



A CASE OF HENOCHE-SCHONLEIN PURPURA IN A COVID-19 PATIENT

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Article Received on 07/04/2021

Article Revised on 28/04/2021

Article Accepted on 18/05/2021

ABSTRACT

Henoch-Schonlein purpura (HSP) is the most common small vessel vasculitis of childhood. It is an IgA-mediated, self-limiting disease that involves multiple organs including skin, kidneys, and the GI tract. HSP usually occurs following upper respiratory tract infections and can occur secondary to vaccinations, medications, malignancies, and insect bites. It is characterized by a clinical triad of palpable purpura, abdominal pain, and arthritis. COVID-19 is caused by the novel coronavirus SARS-CoV-2, which is a single stranded RNA virus. The clinical presentation of COVID-19 ranges from being asymptomatic to severe disease resulting in mortality. Although pulmonary manifestations are the most common, the virus can have an impact on other organ systems as well. However, data related to extra pulmonary manifestations is still rapidly emerging. We herein report a patient with Henoch-Schonlein purpura (HSP) secondary to a COVID-19 infection.

KEYWORDS: palpable purpura, abdominal pain, and arthritis.

INTRODUCTION

Henoch-Schonlein purpura (HSP) is a common IgA-mediated small vessel vasculitis of childhood. It is a self-limiting disease that involves various systems. The pathophysiology of HSP is still not well known; however, it's proposed that it's caused by IgA-mediated immune complexes deposition in the small vessels of the skin, joints, kidneys, and gastrointestinal tract triggered by an underlying upper respiratory tract infection.^[1] It's diagnosed clinically based on the EULAR/PRINTO/PRES criteria which consists of compulsory and supportive criteria.^[2] SARS-CoV-2, the cause of COVID-19 is a single stranded RNA virus that primarily affects the respiratory system ranging from most common clinical manifestations including cough, dyspnea, fever, and sore throat, similar to SARS and MERS to the development of pneumonia with ARDS, acute hypoxic respiratory failure, and/or death. However, the extra-pulmonary manifestations are not uncommon and are rapidly emerging this includes distinct patterns of vasculitis, predominantly related to cutaneous and coronary arteries.^[3,4] The purpose of this report is to present a possible association of COVID-19 with a first presentation of HSP in a previously healthy child.

Case presentation

A 13-year-old Jordanian female patient with no prior medical or surgical history, tested positive for COVID-19, presented with lower limb skin rash for 2 weeks prior

to her admission. It started as a sudden non-pruritic, non-blanchable tender maculopapular rash bilaterally on her lower limbs with crustations in an ascending fashion from soles to knees. Associated with bilateral ankle swelling, pain, and redness however, without any limitation in movement. Patient reported severe generalized abdominal pain but denied any rigors, chills, and hematuria. She also denied a history of insect bite, and any relevant drug history. Patient took paracetamol and placed cold compressors on the rash with no improvement. Patient denies any other symptoms and reports that she never encountered such symptoms in the past. On admission, temperature was 39 °C and all other vitals were normal. Physical examination showed intact mental status, normal S1 and S2 with no added sounds, good bilateral air entry and clear lung sounds, soft lax abdomen with mild abdominal tenderness. Patient was in pain and ill-looking. Abdominal ultrasound revealed sizes of liver, spleen, and kidneys were within normal and no intra-abdominal lymphadenopathy or fluid.

Laboratory testing revealed hematocrit of 33%, platelet count of 430 x 10³/mL, WBC count of 4000/mcL, Bun of 10mg/dL, Creatinine of 0.8mg/dL, AST of 30 U/L, ALT of 50 U/L, ALP of 120 U/L. CRP was positive and Serial elevation of ESR with 37 mm/hr at presentation, then 75 mm/hr and then 100 mm/hr. Urinalysis showed WBC of 5, but RBCs, Albumin, and sugar null. Otherwise, the rest of the labs including CBC

differential, blood film are within normal. COVID-19 nasal swab reverse transcription polymerase chain reaction (RT-PCR) tested positive for the infection. A punch biopsy was obtained from the skin lesion and revealed orthokeratosis in the epidermis, and mild-

moderate perivascular mixed acute and chronic inflammatory cells infiltrate with involvement of wall, with nuclear debris, extravasated RBCs and focal fibrinoid necrosis in the dermis which is consistent with leukocytoclastic vasculitis.



Figure 1: Purpuric papules of lower limb characteristic of HSP.

Treatment

Treatment was conservative before her fever and severe abdominal pain. However, patient then admitted for 12 days, given ceftriaxone intravenously in the 1st four days for the fever, then started on methylprednisolone 30mg twice daily for 6 days and IVIG 60mg per day slowly over 16 hours for 3 days, during her admission she also given omeprazole 30 mg twice daily.

On discharge patient switched to prednisolone 5mg tabs, given six tabs daily at early morning and tapered over two weeks.

Follow up of the patient at the clinic two weeks after discharge revealed no episodes of abdominal pain or fever in the past two weeks and showed excellent improvement in her dermatological lesions (figure 2). CBC, Fluorine analysis, CRP, ESR were done as follow up at the clinic in the 2nd and sixth weeks post discharge which revealed excellent improvement with the abnormal results reverts back to normal at 6 weeks post discharge.



Figure 2: Patient lesions 2 weeks post discharge.

DISCUSSION

Vasculitis is descriptive term covering a group of heterogeneous pathologies characterized by inflammation and necrosis of blood vessels. They occur in various sizes and locations leading to variable clinical and pathological features.^[5] Multiple studies suggested that SARS-CoV2 is associated with systemic vasculitis, cutaneous vasculitis, and vasculitis mimics.^[4] A study from northern Italy observed that the COVID-19 epidemic was associated with a significant rise in numbers of patients with Kawasaki and Kawasaki-like vasculitis with a higher incidence being severe including Kawasaki disease shock syndrome.^[6] The proposed pathophysiology of COVID-19 induced vasculitis is the escalation from type 2 T-helper immune response) to type 3 hypersensitivity with deposition of immune complexes inside the vascular walls inducing a severe inflammatory reaction and a cytokine release syndrome, with interleukin-6 is the key myokine in this outline.^[7] Moreover, it's illustrated that direct viral infection of endothelial cells and diffuse endothelial inflammation superseded with induction of endotheliitis, apoptosis, and pyroptosis in autopsy cases of COVID-19.^[8]

Henoch-Schönlein purpura (HSP) is estimated to affect 10 to 20 per 100,000 children per year and the majority of children present before the age of 10. It is more severe and more likely to cause long-term renal disease in adults.^[9] From the patient's history and lab results, she had no prior viral or bacterial infections that might be a trigger for the underlying disease. With streptococcal infections being the most common causative agent followed by viral infections, the patient only tested positive for SARS-CoV-2 suggesting that it's the only triggering agent. IgA vasculitis associated with COVID-19 infection has been documented in multiple patients. It was reported in a 78-year-old male that developed cutaneous vasculitis, arthritis and nephritic syndrome following COVID-19 pneumonia. PCR for SARS-CoV-2 was negative; however, serology was positive. A kidney biopsy revealed the presence of an IgA vasculitis, thus confirming the diagnosis.^[10] Moreover, another case was described by AlGhoozi et al^[11] of a 4-year-old boy that developed cutaneous, and musculoskeletal manifestations of HSP after a recent recovery from a COVID-19 upper respiratory tract infection. Acute hemorrhagic edema of infancy (AHEI), a rare variant of HSP has been reported in 2016 in an 8-month-old infant who previously tested positive for coronavirus NL63 via Nucleic Acid Amplification test (NAAT).^[12] A coronavirus OC43 vasculitis also has been reported recently in a 63-year-old female patient.^[13] Vasculitis with its various pictures has been reported in response to COVID-19. However, future research is needed to provide more substantial evidence for a potential association on the role of SARS-Cov2 in the pathogenesis of vasculitis in general and IgA-mediated vasculitis in particular.

CONCLUSION

The interface of viruses and endothelial cells had been a topic of interest in many viral diseases, here we report a case of HSP in a Covid patient which could with reporting other cases and studying them signify this relation in Covid 19. This will figure out some pathological events that still not understood completely in Covid 19 infection, and help us to reassess our treatment plans that we overtake in Covid19 pandemic.

Conflict of interest

No conflict of interest.

REFERENCES

1. Roache-Robinson P, Hotwagner DT. Henoch Schönlein Purpura. [Updated 2020 Dec 2]. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK537252/>
2. Ruperto N, et al; Paediatric Rheumatology International Trials Organisation (PRINTO). EULAR/PRINTO/PRES criteria for Henoch-Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part I: Overall methodology and clinical characterisation. *Ann Rheum Dis*, 2010 May; 69(5): 790-7.
3. Huang C, Wang Y, Li X et al: Clinical features of patients infected with 2019 novel coronavirus in Wuhan, China. *Lancet*, 2020; 395(10223): 497-506
4. [https://www.thelancet.com/journals/lanrhe/article/PIIS2665-9913\(20\)30420-3/fulltext](https://www.thelancet.com/journals/lanrhe/article/PIIS2665-9913(20)30420-3/fulltext).
5. Savage CO, Harper L, Cockwell P, Adu D, Howie AJ. ABC of arterial and vascular disease: vasculitis. *BMJ*, 2000 May 13; 320(7245): 1325-8.
6. Verdoni, L., Mazza, A., Gervasoni, A., Martelli, L., Ruggeri, M., Ciuffreda, M., Bonanomi, E., & D'Antiga, L. (2020). An outbreak of severe Kawasaki-like disease at the Italian epicentre of the SARS-CoV-2 epidemic: an observational cohort study. *Lancet (London, England)*, 395(10239): 1771–1778.
7. Roncati, L., Ligabue, G., Fabbiani, L., Malagoli, C., Gallo, G., Lusenti, B., Nasillo, V., Manenti, A., & Maiorana, A. (2020). Type 3 hypersensitivity in COVID-19 vasculitis. *Clinical immunology (Orlando, Fla.)*, 217, 108487. <https://doi.org/10.1016/j.clim.2020.108487>.
8. Varga, Z., Flammer, A. J., Steiger, P., Haberecker, M., Andermatt, R., Zinkernagel, A. S., Mehra, M. R., Schuepbach, R. A., Ruschitzka, F., & Moch, H. (2020). Endothelial cell infection and endotheliitis in COVID-19. *Lancet (London, England)*, 395(10234): 1417–1418.
9. Reamy BV, Williams PM, Lindsay TJ. Henoch-Schönlein purpura. *Am Fam Physician*, 2009 Oct 1; 80(7): 697-704.
10. <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7439008/>.
11. <https://casereports.bmj.com/content/14/1/e239910>.

12. Chesser H, Chambliss JM, Zwemer E. Acute Hemorrhagic Edema of Infancy after Coronavirus Infection with Recurrent Rash. Case Rep Pediatr, 2017; 2017: 5637503.
13. <https://casereports.bmj.com/content/13/9/e237407>.