Anaphylactoid purpura, also known as Henoch–Schönlein purpura (HSP), is an IgA-mediated vasculitis that tends to be a benign disease of childhood. Up to 50% of cases are preceded by an upper tract respiratory infection caused by group-A beta-hemolytic streptococcus and present with the common tetrad of abdominal pain, arthritis, purpuric rash, and renal involvement. The majority of patients recover completely. Here we document a rare case of anaphylactoid purpura which manifested with skin lesions in the form of palpable purpura following a bout of acute gastroenteritis with severe dehydration; it was treated with a short regimen of steroid therapy, which resulted in the complete remission of the disease. We conclude that prompt diagnosis and multidisciplinary intervention will lead to appropriate management—consisting of the installation of early short-course steroid therapy and thus, prevent further complications and the recurrence of the disease. [PR Health Sci J 2015;34:225-227]

Key words: Henoch–Schönlein purpura, Leukocytoclastic vasculitis, Acute gastroenteritis

An 8-year-old boy presented with the following complaints: 8 to 9 episodes per day of greenish black semisolid to watery stool, mild to moderate intermittent abdominal pain, intermittent painful swelling of both his knee and ankle joints and episodes of recurrent vomiting, all of which had been occurring over the course of the month prior to his being seen. Subsequently and 4 days prior to admission, he developed cough, skin rashes over all his limbs, throat pain, and a low grade fever. On physical examination, he had a normal body temperature, a pulse rate of 78 beats per minute and a blood pressure of 80/50 mmHg. He also showed signs of severe dehydration. He was admitted for further management. A lab examination revealed that he had an elevated leukocyte count of 1.5×10⁴/µL, with neutrophilia (82%). Platelet counts and serum complement levels were within normal ranges. A urinalysis showed 10 to 15 crenated RBCs/ high-power field, with an albumin level of greater than 1.0 g/dL. The boy’s stools were positive for occult blood. Anti-streptolysin (ASO) titer was 600 IU/mL (normal: <200 IU/mL), and C-reactive

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Anaphylactoid Purpura

Anaphylactoid purpura, also known as HSP, is an IgA-mediated vasculitis due to antigen–antibody complexes resulting from bacterial (Hemophilus influenza, parainfluenza, mycoplasma, Legionella, Yersinia, Shigella and Salmonella species) and viral (adenoviruses, Epstein–Barr virus, paroviruses and varicella) infections, vaccinations (cholera, measles, paratyphoid A and B, typhoid, and yellow fever), medications (ampicillin, erythromycin, penicillin, quinidine, and quinine) or autoimmune disorders (2). It is usually a benign disease of childhood that tends to be observed following 1 to 3 weeks of an upper respiratory tract infection with group-A β-haemolytic streptococci (associated with upto 50% of all cases). It is usually an acute, self-limiting illness; one third of patients may endure 1 or more recurrences. In the Northern Hemisphere, the disease tends to be most prevalent from November through January. In the US, the prevalence is approximately 14 to 15 cases/100,000 and in the UK, said prevalence is 20.4 cases/100,000 population. In general, patients appear ill and have a body temperature of less than 38°C (100.4°F). Caucasians comprise the ethnic group most affected by HSP. Males are affected more frequently than females, with a ratio of approximately 1.5:1 to 2:1. The incidence peaks in children aged 5 years.

Although new criteria have been proposed by the European League Against Rheumatism (EULAR) and the Pediatric Rheumatology Society (PReS) for use in determining HSP, those criteria originally proposed by American College of Rheumatology remain in use in most current studies (Table 1) (3). Clinical features include cutaneous manifestations in form of skin rashes without thrombocytopenia which evolves from erythematous to urticarial and macular wheels to nonblanching palpable purpura with petechiae and ecchymoses. Palpable purpura is seen in 50% of the cases as the presenting sign. Purpuric lesions occur in groups and may persist up to 3–10 days. Classical HSP is symmetrical in distribution involving dependent areas such as the lower extremities and buttocks but it can also be seen in the upper extremities. Truncal and facial involvement can also be seen. Initially the lesions are single and less than 1 cm, but later coalesce to form ecchymotic areas. Rarely, hemorrhagic bullae, ulcerations or dermal scarring may be seen. On histopathology leukocytoclastic vasculitis, characterized by neutrophil infiltration and prominent nuclear fragmentation, involving the upper and middle layers of the dermis with IgA deposition on immunofluorescence, is seen. Angioedema (nonpitting edema) can be seen in the scalp, back, and extremities (4). Gastrointestinal manifestations occur in about 50% of all HSP cases and usually consist of colicky abdominal pain, melena, bloody diarrhea, and/or intussusception. The most serious complication is IgA nephropathy, and 2 to 5% of all patients progress to end-stage renal failure. Arthralgia is seen in 60 to 84% of cases and when such occurs, commonly affects the knees and ankles and less frequently, the wrists and fingers. Rarely, joint effusions without permanent joint deformities are observed.

### Table 1. Diagnostic criteria of anaphylactoid purpura

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<tr>
<td>Mandatory criteria</td>
<td>≥3 of the following criteria are needed</td>
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<tr>
<td>1) Palpable purpura</td>
<td>1) Age 20 years or younger at disease onset</td>
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<tr>
<td>Plus at least 1 of the following:</td>
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<tr>
<td>1) Diffuse abdominal pain</td>
<td>2) Palpable purpura</td>
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<tr>
<td>2) IgA deposition in any biopsy</td>
<td>3) Acute abdominal pain/gastrointestinal bleeding</td>
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<tr>
<td>3) Arthritis/arthralgia</td>
<td>4) Biopsy showing granulocytes in walls of small arterioles or venules in superficial layers of skin</td>
</tr>
<tr>
<td>4) Renal involvement</td>
<td>(hematuria ≥ proteinuria)</td>
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</table>

HSP is a clinical diagnosis. Patient may have mild leukocytosis with normal platelet counts, normal serum complement levels, and increased IgA (in 50% of cases). A skin biopsy will show leukocytoclastic vasculitis with IgA and C3 deposits. Different treatment options are given in Table 2 (5). Indicators of poor prognosis are being more than 8 years of age, frequent relapses,
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an elevated serum creatinine level at onset, proteinuria greater than 10 g/day, hematuria, anemia, fever on presentation, hypertension, membranoproliferative glomerulonephritis, persistent purpura above the waist, an elevated sedimentation rate, IgA with reduced IgM concentration at the time of diagnosis, and a low factor XIII level (6). Our patient fortunately responded to short course steroid therapy, in spite of his having such poor prognostic factors as being more than 8 years of age, having a fever and presenting with hematuria associated with acute gastroenteritis induced severe dehydration.

**Conclusion**

We conclude that the prompt diagnosis of anaphylactoid purpura followed by early multidisciplinary intervention constitutes appropriate management and prevents the emergence of complications and recurrence.

**Resumen**

Púrpura anafilactoide, también conocido como Henoch-Schönlein purpura (HSP). IgA es una mediada por vasculitis que tiende a ser una enfermedad benigna de la infancia. Hasta el 50% de los casos son precedidas por un tracto superior infección respiratoria causada por grupo de estreptococos beta-hemolíticos y presente con el común tétradas de dolor abdominal, artritis, erupciones purpúricas y compromiso renal. La mayoría de las pacientes se recuperan por completo. Aquí presentamos un caso poco frecuente de púrpura anafilactoide que se manifiesta con lesiones en la piel en forma de púrpura palpable tras un episodio de gastroenteritis aguda con deshidratación severa; se trata de un tratamiento corto de la terapia con esteroides, que dio lugar a la remisión completa de la enfermedad. Llegamos a la conclusión de que el diagnóstico precoz y intervención multidisciplinar dará lugar a una adecuada gestión de consistente en la instalación de curso corto de terapia con esteroides y por lo tanto, prevenir complicaciones y la reaparición de la enfermedad.

**References**