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Case Report

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Henoch-Schönlein Purpura with Arthritis: A Case Report

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Abstract

Background: *Henoch-Schönlein purpura (HSP) is a vasculitis distinguished by purpura, arthritis, abdominal pain, and renal involvement. It has an uncommon cutaneous manifestation, with only a few cases described. The HSP is ubiquitous in infants but can also be observed in adults.*

Case presentation: *We present a 41-year-old Sudanese man with rheumatoid arthritis, abdominal cramps, and non-thrombocytopenic purpura across his extremities who had a negative medical history of HSP. The proportion of immunoglobulin alpha (IgA) was elevated, and occult blood was detected in the stool. There has also been evidence of hematuria and albuminuria. A skin biopsy also revealed leukocytoclastic vasculitis infiltration. The symptoms were satisfactorily cured after treatment.*

Conclusion: *The conventional morphology of these lesions and their location in an adult are the main findings of this case report. Appropriate management and mitigation of potential problems can be achieved with early diagnosis and interdisciplinary intervention. The disease has a broadly favorable prognosis.*

Keywords: *Henoch-Schönlein purpura, Leukocytoclastic vasculitis, Sudan*

Introduction

HSP is a type of systemic vasculitis that can be told apart from other types by its non-thrombocytopenic purpura, arthritis, abdominal pain, and nephritis [1]. It emerges as purpura or petechiae, most usually on the lower limbs but also other parts of the body [2]. HSP is an illness that may be self-reported, but it has been documented in more than 30% of patients. HSP is ubiquitous among children, and can often be recorded in adults [3]. The peak age at diagnosis is 4–6 years, with 90% of HSP cases emerging before the age of 10. The lowest occurrence is among Afro-Caribbeans, whereas the highest is among Asians. Adults had an incidence of 3.4–14.3 per million of the population. Because this is a self-limiting condition, its true prevalence may be underestimated. [4].

Palpable purpura and petechiae seem to be the most prominent dermatologic symptoms, however, certain lesions may also be encountered [5]. In most cases, the disease's exact etiology is self-limiting, except for those with renal problems. Joint and gastrointestinal aches will be relieved by symptomatic treatment with nonsteroidal anti-inflammatory medications combined with steroids [6]. When taken early in the disease, corticosteroids can improve clinical results. The prognosis is good, other than renal damage, which may necessitate a 6-month or longer follow-up [7]. This case scenario may provide valuable evidence for clinicians considering a Sudanese patient with HSP for earlier diagnosis and cure.

Case presentation

A 41-year-old Sudanese man suffered a rash that occurred in his arms and lower limbs. This rash has begun to cluster. The patient associated pruritus and slight pain. He was slightly smoky. He had an uneventful clinical record. No family history of systemic disease or HSP has been reported. Vital signs were estimated as BP 100/78 mmHg, pulse rate 91 beats/min, respiratory rate 16/min, and body temperature 36.8 C°. During the examination, several noticeable purple spots were visible on the patient's arms and legs (see Figure 1). Laboratory investigations explored normal complete hemograms, typical renal and liver profiles, and normal coagulation parameters (Table 1). Serum immunoglobulins for IgM and IgG levels were normal except IgA levels were high. Anti-streptolysin O, random blood glucose, and thyroid function tests were also normal. Complement (C3) and (C4), double-strand DNA (ds-DNA), antineutrophil cytoplasmic antibodies, Sjogren syndrome, and erythrocyte sedimentation rate were also normal. Rheumatoid factor and antinuclear factor (ANA) titers were abnormal. Urine analysis indicates proteinuria and hematuria. Stool general was shown positive occult blood. Screening investigations for cytomegalovirus (CMV), human immunodeficiency virus (HIV), hepatitis B virus, hepatitis C virus, and syphilis were also negative (Table 1).

With H&E staining, a skin biopsy of the hemorrhagic purpuric lesion exhibited leukocytoclastic vasculitis with modest infiltration of polymorphonuclear neutrophils. The patient was referred to a nephrologist who ordered a kidney biopsy due to the presence of proteinuria and hematuria. Mesangial proliferative glomerulonephritis was found after a needle biopsy. A computed tomography of the abdomen was also performed to rule out other abdominal diseases. Hydrocortisone 5 mg was applied for a week and then shifted to oral Azathioprine for 8 weeks. Vitamins B and D were also administered to provide support. The patient purpuric lesions and symptoms were resolved in 2 weeks. Urine and stool analysis one month later showed normal findings. No recurrence was observed in the next three months.

Table 1: Laboratory findings of the patient.

Variables	Patient results	Normal or control
White blood cells **x10^9/l**	6.9	4 – 10
Red blood cells **x10^{12}/l**	5.12	3.5 – 5.5
Hemoglobin **g/dl**	14.8	12 – 16
Hematocrit ***%***	44.7	35 – 47
Mean Corpuscular Volume ***fl***	87.3	78 – 98
Mean Corpuscular Hemoglobin ***pg***	28.9	26 – 35
Mean Corpuscular Hemoglobin Concentration ***%***	33.1	30 – 36
Red Distribution Width-CV ***%***	13.1	11.5 – 14.5
Absolute lymphocyte count ***x10^9/l***	2.0	1.0 – 4.3
Absolute neutrophil count ***x10^9/l***	3.9	1.5 – 7.0
Platelet count ***x10^9/l***	246	150 – 400
Erythrocyte sedimentation rate, 1st hour, ***mm/h***	5	Up to 20
Erythrocyte sedimentation rate, 2nd hour, ***mm/h***	14	Up to 30
Bleeding time, ***min***	3.49	2 – 7
Clotting time, ***min***	7.39	5 – 15
Clot retraction, ***%***	60	30 – 120
Prothrombin time, ***s***	16.2	12 – 16
Partial thromboplastin time, ***s***	39.4	26 – 43
Thrombin time, ***s***	15.2	8 – 18
Antithrombin, ***%***	90	78 – 126
D-Dimer, ***mg/l***	< 0.1	Up to 0.3
Von willebrand factor: Ag**, %**	121	50 – 160
Von Willebrand factor: Activity (CP)**, %**	43	48 – 180
Factor VIII**, *%***	130	70 – 150
FVIII/VWF: Ag ratio	1.07	> 0.60
Creatinine**, *mg/dl***	0.81	0.4 – 1.6
Blood Urea**, *mg/dl***	15.0	10 – 50
Blood Urea Nitrogen**, *mg/dl***	7.0	7 - 21
Uric Acid**, *mg/dl***	6.0	3.4 – 7.0
Total Bilirubin**, *mg/dl***	0.44	0.2 – 1.3
Direct bilirubin**, *mg/dl***	0.09	Up to 0.25
Total protein, ***g/dl***	6.9	6.6 – 8.3
Albumin, ***g/dl***	3.7	3.5 – 5.5
Alanine transaminase, ***U/l***	18	Up to 41
Aspartate transaminase, ***U/l***	28	Up to 40
Alkaline phosphatase, ***U/l***	78	Up to 115
Glycosylated Hemoglobin**, %**	5.8	4.5 – 6.5
Thyroid stimulating hormone, ***µIU/ml***	1.16	0.27 – 4.2
Free T4, ***pmol/l***	16.2	12 – 22
Free T3, ***pmol/l***	4.86	3.1 – 6.8
Hepatitis C virus screening (HCV)	Negative	—
Hepatitis B virus screening (HBV)	Negative	—
Human Immunodeficiency Virus Screening (HIV)	Negative	—
Hepatitis C virus screening (HCV)	Negative	—
Complement C3, ***mg/dl***	129	75 – 135
Complement C4, ***mg/dl***	30	9 – 36
Proteinase 3 (PR3)	0.14	< 1.0
C-reactive protein (high sensitive), ***mg/l***	0.9	< 1.0
Rheumatoid factor, ***IU/ml***	24	Up to 20
Anti cyclic citrullinated peptide, ***U/ml***	33	Up to 20

Antinuclear factor antibody (ANA)	1/320	< 1:100
Anti-double stranded DNA (dsDNA), ***IU/ml***	< 30	30 - 70
Anti-Sjogren syndrome-related antigen A (Ro), ***U/ml***	4	Up to 15
Anti- Sjogren syndrome-related antigen B (La), ***U/ml***	5	Up to 15
Immunoglobulin IgA, ***mg/dl***	543	70 – 400
Immunoglobulin IgM, ***mg/dl***	53	40 – 230
Immunoglobulin IgG, ***mg/dl***	1263	700 – 1600
Carcinoembryonic antigen, ***ng/ml***	0.3	Up to 5.0

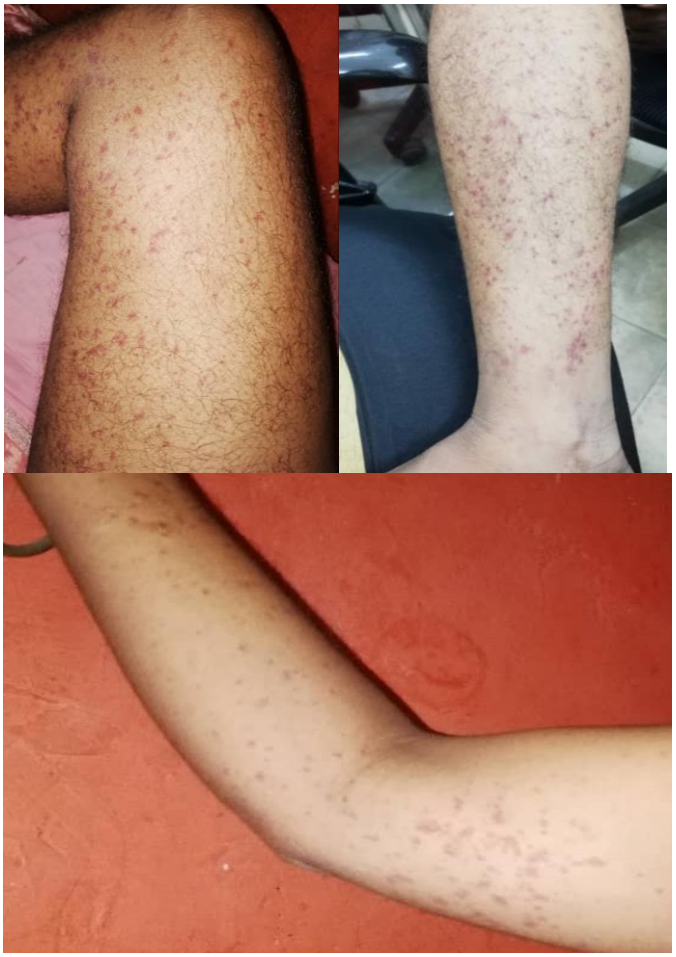

Figure 1: Palpable purpura consistent with leukocytoclastic vasculitis.

Discussion

HSP is the most prevalent kind of vasculitis. It's an immune-mediated vasculitis linked to IgA deposition, but the true pathophysiology is unknown [6]. Our patient fulfilled the HSP standards. HSP is identified by palpable purpura and petechiae on the skin. Joint involvement was the most usual systemic indication (63.2%), followed by gastrointestinal engagement (60.5%), and renal implication (60.5%) [8]. HSP is a leukocytoclastic vasculitis that affects tiny vessels. Neutrophil infiltration surrounding papillary and dermal vessels is the most common histological result.

Joint pain and swelling are the most classic signs of joint involvement, which often primarily affect the knees and ankles. The main signs involving the gastrointestinal tract are intestinal pain, vomiting, and stool hemorrhage. Renal findings encompass microscopic hematuria and albuminuria [7]. Rashes on both the upper and lower limbs, abdominal pain, and sometimes vomiting were all indications in our scenario. Even though arthritis is present, there was no joint involvement. An occult blood test positive for stool was a significant laboratory finding. However, hematuria and albuminuria were reported. There were no conspicuous indications of SLE based on negative dsDNA and or even high titer of ANA testing. Our patient's clinical, biochemical, and histopathologic findings pointed to HSP.

Direct immunofluorescence examinations frequently revealed IgA (particularly IgA1) and C3 deposition. IgA deposition was not present in all of the patients; in fact, only 63 % did. An early biopsy is required to make the diagnosis since the time of the biopsy may alter the discovery of IgA accumulation [9]. IgA and C3 immunoreactants are eliminated in 48 hours [10]. Our patient had IgA but no C3 deposition in the dermis around the vessel walls. In this case, a skin biopsy was conducted 48 hours following the onset of symptoms. Although leukocytoclastic vasculitis is normally limited to the upper layer of the dermis, one study found that it extended to the deeper layer of the dermis, resulting in scar formation [9]. Our patient has not developed any scars.

There is no consensus on the optimum treatment for HSP in terms of therapy. Due to the rarity of HSP, no randomized trials have been done. Corticosteroids have been shown in certain studies to lower the severity of abdominal discomfort and the tendency to develop renal impairment, but not to prevent recurrence [10, 11]. After hydrocortisone treatment, our patient's intestinal discomfort and skin sores improved drastically within days. Intending to reduce the intensity of his abdominal pain, proteinuria, and skin manifestations, we launched azathioprine and corticosteroid therapy. There was no hint of a recurrence. Two studies reported the use of azathioprine with corticosteroids in two patients due to uncontrolled skin lesions and progressive heavy proteinuria [12, 13]. Dapsone has been used to turn a patient off prednisone, according to Chen et al [10]. In most cases, conservative care improves HSP, but the treatment of HSP is still debatable [10,14]. Although HSP is primarily a self-limiting condition, one-third of patients will encounter one or more flare-ups of symptoms [2]. The severity of renal involvement tends to affect the long-term morbidity of HSP [4]. During follow-up, our patient had no repeat skin manifestations or hyperpigmentation, and there was no scarring.

HSP has a favorable prognosis, and full recovery with no side effects is usual. Approximately 90% of pathological changes will resolve spontaneously within weeks to months. Up to 3% of HSP patients may experience recurrent flare-ups. Up to 5% of patients with nephritis may develop end-stage renal disease [8].

Conclusion

A case of HSP with various sequelae, including arthritis, abdominal discomfort, and palpable purpura, is described. Appropriate management and mitigation of potential problems can be achieved with early diagnosis and interdisciplinary intervention. The disease has a broadly favorable prognosis.

Informed consent

The patient permitted the material to be published in a scholarly journal.

Ethics statement

The patient provided consent to be published, and the data is confidential.

Authorship

The author contributed to the article's drafting and critical revision. The author has approved the final draft.

Declaration of Competing Interest

None

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