed high persistent fever (40c°) despite antipyretics, severe hypotension, confusion, anuria, cardiac arrhythmia, rash on the chest and abdomen, mucositis, red dry cracked lips and swollen tongue, conjunctivitis and skin ulcerations on genital area (Images were obtained with patient consent and are shown in the appendix).Blood tests revealed an inflammatory state: C-reactive protein (CRP) level was 266 mg/L, anemia (Hb: 8.1 g/dL), thrombocytopenia (Platelets: 57.000), low white blood cell count (WBC: 2800), acute kidney failure (serum creatinine level was 506 µmol/L, uremia: 43 mmol/L), low prothrombin level (25%), elevated lactate dehydrogenase (696 UI/L), high lactatemia (4.6 mmol/L), elevated pro BNP (5800 ng/L), elevated troponin (162 µg/L), mild hepatic cytolysis (SGOT:149 UI/L, SGPT: 49 UI/L), high ferritinemia level:1983 ng/mL, low serum albumin: 25g/L, high triglyceride level 2,53 g/L. Blood smear showed Schistocytes, with normal Haptoglobin: 4g/L. Thoracoabdominopelvic CT scan showed: micronodules of the lungs and a splenic infarction. Electrocardiogram displayed tachycardia with episodes of atrial fibrillation. Heart ultrasound showed no signs of endocarditis. Left ventricular ejection fraction was normal. After fluid resuscitation, high doses of noradrenalin were needed in order to restore convenient blood pressure (9 mg per hour of noradrenaline). The patient was prescribed broad-spectrum antibiotics, antiviral drugs. Intermittent kidney replacement was also needed. On third day of symptoms, fever kept tray (39°-40°C), blood pressure was low with high doses of noradrenalin along with kidney failure. Blood, sputum and urine cultures were all negative for bacteria. Viral infection was ruled out via viral panel. SARS-CoV-2 PCR of nasopharyngeal swab was negative. Blood screening for CMV, aspergillosis, pneumocystosis, hepatitis and HIV were all clear. Invasive Candidiasis was also ruled out.

Further interrogation revealed a history of uncomplicated Covid 19 infection detected with PCR of nasopharyngeal swab a month ago. Hence, multisystem inflammatory syndrome in adults was suspected. Accordingly, the patient was prescribed intravenous immunoglobulin during three days (0.4g/kg/day)as well as high doses of Methylprednisolone (1 g/day) for three days followed by tapering. Two days after the initiation of treatment, fever dropped down, hemodynamics improved allowing noradrenaline with drawal, urine output increased, and blood creatinine decreased (serum creatinine level: 116 µmol/L, uremia: 18 mmol/L, inflammatory markers decreased (CRP: 20 mg/L), but low blood count persisted (Hb:7.1 g/dL, WBC: 500, neutrophil count: 400, Platelets: 26.000). Bone marrow punction revealed an hemophagocytosis, the patient was prescribed Folate and Vitamin B 12 in adjunction with corticosteroids. After 11 days of stabilization, the patient developed a septic shock due to low neutrophil count, and died shortly after.

Discussion

SARS-CoV-2 can trigger a range of inflammatory syndromes. This serious hyper inflammatory condition presents approximately 4 weeks after onset of acute Covid-19 with extrapulmonary multi organ dysfunction [5]. In our case, the onset of the symptoms is exactly one month after confirmed Covid 19 infection.

The virulent strain of SARS-CoV-2 appears to cause a post-infectious inflammatory syndrome similar to Kawasaki disease in pediatric population [6]. After SARS-CoV-2 infection, Kawasaki-like multisystem inflammatory syndrome can also occur among adults (MIS-A) [7]. Typically, these patients had high-grade fever, severe asthenia, abdominal pain and diarrhea, hypotension related to capillary leak syndrome and vasoplegia, and pronounced biologic inflammatory syndrome [4].

In our case, after ruling out possible infection, given the patient's constellation of signs (fever for more than 5 days, erythema multiforme-like rash, bilateral non-exudative conjunctivitis, erythema or cracking of the lips), he matches the American Heart Association (AHA) criteria for Kawasaki disease [8]. Subsequently, he was diagnosed with Kawasaki-like multisystem inflammatory syndrome associated with COVID-19. Given low systolic blood pressure requiring vasoactive drugs, the patient presented with Kawasaki Disease Shock Syndrome [9]. It's important to highlight that the patient had no active Covid 19 infection at the time of diagnosis.

In some cases, patients diagnosed with after Covid MIS-A show prominent cardiac dysfunction [10], some of them show signs of sever myocarditis [4]. In our case, the patient had rapid atrial fibrillation, with elevated pro BNP (5800 ng/L), high troponin level (162 µg/L). But transthoracic heart ultrasound displayed normal left ventricular function.

In our case, splenic infarction found on CT scan can be correlated to Kawasaki disease's angiitis [11]. As for acute kidney failure in MIS-A, it is thought to be mainly related to cytokine-mediated hypotension and cardiac dysfunction, leading to renal hypoperfusion [12]. However, kidney thrombotic microangipoathy have been reported in some cases [7]. In our case, in addition to high uremia and high serum creatinine level, Schistocytes were found on blood smear with no proteinuria.

Although the cause of Kawasaki disease remains unknown, the most widely accepted theory is an aberrant immune response to an infectious trigger [10]. Emerging reports depict the phenotype of MIS-C as a combination of Kawasaki disease, toxic shock syndrome, and macrophage activation syndrome [10]. In our case, macrophage activation syndrome was confirmed via bone marrow punction. The resolution of symptoms and normalization of vital signs within two days after treatment with corticosteroids and intravenous immunoglobulin is supports the diagnosis. Aspirin was not prescribed given the patient had severe thrombocytopenia.

Figure 1: Rash of the chest and abdomen.

Figure 2: Swollen tongue and cracked lips.

Figure 3: Conjunctivitis.

Figure 4: Ulcerations around genital area.

Conclusion

Multisystem inflammatory syndrome in adults (MIS-A) is a delayed complication due to Covid 19 infection. In fact, distinguishing MIS-A from other severe infectious or inflammatory disease is challenging. We report a rare case of multisystem inflammatory syndrome with Kawasaki like Disease associated with Kawasaki Shock Syndrome and macrophage activation syndrome. Despite being rare, this clinical presentation requires urgent recognition, hemodynamic support, and immediate management.

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