Acad Med J 2025;5(Suppl 1):S127-S132.
UDC: 616.2-022.782.11-097-053.2
https://www.doi.org/10.53582/AMJ2551127a
Case Report

A CASE OF RECURRENT CHLAMYDIA PNEUMONIAE INDUCED HENOCH-SCHÖNLEIN PURPURA IN 8-YEAR-OLD BOY

Adjievska Nevenka[1], Risteska Kristina[1,2], Pollozhani Nora[1], Rushiti Mehmeti Kujtime[1], Boshkovska Spasovska Ivona[1,3], Popovski Tomche[1]

[1]University Clinic for Dermatology, Faculty of Medicine, Ss. Cyril and Methodius University in Skopje, Republic of North Macedonia
[2]Department of Dermatovenerology, General Hospital - Ohrid, Ohrid, Republic of North Macedonia
[3]PHI Ss. Cyril and Methodius University Polyclinic, Skopje, Republic of North Macedonia
e-mail: nadzievska@yahoo.com

Abstract

Henoch-Schönlein purpura (HSP), synonymous with the recent term immunoglobulin A vasculitis (IgAV), is an acute autoimmune IgA-mediated disorder classified under the group of small-vessel vasculitis (SVV). It clinically manifests with the pathognomonic tetrad of palpable purpura, abdominal pain, hematuria or proteinuria, and arthritis. Although the full spectrum of clinical features is not invariably present and additional clinical manifestations may be seen, cutaneous manifestations are observed in 100% of cases, thereby serving as the one most definitive diagnostic criterion.

Young children, predominantly between ages of 2 and 10, are generally affected by the disease (90% of the cases), thereby establishing it as the most common vasculitis of childhood.

We describe a case of two episodes of *Chlamydia pneumoniae* induced HSP in an 8-year-old boy with an acute onset of a rash and an upper respiratory tract infection preceding the onset, and with possible simultaneous multi-factorial infective etiology contributing to the severity of the disease.

Given the rarity of reported cases, only two to our knowledge, it is challenging to establish a definitive causal relationship between *Chlamydia pneumoniae* and HSP.

However, this uncommon association highlights the importance of considering this atypical pathogen in the etiological differential diagnosis of HSP, especially in patients presenting with preceding respiratory symptoms. Early diagnosis is crucial for preventing complications and ensuring optimal management, with prompt recognition of diverse triggers, such as infections, being essential for tailored therapy and improved patient outcomes.

Keywords: Henoch-Schönlein purpura (HSP), IgA vasculitis (IgAV), *Chlamydia pneumoniae*, atypical pneumonia, children

Introduction

Henoch-Schönlein purpura (HSP), synonymous with the recent term immunoglobulin A vasculitis (IgAV), is an acute autoimmune IgA-mediated disorder classified under the group of small-vessel vasculitis (SVV). This disease is characterized by generalized vasculitis affecting the skin, gastrointestinal tract, kidneys, and joints, and, as a result, clinically manifests with the pathognomonic tetrad of palpable purpura, abdominal pain, hematuria, and arthritis. Although the full spectrum of clinical features is not invariably present and additional clinical manifestations may be seen, cutaneous manifestations are observed in 100% of cases, thereby serving as the one most definitive diagnostic criterion.

Young children, predominantly between ages of 2 and 10, are generally affected by the disease (90% of the cases), thereby establishing it as the most common vasculitis of childhood[1]. Adults can rarely be affected, carrying higher risks of more severe presentations and complications of the disease. In this age group, it is generally associated with worse outcomes[2].

The exact etiology remains unclear, but in the most well-documented cases, it is subsequent to upper respiratory tract infections and occasional reports attributable to various noxae such as streptococci, mycoplasma and variety of viruses, but, in general, no organism is consistently linked with the disease occurrence. The symptoms and signs triggered from the pathogenic factor take days to weeks to develop. Moreover, the administration of medications, immunization procedures, and the presence of malignant neoplasms are additionally considered as potential contributing factors[3]. This suggests that the individual immune response is pivotal in the development of the disease, and evidence also support a genetic predisposition, as demonstrated in parents, offspring, and siblings of affected individuals, with episodes occurring years apart.

The recent genome-wide association studies have established a robust association between Henoch-Schönlein purpura (HSP) and HLA class II haplotypes, notably DQA101:01, DQB105:01, and DRB1*01:01, suggesting that antigen presentation pathways may underlie disease susceptibility[4]. Concurrently, polymorphisms in genes regulating endothelial function, the renin-angiotensin system (RAS), and innate immunity (MEFV) have been identified as potential modifiers of HSP risk[5,6].

We describe a case of two episodes of *Chlamydia pneumoniae* induced HSP in an 8-year-old boy with an acute onset of a rash and an upper respiratory tract infection preceding the onset, and with possible simultaneous multi-factorial infective etiology contributing to the severity of the disease.

The diagnostic criteria of childhood HSP were developed in 2005 and revised in 2010 by the European League Against Rheumatism, the Paediatric Rheumatology International Trials Organization and the Paediatric Rheumatology European Society (EULAR/PRINTO/PRES) resulting in a high sensitivity (100%) and specificity (87%) in differentiating IgAV from other forms of vasculitis[7]. The previously mentioned involves fulfilling well-defined criteria including mandatory presence of cutaneous lesions such as purpura or petechiae with lower limb predominance in the absence of thrombocytopenia, along with at least one of the following: acute onset diffuse abdominal colicky pain, acute onset arthritis or arthralgia, renal involvement (either hematuria or proteinuria), or biopsy showing IgA deposits at any anatomical location[8].

The diagnostic significance of IgA immune deposits in small vessel walls (usually postcapillary venules), detected via direct immunofluorescence, remains incompletely defined, as similar deposits are also found in other forms of cutaneous vasculitis. Handler *et al*. demonstrated that while IgA immune vascular deposits are sensitive, they lack specificity for diagnosis of the disease. However, their presence, when combined with relevant clinical data (outlined earlier), enhances diagnostic accuracy[1].

Although the majority of cases are self-limiting, with an average disease duration of approximately 4 weeks, misdiagnosis may lead to adverse outcomes, particularly when an infectious etiology is involved, as such cases could benefit from targeted therapy.

Case description

An 8-year-old male foreign patient presented to the University Clinic for Dermatology with palpable purpuric rash on lower extremities and abdomen that appeared one week after a single febrile episode (39°C), sore throat, and dry cough. The patient's mother additionally provided photographic documentation, revealing erythematous wheals localized to the upper extremities (Figure 1, A) that manifested the initial rash, preceding the development of the characteristic palpable purpuric lesions.

Initial management was provided in the patient's home country, and it consisted of antipyretic therapy with paracetamol and ibuprofen. Seven days after the onset of symptoms, the patient developed joint pain, particularly in the knees, associated with mild edema resulting in difficulty in weight-bearing, leading to limping with walking and limited mobility. The mother reported that the child was complaining of abdominal pain and had constipation. This was followed by the onset of palpable purpura on the lower extremities (Figure 1, B), leading to hospitalization for further evaluation. During his four-day stay, he received symptomatic treatment with NSAID and corticosteroids taper. Following discharge, the patient continued to experience joint pain and swelling, and was prescribed ibuprofen for pain management.

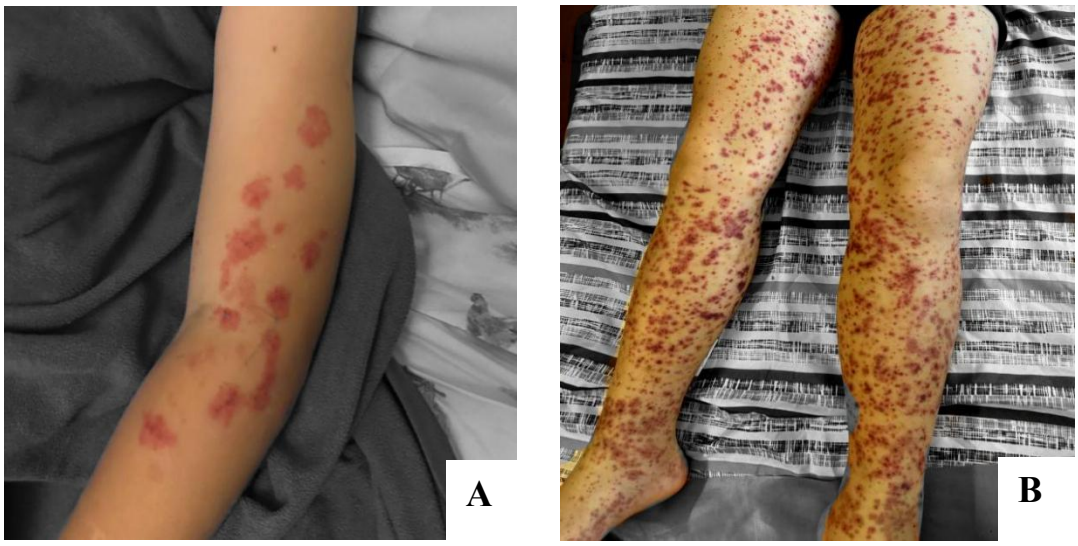

Fig. 1. A. Erythematous wheals localized to the upper extremities, **B.** Palpable purpura on the lower extremities

Upon presentation at our Dermatology Clinic, two weeks after the onset of the rash, the patient was admitted for further investigation. On physical examination at our hospital, the patient was clinically stable and with vital signs within normal limits. Pulmonary auscultation revealed wet crackles over the left lung field. Abdominal examination showed soft and painful abdomen upon palpation of the left side, with clinical suspicion of splenomegaly. The liver was neither palpable nor enlarged.

Examination of the skin revealed hemorrhagic palpable purpura predominantly localized on lower extremities, less in the gluteal region, lower abdomen, upper extremities, and only a few in facial area. The lesions on the lower extremities demonstrated a tendency to coalesce.

Laboratory tests were performed, including throat and nasal swabs, and abdominal imaging because of the persistent abdominal pain. The abdominal CT scan revealed no signs of intestinal invagination, but extensive fecal masses were observed throughout the colon. Laboratory results showed positive findings for *Mycoplasma pneumoniae* (IgM), *Chlamydia pneumoniae* (IgA and IgG), and SARS-CoV-2 IgG. Further testing revealed mild elevation in CRP (12.0mg/L) and ASO (255 U/ml). D-dimer levels were significantly elevated at 2340 ng/mL, suggesting secondary fibrinolysis, but overall hemostasis remained intact.

The patient was treated with parenteral corticosteroid therapy, oral antibiotic (tbl. Azithromycin for 3 days), gastroprotective agents, antihistamines, and vitamin supplementation. Topical corticosteroids and emollients were also applied to manage cutaneous lesions. Upon stabilization, the patient was discharged in good condition, with minimal residual skin changes and favorable laboratory parameters.

However, approximately one month later, the patient experienced a recurrence of symptoms and signs (Figure 2), including fever and arthralgia. New laboratory tests revealed the reappearance of *Chlamydia pneumoniae* (IgM and IgG), along with a marked increase in CRP (39.34 mg/L) and a slight elevation in ASO (Figure 3). Given the clinical presentation and laboratory findings, antibiotic therapy was promptly administered, resulting in rapid improvement within a few days. Upon completion of a three-month follow-up period, no evidence of recurrence of the disease was identified.

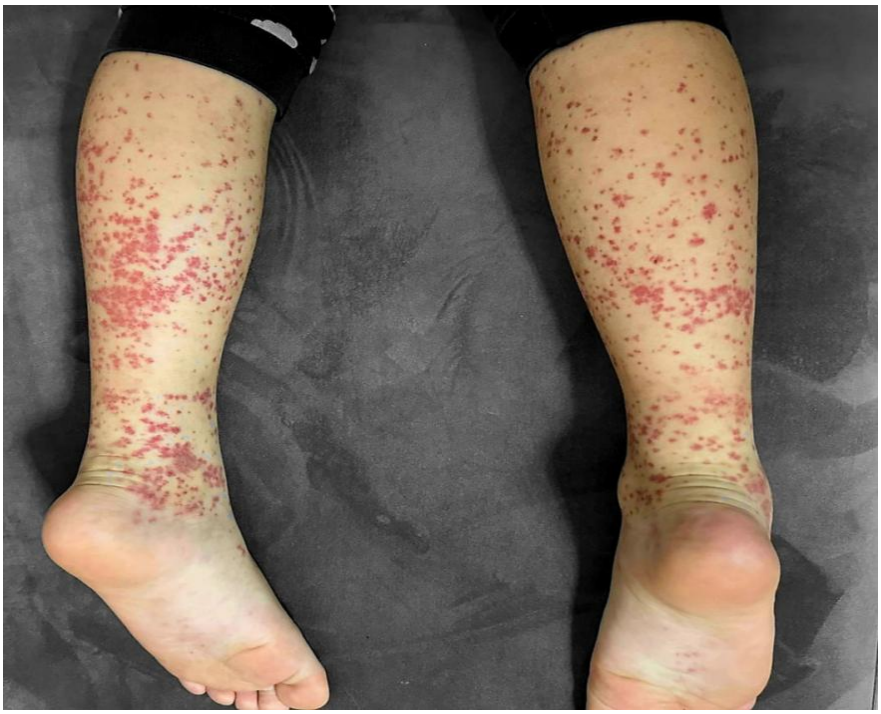

Fig. 2. Recurrence of palpable purpura on the lower extremities

CHLAMYDIE

Analiza	Rezultati	Norma	Njësia	Përshkrimi
Chlamidia Pneumoniae IgA	12.8 Negativ	< 20.00 20.10 - 25.00 > 25.00	U/mL	Negativ Intermediar Pozitiv
Chlamydia Pneumoniae IgM	> 200.0 Pozitiv	< 20.00 20.10 - 25.00 > 25.00	U/mL	Negativ Intermediar Pozitiv
Chlamidia Pneumoniae IgG	70.9 Pozitiv	< 25.00 20.10 - 25.00 > 25.00	U/mL	Negativ Intermediar Pozitiv

BIOKIMIA

Analiza	Rezultati	Norma	Njësia	Përshkrimi
CRP (Gjak)	**39.34 H**	< 6.00	mg/L	Analiza është punuar me metoden: FIA

Fig. 3. Repeated *Chlamydia pneumoniae* IgA, IgM and IgG serology results, along with C-reactive protein analysis

Discussion

This case illustrates a typical presentation of childhood HSP fulfilling the EULAR/PRINTO/PRES criteria. The patient's clinical manifestations included palpable purpura, joint pain and abdominal pain, which are consistent with the classic triad of this disease, without renal involvement or complications. Bacterial pathogens that cause atypical pneumonia have been linked to HSP as a potential causative etiologic factor. The initial presentation with accompanying multiple infections underscores the multi-factorial etiology, often observed in HSP cases.

To the best of our knowledge, based on an extensive review of the literature, including searches in PubMed, academic databases, and web-based sources, only two reported cases exist in which *Chlamydia pneumoniae* has been identified as the etiological factor triggering Henoch-Schönlein purpura[2,9]. Of the two described cases, only one involved a pediatric patient. While the role of infections, particularly respiratory ones, as a trigger for HSP is well-documented, it is important to consider other potential factors, including genetic predisposition and immune dysregulation. Furthermore, the recurrent nature of the disease in this patient highlights the need for long-term monitoring and timely treatment.

The identification of *Chlamydia pneumoniae* IgM and IgG during the second episode reinforces the hypothesis that various infections can trigger HSP flares, especially in susceptible individuals. This emphasizes the importance of early diagnosis and appropriate management, as untreated or misdiagnosed cases may lead to long-term complications.

This case shows rare etiology of *Chlamydia pneumoniae* in the development of HSP and a pathogenesis that has yet to be investigated. However, the presence of concurrent infections highlights the complex interplay between infections, so it is worth considering both timing and severity of the infection in relation to symptom onset. The individual immune response likely predisposes certain individuals to the disease.

Given the rarity of reported cases, it is challenging to establish a definitive causal relationship between *Chlamydia pneumoniae* and HSP[2]. However, this uncommon association highlights the importance of considering this atypical pathogen in the etiological differential diagnosis of HSP, especially in patients presenting with preceding respiratory symptoms. Early diagnosis is crucial for preventing complications and ensuring optimal management. Prompt recognition of diverse triggers, such as infections, is essential for tailored therapy and improving patient outcomes.

Conclusion

Chlamydia pneumoniae can be considered as a pathogen that is rarely associated with HSP. Although the current literature does not provide specific guidelines on the optimal duration of azithromycin treatment for the management of the disease and preventing HSP recurrence, we could consider extending the course to approximately two doses as a more effective strategy.

Conflict of interest statement. None declared.

References:

1. Watts RA, Robins J. *Oxford Textbook of Vasculitis*. Oxford University Press eBooks; 2014. doi:10.1093/med/9780199659869.001.0001.
2. Kang SA, Khalaf SA, Nelson T. Immunoglobulin A vasculitis induced by atypical pneumonia infection with *Chlamydophila pneumoniae*. *IDCases*. 2022 Jan 1;30:e01624. doi:10.1016/j.idcr.2022.e01624.
3. Wang B, Xiang W, Zhu Z, et al. The influence of respiratory infections on Henoch-Schönlein purpura in children. *BMC Infect Dis*. 2025;25:118. doi:10.1186/s12879-025-10514-4.
4. Tauriainen S, et al. HLA class II associations in Henoch-Schönlein purpura. *Arthritis Res Ther*. 2020;22(1):108. doi:10.1186/s13075-020-02180-x.
5. He P, et al. Association of nitric oxide synthase gene polymorphisms with Henoch–Schönlein purpura in Chinese children. *Eur J Pediatr*. 2017;176(8):1035–1040. doi:10.1007/s00431-017-2940-2.
6. Yang YH, et al. Genetic polymorphisms and susceptibility to Henoch-Schönlein purpura. *Genet Test Mol Biomarkers*. 2017;21(6):373–377.
7. Hetland LE, Susrud KS, Lindahl KH, et al. Henoch-Schönlein purpura: a literature review. *Acta Derm Venereol*. 2017;97(9):1160–1166. doi:10.2340/00015555-2733.
8. AlGhoozi DA, AlKhayyat HM. A child with Henoch-Schönlein purpura secondary to a COVID-19 infection. *BMJ Case Rep*. 2021;14(1):e239910. doi:10.1136/bcr-2020-239910.
9. Beltrán García S, Domínguez Cajal M, Lafuente Hidalgo M, García Sánchez N, Benito Ruesca R. Púrpura de Schönlein-Henoch y *Chlamydophila pneumoniae*. *An Pediatr*. 2008;68(2):196–197.