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Case Report

A case of Henoch Schoenlein purpura and IgA nephropathy in an adult

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Key words: **vasculitis, IgA nephropathy, haematuria, systemic lupus erythematosus, skin biopsy, fibrinoid necrosis**

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Introduction

Henoch Schoenlein purpura is a small vessel vasculitis which usually presents as erythematous macular lesions, arthralgia, abdominal pain and gross haematuria. The most common symptom is rash which usually involves the extremities [1]. It occurs as result of fibrinoid necrosis of the blood vessels. Apart from the skin, the other organs involved include the gastrointestinal system, joints, kidney, lung and myocardium [2]. Deposition of immunoglobulin A accounts for the multiple organ involvement. No specific laboratory test is available to establish a diagnosis. The palpable purpura and suggestive clinical symptoms along with skin biopsy findings are helpful to make the diagnosis.

Case presentation

We report a case of an 18-year-old female who presented with non-itchy, purple spots which started on the extensor surfaces of the legs symmetrically [Figure 01] 2 weeks previously and extended to involve the upper limbs [Figures 02 & 03] and buttocks thereafter. There was no prodromal upper respiratory or gastrointestinal illness. She had intermittent abdominal cramps, gross haematuria and arthralgia. She denied any tarry-coloured stools or any symptoms of anaemia. Features suggestive of autoimmune aetiology, muscle pain and muscle weakness were absent.

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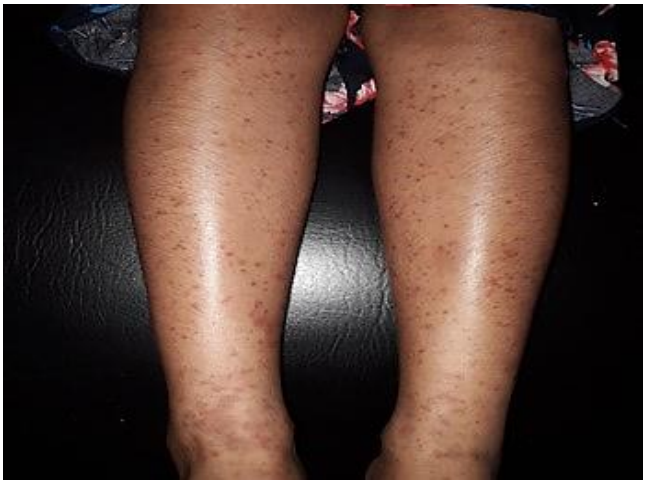

Figure 01: non-itchy, purple spots which started on the extensor surfaces of the legs symmetrically

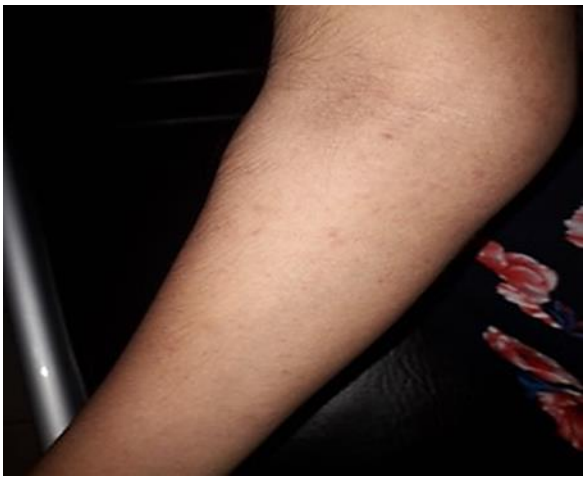

Figure 02: non-itchy, purple spots involving the upper limbs

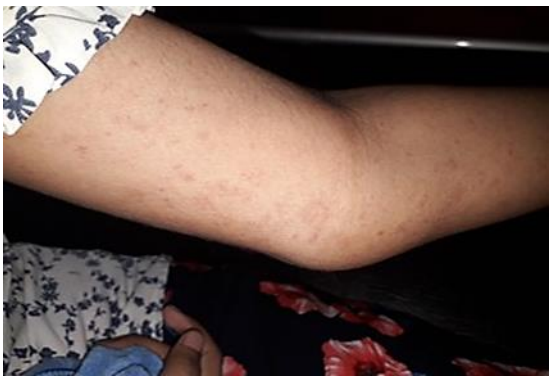

Figure 03: non-itchy, purple spots involving the upper limbs

On examination, she was afebrile and not pale. She had no malar or discoid rash. She had a more than 2mm sized, blanching, palpable rash symmetrically distributed on the upper, lower limbs and buttocks sparing the trunk and face. There was no lymphadenopathy. She had no peripheral stigmata of infective endocarditis. Ankle oedema was absent.

Cardiovascular examination showed a blood pressure of 120/ 70 mmHg and a pulse rate of 88 bpm which was regular and of good volume. Chest examination was normal with no

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detectable murmur. The abdomen, respiratory and neurology examinations were unremarkable.

Table 01: Investigations done

Test	Reference values	Results on admission	Results after 1 Week
Full blood count			
White cell counts (10^3/ µl)	4- 11	10.7	8.3
Neutrophils 10^3/ µl	2- 7	6.5	4.2
Lymphocytes 10^3/ µl	1- 5	3.2	3.4
Eosinophils 10^3/ µl	< 0.5	0.6	0.4
Monocytes 10^3/ µl	0.2- 0.8	0.4	0.3
Platelets 10^3/ µl	150- 400	368	299
Haemoglobin g/dl	11- 15	14	13.6
CRP mg/l	< 5	7	3.1
ESR mm/hr	< 22	5	6
LDH U/L	< 234	154	142
ASOT IU/ ml	< 200	< 200	
Serum Na^+ mmol/l	135- 145	139	137
Serum K^+ mmol/l	3.5- 5.1	3.9	3.9
Serum Ca^{2+} mmol/L	2.1- 2.6	2.4	2.3
Serum Creatinine µmol/L	60- 85	63	74
Blood Urea mmol/L	2.5- 6	3.2	3.2
ALT U/L	10- 50	24	20
AST U/L	10- 40	22	12
ALP U/L	25- 150	45	35
Gamma- glutamyl transferase U/L	10 - 65	34	21
Total protein g/l	65- 83	54	55
Serum Albumin g/l	35- 50	36	35
Serum Globulin g/l	20- 40	18	20
Total Bilirubin µmol/L	5- 17	7	6.4
Free Thyroxine ng/dl	1-1.7	1.2	
TSH mIU/ L	10- 28	14	
Anti-nuclear antibody	< 1/80	Negative	
Double stranded DNA IU/ ml	< 10	5	
Complement 3 mg/ dl	80-180	85	
Complement 4 mg/dl	15- 45	30	
Serum IgA level mg/dl	80- 350	450	
Hepatitis B surface antigen		Negative	
Anti-Hepatitis C IgG		Negative	
Cytomegalovirus antibody		Negative	
INR	< 1.1	1	
APTT	30-40 seconds	20	

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UFR: Pus cells	Nil	2-3/ hpf	Nil
UFR: Red cells	Nil	Field full/ hpf	Nil
UFR: Albumin	Nil	Nil	Nil
Urine for dysmorphic cells	Negative	10%	Not detected
UPCR	< 30	< 30	< 30
Blood Culture		No growth	
Urine Culture		No growth	
Blood picture		Normal	
ECG		Sinus rhythm	
2D echocardiography		Ejection fraction: 60% with normal valves	
US abdomen		Normal kidneys No hepatosplenomegaly or lymphadenopathy	

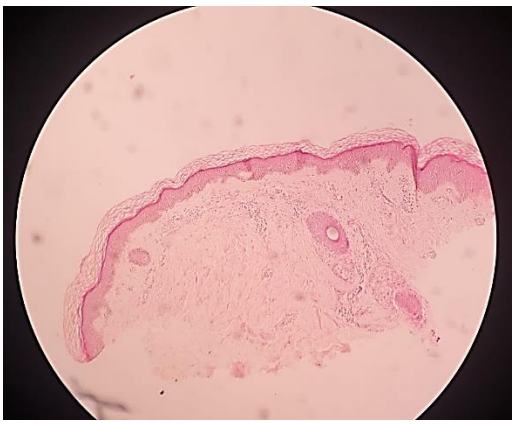

Figure 04: (H & E stain x 100) Punch biopsy from the lateral aspect of the lower limb

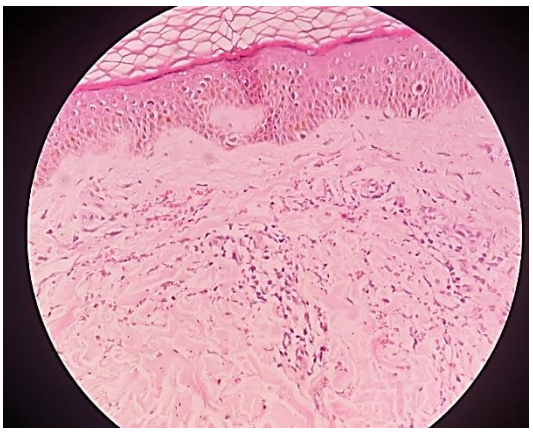

Figure 05: (H & E stain x 400) Punch biopsy from the lateral aspect of the lower limb

As the stool occult blood test was negative, she was not offered an upper gastrointestinal endoscopy. Skin biopsy was done which showed a characteristic pattern of leukocytoclastic vasculitis [Figure 04 & 05] with small vessel vasculitis evidenced by extravasated red cells and the presence of neutrophil infiltration along the vessel wall. A multidisciplinary team was employed with the participation of the consultant physician, consultant dermatologist, consultant histopathologist, consultant rheumatologist and consultant nephrologist to discuss the management plan. She was treated for Henoch Schoenlein purpura and acute glomerular nephritis (Immunoglobulin A related) with oral prednisolone 30 mg daily for 5

days which was planned to be tapered off over two weeks. After 3 days of therapy, her haematuria settled and the rash started to resolve. She was free of symptoms in 5 days and was discharged on day 7 of admission. She was reviewed at the ward in 1 weeks' time where she was found to be completely symptom free and the investigations were all normal.

Discussion

Henoch Schoenlein purpura is an immunoglobin A-mediated vasculitis which characteristically involves the small vessels of the skin, gastrointestinal tract and kidney. It is common in children aged between 3- 10 years. Its occurrence declines with age, but its course is more complicated in adults than in children [3]. Approximately 75% of patients have a history of prodromal gastrointestinal or upper respiratory illness [3].

The prodromal period comprises of mostly fever and anorexia. The most obvious symptoms include the typical non- blanching rash which commonly occurs in the lower limbs, abdominal pain, bloody stool and haematuria [3].

The EULAR criteria for diagnosis comprise a palpable purpura and petechiae in the absence of low platelets or coagulopathy with an addition of either diffuse abdominal pain, suggestive skin biopsy findings, acute arthritis or renal involvement [4]. Our patient certainly fulfilled the criteria with the typical rash and abdominal pain along with characteristic skin biopsy findings and glomerular involvement in the absence of thrombocytopenia or coagulation abnormalies.

The skin biopsy in Henoch Schoenlein purpura shows necrotizing vasculitis with fibrinoid destruction and infiltration of leukocytes which is termed as leukocytoclastic vasculitis. Other connective tissue diseases must be excluded, and the renal function must be thoroughly evaluated whilst evaluating the patient for complications [5].

Renal involvement occurs in approximately 20-55% of children and 45- 85% of adults with Henoch Schoenlein purpura [6] IgA deposition in the glomerular mesangium with varying degrees of mesangioproliferative nephritis is seen [7]. The most common presentation in children and young adults is gross haematuria followed by invisible haematuria, nephrotic syndrome, acute kidney injury and, finally, chronic kidney disease [7]. 25% of the patients with progressive renal involvement develop end stage renal disease within about 20 years from onset [7]. The most dreaded complications include acute renal shut down and severe bowel ischaemia [5]. Elevated serum immunoglobulin A levels are seen in the initial active stage of adult Henoch Schoenlein purpura [8]

Due to the self-limiting nature of the illness, the treatment can be simply supportive. The supportive measures are mainly adequate hydration and adequate pain relief and elevation of the swollen limbs. Non-steroidal anti- inflammatory medications may suffice if the

patient's main complaint is arthralgia [9] . But caution should be exercised in case of renal insufficiency. Corticosteroids are warranted when there is profound renal impairment, significant arthritis, gastrointestinal involvement and subcutaneous oedema. Refractory cases may be treated with intravenous methylprednisolone pulse therapy or immunosuppressive agents such as azathioprine and cyclophosphamide [9] .

Conclusion

Though Henoch Schoenlein purpura is rare in adolescents, it can be associated with more renal and gastrointestinal complications than in children. Therefore, it is important to make the diagnosis early and initiate corticosteroids promptly, if indicated, as in cases with profound renal impairment, significant arthritis, gastrointestinal impairment and prominent peripheral oedema, in order to minimize ~~the~~ complications.

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