CASE REPORT

Oman Medical Journal [2025], Vol. 40, No. 2: e740

Immunoglobulin A Vasculitis Following COVID-19 Infection in an Adolescent Girl

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ARTICLE INFO

Article history:
Received: 17 May 2023
Accepted: 22 June 2023

Online:
DOI 10.5001/omj.2025.09

Keywords:
COVID-19; Henoch-Schoenlein Purpura; Immunoglobulin A Vasculitis; Azathioprine; Steroids; Oman.

ABSTRACT

Immunoglobulin A vasculitis, formerly known as Henoch-Schoenlein purpura, is a rare acute auto-immune condition often associated with infections. We describe an adolescent girl who had recently recovered from COVID-19 infection. She presented with painless hematuria, high blood pressure, purpuric skin rash, bilateral ankle pain and swelling, abdominal pain, and inability to walk. She was admitted and clinically diagnosed with immunoglobulin A vasculitis and started on steroid treatment, to which there was a dramatic response, only to relapse as the steroid was being tapered off. Treatment with azathioprine as a steroid-sparing agent led to complete remission without recurrence. This case also suggests that SARS-CoV-2 infection may trigger the development of autoimmune diseases.

Immunoglobulin A vasculitis (IgAV) is a self-limiting disease systemic, non-granulomatous, and autoimmune complex of small vessel vasculitis, with multiorgan involvement.[1] Though it can occur at any age, it is most frequently seen in children. IgAV is the most common systemic vasculitis in childhood, with an incidence of approximately 20 per 100 000 children.[2,3]

The etiology of IgAV is unclear, but it is generally associated with infections, medications, vaccinations, cancers, alpha-1-antitrypsin deficiency, and autoimmune diseases such as familial Mediterranean fever.[4] IgAV is characterized by a classic tetrad of nonthrombocytopenic palpable purpura, arthritis or arthralgias, and gastrointestinal and renal involvement, but rarely involves the lungs, central nervous system, or genitourinary tract.[5]

We report a case of IgAV diagnosed in a young girl who had recovered from a recent COVID-19 infection. She was treated initially with steroids, which led to dependency, and later with azathioprine, leading to complete remission.

CASE REPORT

A 14-year-old girl with known chronic eczema, was admitted to our tertiary hospital with painless hematuria, high blood pressure, and purpuric skin rash. The rash started over her lower limbs and then progressed to the abdomen and upper limbs. It was associated with bilateral ankle pain and swelling, abdominal pain, and inability to walk. She also had developed hair loss and oral ulcers, but there were no genital ulcers. She also reported having irregular menstruation for 15 days. All these symptoms began following a bout of COVID-19 infection, which she had contracted eight weeks before the current presentation.

Clinical examination revealed a young girl with morbid obesity, a body mass index of 45 kg/m^2, and relatively high blood pressure (130/80) for her age. She was afebrile and had active, palpable purpuric erythematous lesions on her thighs, abdomen, arms, and forearms. She also had tender and swollen ankles, wrists, and shoulder joints, with malar flush [Figure 1].

Laboratory investigations revealed a total leukocyte count within the upper normal range level and a high urinary red blood cell count of > 60 cells/uL, but all remaining investigations were normal including virology, immunology, and inflammatory markers [Table 1].

The patient was put on steroid treatment (prednisolone 60 mg/day) along with lisinopril (5 mg/day) for two weeks. However, once her condition improved, the family discontinued the medications.

One month later, she was seen by a dermatologist who took skin punch biopsies for histopathological examination. Its findings revealed leukocytoclastic vasculitis, consistent with IgAV with no dysplasia

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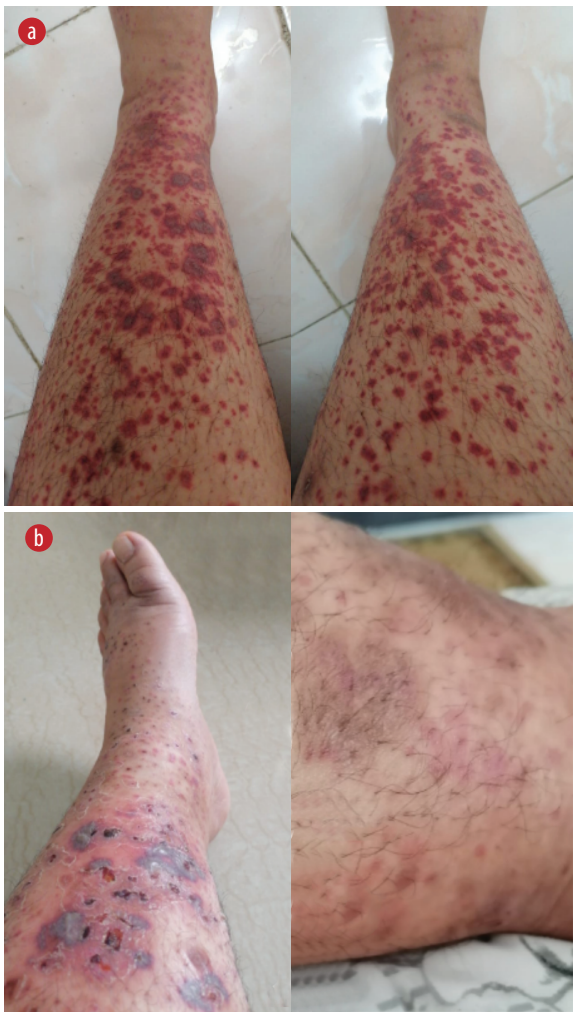


Figure 1: Active purpuric erythematous lesion on the leg. **(a)** Acute phase of polymorphic Gutate non-blanching erythematous purpuric lesions along with large dusky purpuric lesions with hemorrhagic centers. **(b)** Healing phase with crusted and fading purpuric lesions, along with others that have healed with post-inflammatory hyperpigmentation.

Table 1: Results of laboratory tests on admission.

Variables	Laboratory results	Normal values
Hemoglobin, g/dL	14	11.5–15.5
Total leukocytes, × 10^3/μL	10	2.2–10
Platelets, × 10^9/μL	419	150–450
International normalized ratio	0.95	0.8–1.0
Serum creatinine, umol/L	58	53–97.2
Serum urea, mmol/L	3.6	2.1–8.5
Urine protein-creatinine ratio, mg/mmol	6.9	< 20.0
Urine red blood cells, cells/uL	> 60	< 2.0
Albumin, g/L	36	34–50
Erythrocyte sedimentation rate, mm/h	17	2–30
C-reactive protein, mg/L	5	< 10
Complement level C3, mg/L	1927	850–1600
Complement level C4, mg/L	335	120–360
Antinuclear antibody	Negative	≤ 1:80
Anti-neutrophilic cytoplasmic autoantibody, AU/mL	Nonreactive	< 19
Immunoglobulin A serum, mg/dL	Not available	> 368
HIV 1 and 2 antibodies	Nonreactive	Not detected
Hepatitis B surface antigen, log IU/mL	0.32 nonreactive ratio	1–9
Anti-hepatitis C virus Abs, log IU/mL	Nonreactive	1–8
Thyroid stimulating hormone, mIU/L	2.8	0.4–4
Free T4, pmol/L	14	11–18
Glycated hemoglobin, %	5.4	< 6.0
QuantiFERON TB test, IU/mL	Not detected	0.2–0.99

or malignancy. The oral prednisone (60 mg/day) treatment was restarted. Within one month, there was a dramatic response, but when over the subsequent two months, the steroid dosage was reduced to < 15 mg daily, she began to experience relapses.

The post-relapse skin punch biopsy findings in the hematoxylin and eosin-stained slides are shown in Figure 2. It shows the histopathological changes in hematoxylin and eosin-stained slides of IgAV. While the immunofluorescence findings revealed superficial dermal vessels with positive staining for IgA++ and C3++, the results for IgG and IgM were negative.

Over the next three months, with tapering of steroid dose, once again, the patient developed frequent relapses of skin eruption and arthralgia. Hence, azathioprine 100 mg per day was started as a steroid-sparing agent along with the gradual withdrawal of steroids over the next three months when no relapses occurred. During the next six months of the follow-up period, the patient continued to take azathioprine 100 mg per day and achieved complete remission without any recurrence.

Figure 3 summarizes the timeline of the current case events from the initial development of IgAV post-COVID-19 infection till complete remission.

DISCUSSION

COVID-19 is known to cause vasculitis-like syndromes, as seen in our patient, as reported in previously published cases listed in Table 2.[6–12]

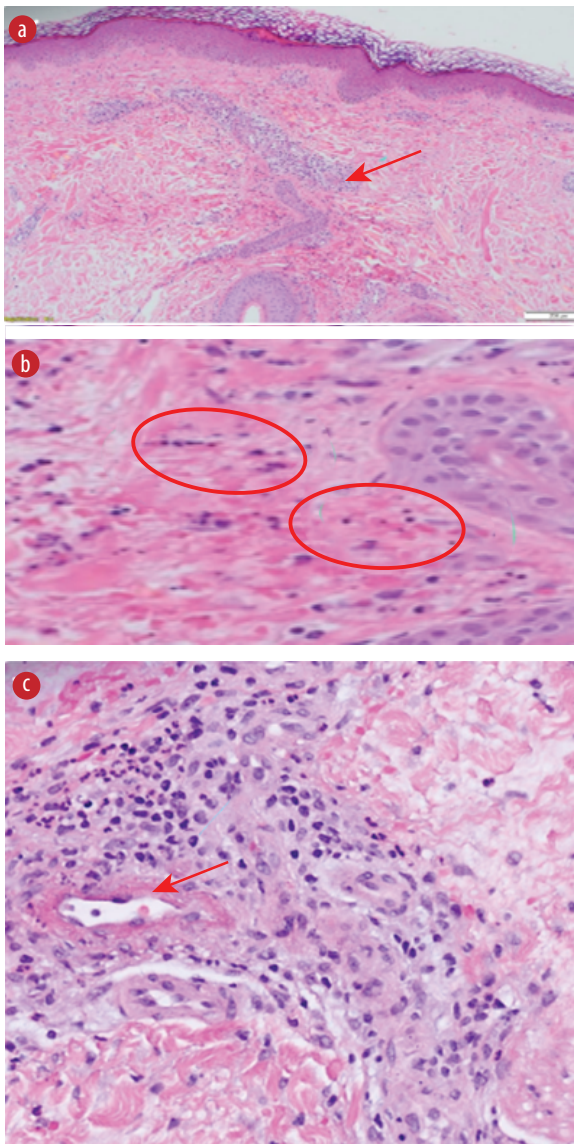
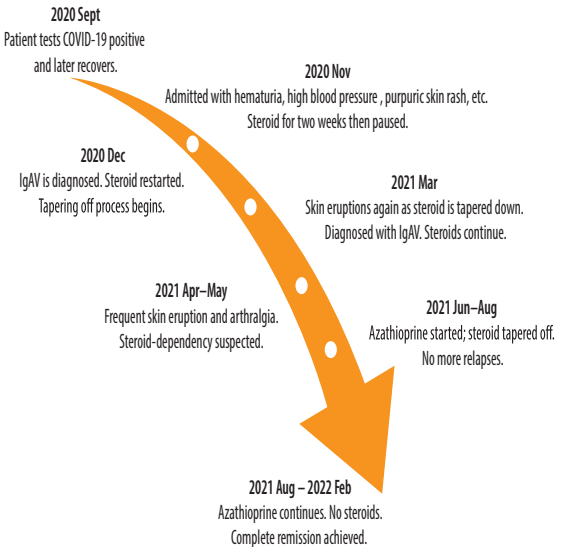


Figure 2: Histopathological findings from the skin punch biopsy taken after several relapses. **(a)** Superficial perivascular inflammatory infiltrate. **(b)** Extravasation of red blood cells and leukocytoclasia (nuclear dust). **(c)** Fibrinoid necrosis of the vessel.

The European League Against Rheumatism, Pediatric Rheumatology International Trials Organization, and Pediatric Rheumatology European Society (EULAR/PRINTO/PRES) collaboratively published in 2010, the following classification criteria for childhood vasculitides, including IgAV: purpura or petechiae and one of the following four criteria: abdominal discomfort, arthritis or arthralgia, kidney association, leukocytoclastic vasculitis with predominant IgA deposits, or proliferative glomerulonephritis with predominant IgA deposits (sensitivity 100%; specificity 87%).[13] Our patient had all these characteristics.



IgAV: immunoglobulin A vasculitis.

Figure 3: Timeline of the events until complete remission.

As IgAV is usually diagnosed clinically, laboratory tests are often not considered essential and skin biopsies are rarely taken.[14] However, chronic or recurrent IgAV symptoms should always raise concern for alternative causes of cutaneous small vessel vasculitis, such as anti-neutrophil cytoplasmic antibodies-associated vasculitis or systemic lupus erythematosus.[15,16]

To investigate the several relapses in our patient, various laboratory tests were conducted, all of which came back negative. Blood results were also within normal levels. Therefore, we obtained a skin biopsy, which revealed leukocytoclastic vasculitis involving the skin (palpable purpura).

Earlier studies performed in 2004 and 2006 (including an eight-year follow-up) showed similar results as ours, with corticosteroids providing no long-term benefits.[17–19] Renal manifestations such as hematuria and proteinuria, were not resolved after 28 days of corticosteroid treatment but were reduced in comparison with placebo. At the six-month follow-up, the study found that 61% of patients had resolved kidney manifestations compared with 34% of placebo group. This study recommended corticosteroids for patients older than six years with mild kidney manifestations.[17] The study also found statistically significant results regarding the treatment of extra-renal symptoms. Joint pain and abdominal involvements were reported with less frequency among people taking corticosteroid treatment compared to placebo. There

Table 2: Summary of published cases of immunoglobulin A vasculitis following COVID-19.

Author	Age, years	Sex	Period after COVID-19 positivity	Organs involved	Treatment
Suso et al[6]	78	male	5 weeks later	Skin; nephritis	Steroid pulse and Rituximab
Hoskins et al[7]	2	male	Same time	Skin; abdominal pain	Intravenous steroid
Allez et al[8]	24	male	unknown	Skin; abdominal pain	Methylprednisolone 0.8 mg/day
Sandhu et al[9]	22	male	Same time	Skin; nephritis	Prednisolone 1 mg/kg
AlGhoozi et al[10]	4	male	37 days later	Skin	Not stated
Jacobi et al[11]	3	male	Same time	Skin; abdominal pain	Antibiotics
Li et al[12]	30	male	Same time	Skin; nephritis	Losartan 25 mg following prednisolone 40 mg for 7 days
Atris et al (present case)	14	female	eight weeks	Skin; abdominal pain; nephritis	Lisinopril, steroid, azathioprine

was no difference between the groups in terms of skin manifestations.[18,19]

Our patient suffered several relapses while she was on steroids alone. Hence, azathioprine was added along with the steroid. A clinical and histopathological study by Foster et al,[20] found that azathioprine therapy with corticosteroids had a therapeutic effect on IgA nephritis cases. In steroid-dependent patients, such cases could be managed by tapering steroids within three months and azathioprine continued as monotherapy for six months with minimal relapses or adverse events. This approach is similar to the management of inflammatory bowel disease.[21,22]

CONCLUSION

IgAV is typically self-limiting, therefore; our patient's unusual relapses while on steroid therapy might be attributable to her history of COVID-19. We found azathioprine to be an effective therapeutic agent as it was curative while facilitating the gradual extinction of our patient's steroid dependence.

Disclosure

The authors declare no conflicts of interest. Patient consent was obtained for the publication of this case report.

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