Henoch-Schönlein Purpura in Primary Care: The Importance of a Holistic Assessment

Púrpura de Henoch-Schönlein nos Cuidados de Saúde Primários: A Importância de uma Avaliação Holística

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ABSTRACT

Henoch-Schönlein purpura (HSP) is a rare vasculitis in adults, requiring a high index of suspicion. This case highlights the role of Primary Healthcare and a comprehensive assessment in early recognition.

A healthy 27-year-old female presented to the Emergency Department (ED) with abdominal pain and vomiting and was discharged with suspected gastroenteritis. Four days later, she consulted her Family Doctor for persistent symptoms. A thorough review revealed arthralgia, rectal bleeding and palpable purpura on both lower limbs. She was promptly referred back to the ED. Laboratory results and abdominal ultrasound had no major alterations and abdominal computed tomography (CT) suggested inflammatory enteritis.

She was admitted for a diagnostic skin biopsy and initiated on corticosteroid therapy for suspected HSP. The patient showed a favorable clinical response to prednisolone, with stable renal function and blood pressure throughout hospitalization.

This case reinforces the value of Primary Health Care in ensuring comprehensive evaluation and facilitating early diagnosis to prevent complications.

KEYWORDS: Holistic Health; IgA Vasculitis; Primary Health Care

RESUMO

A púrpura de Henoch-Schönlein (PHS) é uma vasculite rara em adultos, exigindo elevada suspeição. Este caso destaca o papel dos Cuidados de Saúde Primários (CSP) e da avaliação holística no seu reconhecimento precoce.

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Mulher, 27 anos, saudável recorre ao Serviço de Urgência (SU) por dor abdominal e vómitos, interpretada como gastroenterite. Quatro dias depois, recorre à Médica de Família por persistência dos sintomas. Uma avaliação cuidada revelou artralgias, retorragia e púrpura palpável nos membros inferiores. Foi referenciada ao SU, onde realizou análises e ecografia abdominal (sem alterações relevantes) e tomografia computorizada (TC) abdominal sugestiva de enterite inflamatória.

Foi internada para realizar biópsia cutânea e iniciar corticoterapia por suspeita de PHS. A doente apresentou uma evolução clínica favorável com prednisolona, mantendo função renal e tensão arterial estáveis durante o internamento.

Este caso reforça o valor dos CSP na realização de uma avaliação holística e diagnóstico precoce da PHS, essencial para prevenir complicações.

PALAVRAS-CHAVE: Cuidados de Saúde Primários; Saúde Holística; Vasculite por IgA

INTRODUCTION

Henoch-Schönlein purpura (HSP), also known as IgA vasculitis, is a rare systemic small-vessel vasculitis, resulting from the accumulation of immune complexes and immunoglobulin A-1 within the walls of arterioles, venules and capillaries. This condition involves multiple organs, commonly impacting the skin, joints, gastrointestinal system and kidneys.[1]

HSP is the most frequent acute vasculitis in children, occurring at a rate of about 10 cases per 100 000 children annually. The mean age of onset of HSP is six years old and it is twice more common in males than in females. Although more common in children, it can also occur in adults, with an approximate annual incidence of about one-tenth that of children (13-14 cases per 1 000 000 adults per year). In adults, it equally affects both men and women.[2]

In adults, its clinical presentation is often more variable and potentially severe. The diagnosis remains essentially clinical but the signs may not appear simultaneously, making early recognition particularly challenging. In adults, complications related to the gastrointestinal tract and renal involvement are the primary contributors to illness and death.[3]

This clinical report illustrates a suspected case of HSP initially identified in the context of Primary Health Care (PHC). The present article was written according to CARE guidelines for case reports.[4]

CASE REPORT

A 27-year-old female patient presented with a week-long history of intense, diffuse upper abdominal pain associated with persistent vomiting. She had no significant medical history, apart from the use of a combined oral contraceptive. She was an active smoker with a cumulative exposure of approximately 6 pack-years. There was no recent travel or relevant epidemiological context.

The patient initially sought care at the Emergency Department (ED), where the physical examination was non-specific and preliminary workup was normal. She was discharged with symptomatic treatment, including a proton pump inhibitor and sucralfate, under the presumptive diagnosis of gastritis or functional dyspepsia.

Four days later, she presented to her PHC clinic with no improvement in symptoms. During this second evaluation, a detailed systemic review and comprehensive physical examination revealed additional findings. The patient reported bilateral arthralgia involving the elbows and knees, with mixed mechanical and inflammatory characteristics, including morning stiffness lasting approximately 10 minutes. She also described episodes of hematochezia. On physical examination, palpable purpura and petechiae were observed on both lower limbs, extending proximally to the root of the thighs (Figs. 1A and 1B). Abdominal examination revealed diffuse tenderness on palpation and there was no clinical evidence of arthritis, namely joint swelling, erythema or warmth.

Given this constellation of systemic symptoms, the patient was referred again to the ED with the clinical suspicion of vasculitis for further evaluation.

At the ED, a full analytical workup was conducted (Table 1). This included a complete blood count showing mild leukocytosis and elevated inflammatory markers (C-reactive protein at 15.4 mg/L), but no other hematological abnormalities. Liver and renal function tests were within normal limits. Urinalysis showed no rele-

vant hematuria or proteinuria and urinary β-hCG was negative. Viral serologies (including hepatitis B and C, human immunodeficiency virus - HIV, cytomegalovirus and Epstein-Barr virus) were negative. Autoimmune markers, including antinuclear antibodies (ANA) and antineutrophil cytoplasmic antibodies (ANCA), were also negative. Serum immunoglobulin levels, including IgA, were within normal range (IgA: 202 mg/dL). An abdominal ultrasound showed no relevant findings, but an abdominal computed tomography (CT) scan revealed signs suggestive of inflammatory enteritis.

In light of the clinical suspicion of Henoch-Schönlein purpura, the patient was admitted to the Internal Medicine ward for close monitoring, further diagnostic clarification and initiation of appropriate treatment.

A skin biopsy of a purpuric lesion on the lower limb was performed, more than one week after the onset of cutaneous lesions. Histological examination showed superficial dermal capillary ectasia, mild perivascular mononuclear inflammatory infiltrate and focal erythrocyte extravasation, without evidence of fibrinoid necrosis or leukocytoclasia. Direct immunofluorescence did not demonstrate immune complex deposition, including IgA, IgG, IgM, C3 or fibrinogen.

Systemic corticosteroid therapy was initiated with oral prednisolone at a dose of 60 mg/day, resulting in rapid and marked clinical improvement, particularly of abdominal pain and cutaneous manifestations. During hospitalization, the patient maintained stable blood pressure and normal renal function, with no evidence of renal involvement throughout admission.

She was discharged on prednisolone 50 mg/day, associated with a proton pump inhibitor. Corticosteroid tapering was conducted on an outpatient basis, with dose reductions of 10 mg every 10 days until reaching 10 mg/day, followed by tapering to 5 mg and subsequently 2.5 mg. Approximately five months after the initial presentation, while on prednisolone 2.5 mg/day, the patient developed a mild cutaneous relapse with recurrence of petechiae, but no other symptoms. She temporarily increased the prednisolone dose to 5 mg/day for 15 days, with complete resolution of symptoms. Given this relapse, colchicine at a dose of 1 mg/day was initiated as a steroid-sparing agent; however, it was discontinued due to gastrointestinal intolerance, namely diarrhea. The patient subsequently maintained low-dose prednisolone with gradual tapering and experienced no further relapses.

TABLE 1. Laboratory results from the emergency department and hospitalization

TEST	RESULT	UNIT	REFERENCE INTERVAL
HEMATOLOGY			
Erythrocytes	4.79	x 10^{12}/L	4.00-5.00
Hemoglobin	13.6	g/dL	12.0-16.0
Hematocrit	39.5	%	37.0-49.0
Mean Corpuscular Volume	82.5	fL	87.0-103.0
Mean Corpuscular Hemoglobin	28.4	pg	27.0-35.0
Mean Corpuscular Hemoglobin Concentration	34.4	g/dL	28.0-36.0
Red Cell Distribution Width - Coefficient of Variation	12.0	%	11.0-16.0
Leukocytes	12.76	x 10^{9}/L	4.00-11.00
Neutrophils	73.8	%	53.8-69.8
Eosinophils	0.5	%	0.6-4.6
Basophils	0.3	%	0.0-1.5
Lymphocytes	21.2	%	22.6-36.6
Monocytes	3.8	%	4.7-8.7
Immature Granulocytes	0.4	%	< 0.5
Platelet Count	245	x 10^{9}/L	150-400
IMMUNOGLOBULINS			
Immunoglobulin G	728	mg/dL	650-1500
Immunoglobulin A	202	mg/dL	78-312
Immunoglobulin M	43	mg/dL	55-300
Immunoglobulin E	4	kU/L	< 114
COMPLEMENT			
Complement Fraction C3c	113	mg/dL	83-177
Complement Fraction C4	48	mg/dL	dez/36

TEST	RESULT	UNIT	REFERENCE INTERVAL
SERUM PROTEIN ELECTROPHORESIS			
Total Proteins	66.7	g/L	64.0-83.0
Albumin	57.6	%	49.7-64.4
Alpha 1 Globulin	6.9	%	4.8-10.1
Alpha 2 Globulin	12.8	%	8.5-15.1
Beta Globulin	12.6	%	7.8-13.1
Gamma Globulin	10.1	%	10.5-19.5
AUTOIMMUNITY			
Anti-Nuclear Antibodies (Indirect Immunofluorescence) Titration	Negative		< 1/160
Anti-Double Stranded Deoxyribonucleic Acid Antibody	< 10.0	International Units/mL	< 100.0
Anti-Neutrophil Cytoplasmic Antibodies Proteinase 3 (Enzyme-Linked Immunosorbent Assay)	< 2	Units/mL	< 20
Anti-Neutrophil Cytoplasmic Antibodies Myeloperoxidase (Enzyme-Linked Immunosorbent Assay)	< 2	Units/mL	< 20
Rheumatoid Factor	< 9.7	International Units/mL	< 30.0
Extractable Nuclear Antigen Screening (Enzyme-Linked Immunosorbent Assay)	Negative		Negative
GENERAL CHEMISTRY			
Total Proteins	74.9	g/L	64.0-83.0
Albumin	43.6	g/L	38.0-51.0
Aspartate Aminotransferase	18	U/L	out/31
Alanine Aminotransferase	12	U/L	out/31
Gamma-Glutamyl Transferase	23	U/L	jul/32
Alkaline Phosphatase	49	U/L	30-120
Total Bilirubin	0.75	mg/dL	< 1.20
Direct Bilirubin	0.14	mg/dL	< 0.40
Amylase	54	U/L	22-80
Lipase	33	U/L	jul/60
Lactate Dehydrogenase	152	U/L	135-225
Creatine Kinase	57	U/L	10-149
Glucose	78	mg/dL	75-110
Urea	20	mg/dL	10.00-50
Creatinine	0.70	mg/dL	0.51-0.95
Estimated Glomerular Filtration Rate (CKD-EPI)	116	mL/ min/1.73 m^2	> 90
Sodium	136	mEq/L	135-147
Potassium	3.9	mEq/L	3.5-5.1
Chlorides	102	mEq/L	101-109
C-Reactive Protein	15.4	mg/L	< 3.0
Ferritin	212.5	ng/mL	20.0-250.0
URINALYSIS			
Density	1.025		1.002-1.030
pH	6.0		5.0-8.0
Glucose	Negative	mg/dL	<100
Cetonic bodies	80	mg/dL	<10
Urobilinogen	2	mg/dL	<1.0
Bilirrubin	Negative	mg/dL	
Nitritis	Negative		
Proteins	0.15	g/L	<0.15
Albumin	30.0	mg/L	<30
Creatinine	2000	mg/L	
Ratio Protein/Creatinine	<0.15	g/g creatinine	<0.15
Ratio Albumin/Creatinine	<30.0	mg/g creatinine	<30

TEST	RESULT	UNIT	REFERENCE INTERVAL
Leukocytes	51.8	/µL	<30
Erythrocytes	22.4	/µL	<27
OCASIONAL URINE			
Total Proteins	0.15	g/L	<0.15
Albumin	17.5	mg/L	<20.0
Creatinine	2866	mg/L	160-3270
Ratio Albumin/Creatinine	6.1	mg/g creatinine	<30.0
ENDOCRINOLOGY (Urine)			
β-hCG	<1.20	mUI/mL	0.00-5.00
SEROLOGY			
Cytomegalovirus Antibodies - IgG	< 1.0	AU/mL	Negative: < 6.0
Cytomegalovirus Antibodies - IgM	Negative		Negative
Epstein-Barr Virus Antibodies - IgM	Negative		Positive ≥ 1.1 RU/mL
Epstein-Barr Virus Antibodies - IgG	Positive 6.0 RU/mL		Positive ≥ 1.1 RU/mL
Serodiagnosis of Syphilis (*Treponema pallidum* TPPA)	Negative		Negative
MOLECULAR BIOLOGY (Pharyngeal Exudate)			
Neisseria gonorrhoeae DNA	Negative		Negative
Chlamydia trachomatis DNA	Negative		Negative
MOLECULAR BIOLOGY (Rectal Exudate)			
Chlamydia trachomatis DNA	Negative		Negative
MOLECULAR BIOLOGY (Endocervical Exudate)			
Chlamydia trachomatis DNA	Negative		Negative
Neisseria gonorrhoeae DNA	Negative		Negative
Trichomonas vaginalis DNA	Negative		Negative
BACTERIOLOGICAL EXAMINATION (Urine)			
Cultural Exam	Negative		Negative

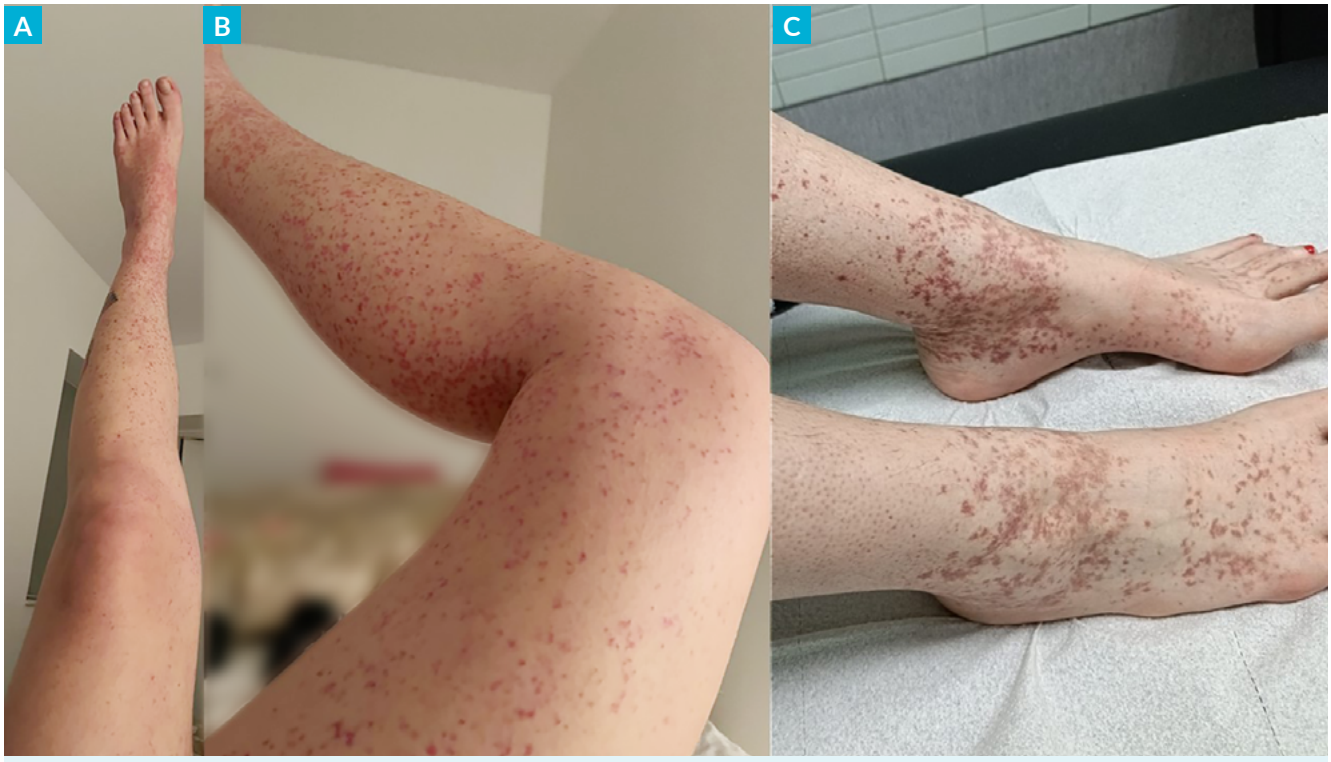

FIGURE 1. Progression of palpable purpura in an adult-onset case of Henoch-Schönlein purpura
1A and B. Initial presentation: Palpable purpura on the lower limbs observed at the primary care consultation, before any diagnosis and/or treatment.

DISCUSSION

Henoch-Schönlein purpura is the most common systemic vasculitis in children, but there are few reports presenting HSP in female adults.[5,6]

HSP diagnosis remains clinical but requires a high index of suspicion, particularly in adults. According to both the EULAR and European consensus-based SHARE recommendations, the presence of palpable purpura (not due to thrombocytopenia) in combination with at least one of the following - abdominal pain, arthritis or arthralgia, renal involvement or biopsy-proven leukocytoclastic vasculitis with predominant IgA deposition - supports the diagnosis of IgA vasculitis.[7–9]

While HSP is primarily identified through clinical presentation, atypical instances may necessitate the use of laboratory tests and imaging. Beyond standard laboratory analyses, a comprehensive immunological blood panel, including assessments for ANA, ANCA, rheumatoid factor and factors VIII and XIII, might be required to corroborate the diagnosis and exclude alternative underlying conditions. Imaging, such as renal or skin biopsies, can also be considered in situations where the diagnosis remains unclear or for surveillance of potential complications and systemic involvement.[8,9]

Although skin biopsy with direct immunofluorescence may support the diagnosis of IgA vasculitis by demonstrating IgA-dominant immune complex deposition in small vessels, its diagnostic yield is variable and influenced by several factors, particularly the timing of the procedure and lesion selection. While IgA deposition can be identified in the majority of clinically confirmed cases (75%-100%), a consistent subset of patients has been described in whom direct immunofluorescence fails to demonstrate vascular IgA deposits.[10,11] In cutaneous vasculitis, the temporal relationship between lesion onset and biopsy is particularly relevant, as the detectability of immunoreactant deposits is higher in early lesions and progressively decreases as lesions evolve or resolve. Indeed, one study demonstrated a marked reduction in direct immunofluorescence positivity for immunoglobulin deposits, from approximately 85% when biopsies were performed within the first 7 days of lesion onset, to around 14% when obtained during the second week.[10] In adult patients, biopsies obtained from older lesions may therefore show non-specific histopathological changes, as observed in this case. Consequently, negative immunofluorescence findings should be interpreted cautiously and always in conjunction with the overall clinical presentation, as they are not mandatory to establish the diagnosis according to the classification criteria.[8]

The disease is usually self-limited, so the approach is largely supportive, including symptom relief (analgesia, rest and eventually compression stockings) and vigilant monitoring for complications.[3,12] Cutaneous manifestations typically do not require specific management; however, corticosteroids can be beneficial in suppressing the inflammatory response. An oral prednisolone regimen of 1-2 mg per kg daily for two weeks might be effective in symptomatic relief, especially abdominal and joint symptoms, despite not preventing the onset of renal involvement.[3,6,12,13] In cases of relapsing or corticosteroid-dependent IgA vasculitis, particularly in adults, second-line immunosuppressive agents may be considered. Colchicine has been used mainly for recurrent cutaneous manifestations and arthralgia, with a favorable safety profile. Other options described in refractory disease include azathioprine, methotrexate or mycophenolate mofetil, especially in patients with persistent systemic involvement. In severe or organ-threatening disease, such as progressive renal involvement, calcineurin inhibitors, cyclophosphamide or rituximab have been reported in selected cases and observational studies.[12] The choice of therapy should be individualized, taking into account disease severity, organ involvement and recurrence pattern.[3,6,12,13]

This case highlights the fact that IgA levels may be within normal range in a subset of patients and that negative autoimmune or serological panels do not exclude the diagnosis.[7] Similarly, skin biopsy and direct immunofluorescence may be non-diagnostic in some cases, particularly in adult patients or when performed on non-early lesions, and should be interpreted in the context of the overall clinical picture. Recent consensus recommendations emphasize a treat-to-target approach for IgA vasculitis, including the early initiation of systemic corticosteroids in patients with significant gastrointestinal or renal manifestations, aiming to prevent progression and organ damage.[14]

This case also underscores the diagnostic challenges posed by adult-onset Henoch-Schönlein purpura, particularly when the initial clinical presentation is non-specific. The absence of skin findings and the lack of a systematic full body physical examination during the

initial ED visit contributed to a delay in diagnosis. Only after a systematic reassessment in the PHC setting was the correct diagnostic hypothesis raised.

Primary Care can play a central role in the early detection of systemic diseases, especially in young adults who may initially present with vague or non-specific symptoms. In this case, the continuity of care and clinical acumen demonstrated in the second evaluation at the PHC clinic were decisive in the identification of an underlying systemic process.

CONCLUSION

This case illustrates how a rare systemic condition in adults like Henoch-Schönlein purpura, can be initially misinterpreted as a benign gastrointestinal issue, especially in young adults without significant comorbidities. It highlights the importance of the family physician's role in maintaining a holistic, system-wide view of the patient and the value of a meticulous physical examination. The case also reinforces the utility of structured referral and communication between levels of care, ensuring that findings from PHC evaluations are integrated into the diagnostic process at the hospital level.

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ARQ, AP - Follow-up consultations, literature review and manuscript writing

CBF - Initial patient assessment, manuscript revision

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REFERENCES

1. González-Gay MA, López-Mejías R, Pina T, Blanco R, Castañeda S. IgA Vasculitis: Genetics and Clinical and Therapeutic Management. Curr Rheumatol Rep. 2018;20:24. doi: 10.1007/s11926-018-0735-3.
2. Saulsbury FT. Epidemiology of Henoch-Schönlein purpura. Cleve Clin J Med. 2002;69 Suppl 2:SII87-9. doi: 10.3949/ccjm.69.suppl_2.sii87.
3. Audemard-Verger A, Pillebout E, Guillevin L, Thervet E, Terrier B. IgA vasculitis (Henoch-Shönlein purpura) in adults: Diagnostic and therapeutic aspects. Autoimmun Rev. 2015;14:579-85. doi: 10.1016/j.autrev.2015.02.003.
4. The CARE guidelines: consensus-based clinical case reporting guideline development | BMJ Case Reports. [Accessed January 20, 2026]. Available at: https://casereports.bmj.com/content/2013/bcr-2013-201554
5. Sood R, Parekh P, Raj N, Saani I. A case report on an adult presentation of Henoch-Schönlein purpura. Cureus. 2022;14:e26385. doi: 10.7759/cureus.26385.
6. Camacho-Péreza LC, Rubio-Rivera M, Camacho-Péreza LC, Rubio-Rivera M. Schönlein-Henoch purpura in adults: Report of four cases. Rev Colomb Reumatol. 2023;30:72-7. doi:10.1016/j.rcreu.2021.05.012
7. Ozen S, Marks SD, Brogan P, Groot N, de Graeff N, Avcin T, et al. European consensus-based recommendations for diagnosis and treatment of immunoglobulin A vasculitis-the SHARE initiative. Rheumatology. 2019;58:1607-16. doi: 10.1093/rheumatology/kez041.
8. Ruperto N, Ozen S, Pistorio A, Dolezalova P, Brogan P, Cabral DA, et al. 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;69:790-7. doi: 10.1136/ard.2009.116624

9. Hočevar A, Rotar Z, Jurčić V, Pižem J, Čučnik S, Vizjak A, et al. IgA vasculitis in adults: the performance of the EULAR/PRINTO/PRES classification criteria in adults. Arthritis Res Ther. 2016;18:58. doi: 10.1186/s13075-016-0959-4.

10. Nandeesh B, Tirumalae R. Direct immunofluorescence in cutaneous vasculitis: experience from a referral hospital in India. Indian J Dermatol. 2013;58:22-5. doi: 10.4103/0019-5154.105280.

11. oudel P, Adams SH, Mirchia K, Jain H, Eranki A. IgA negative immunofluorescence in diagnoses of adult-onset Henoch-Schönlein purpura. Proc. 2020;33:436-7. doi: 10.1080/08998280.2020.1770526.

12. Weiss PF, Feinstein JA, Luan X, Burnham JM, Feudtner C. Effects of corticosteroid on Henoch-Schönlein purpura: a systematic review. Pediatrics. 2007;120:1079-87. doi: 10.1542/peds.2007-0667.

13. Castañeda S, Quiroga-Colina P, Floranes P, Uriarte-Ecenarro M, Valero-Martínez C, Vicente-Rabaneda EF, et al. IgA Vasculitis (Henoch-Schönlein Purpura): An Update on Treatment. J Clin Med. 2024;13:6621. doi: 10.3390/jcm13216621.

14. Abu-Zaid MH, Salah S, Lotfy HM, El Gaafary M, Abdulhady H, Tabra SAA, et al. Consensus evidence-based recommendations for treat-to-target management of immunoglobulin A vasculitis. Ther Adv Musculoskelet Dis. 2021;13:1759720X211059610. doi: 10.1177/1759720X211059610.