

**Gebelik Sırasında Henoch-Schönlein Purpurası: Olgu Sunumu****Henoch-Schönlein Purpura During Pregnancy: A Case Report**

Atakan TANACAN  
Fatih AKTOZ  
Gokcen ORGUL  
M. Sinan BEKSAC

<https://orcid.org/0000-0001-8209-8248>  
<https://orcid.org/0000-0003-3210-849X>  
<https://orcid.org/0000-0003-0578-4230>  
<https://orcid.org/0000-0001-6362-787X>

Division of Perinatology, Department of Obstetrics and Gynecology, Hacettepe University, Ankara, Turkey

**ÖZ**

Yirmi beş yaşında bir hasta, her iki bacakta palpe edilebilen purpura ve bipedal ödem şikayetleri ile dördüncü gestasyonel haftada hastaneye başvurdu. Hastanın 12 yaşında tanısı konulmuş olan Henoch-Schönlein purpura (HSP) hikayesi mevcuttu. Erkek kardeşine de aynı hastalık için tanı konulmuştu. Hastaya gebeliği süresince aktif müdahalede bulunulması gerekmedi. Ayrıca ciddi bir obstetrik komplikasyon da gözlenmedi. Dikkatli şekilde takip edilen HSP tanılı gebelerin başarı ile doğurtulabileceğine inanıyoruz.

**Anahtar Kelimeler:** Henoch-Schönlein purpurası, gebelik, vaskülit

**ABSTRACT**

A 25-year-old primiparous woman in the fourth gestational week with palpable purpuras on both legs and bipedal edema was admitted to the hospital. She had a history of Henoch-Schönlein purpura (HSP) that was diagnosed at the age of 12 years. Her brother had also received the same diagnosis. The patient did not necessitate active intervention during pregnancy. In addition, we did not observe severe obstetrical complications. We believe that patients with HSP undergoing careful follow-up (pregestational and gestational) may deliver successfully.

**Keywords:** Henoch-Schönlein purpura, pregnancy, vasculitis

**Introduction**

Henoch-Schönlein purpura (HSP) is an immune-mediated systemic vasculitis concomitant with immunoglobulin A (IgA), complement component 3 (C3), and fibrin deposition within the walls of the affected small vessels (1). It is the most common type of systemic vasculitis in the pediatric population, with an incidence of approximately 20 per 100,000 children, but its occurrence is less common in the adult population, with ethnic and regional alterations (2, 3).

The American College of Rheumatology developed the classification criteria for the diagnosis of HSP in 1990 (4). Diagnosis is made in 90% of patients when two or more of the following conditions are present (4): 1) palpable purpura, 2) age at onset  $\leq$  20 years, 3) acute abdominal pain, and 4) biopsy results showing granulocytes in the walls of small arterioles and/or venules. The European League Against Rheumatism and the Pediatric Rheumatology European Society together with the Pediatric Rheumatology International Trials Organization developed a different classification criterion (5). HSP is diagnosed in the presence of one or more of the following conditions with purpura (usually palpable and in clusters) and/or petechiae with lower limb predominance without thrombocytopenia or coagulopathy: 1) abdominal pain (usually diffuse with acute onset), 2) arthritis or arthralgia (acute onset), 3) renal involvement (proteinuria and hematuria), and 4) leukocytoclastic

vasculitis or proliferative glomerulonephritis with predominant IgA deposition. The Chapel Hill Consensus Group recommends a histologic criterion including IgA-dominant immune complex deposition on the walls of small vessels (6).

The exact cause of HSP has not yet been clarified, but immunologic, genetic, and environmental factors have contributed to its pathogenesis (7). Palpable purpura, arthralgia/arthritis, abdominal pain, and symptoms related to renal involvement are the main clinical characteristics. Although HSP is self-limited in most cases, renal complications are more prominent in the adult population (8).

Therapeutic targets are aimed at symptomatic therapy and management of complications because a definitive treatment for the disease is yet to be reported. Prognosis is good in the absence of severe renal complications (9).

The prognosis and management of HSP in pregnancy are debatable because only a small number of cases (21) have been reported in the literature (10-12).

**Case Report**

A 25-year-old primiparous woman, in the fourth gestational week, with palpable purpuras on both legs and bipedal edema was admitted to the hospital. She received a diagnosis of HSP at the age of 12 years. Her brother also

Yazışma Adresi/ Correspondence Address:

Atakan Tanacan, <https://orcid.org/0000-0001-8209-8248>

Division of Perinatology, Department of Obstetrics and Gynecology, Hacettepe University, Ankara, Turkey

Tel/Phone: 0312 305 18 01

E-mail: atakantanacan@yahoo.com

Geliş Tarihi : 26.10.2017

Kabul Tarihi : 14.12.2017

had the same diagnosis. The laboratory results were as follows: hemoglobin, 10.2 g/dL; hematocrit, 33%; white blood cell count,  $9.2 \times 10^9/\text{ml}$ ; and platelet count,  $290 \times 10^9/\text{ml}$ . The serum biochemistry values were within the reference range, with sedimentation, complement C3, and complement C4 at 25 mm/h, 130 mg/dL, and 20.3 mg/dL, respectively. She had proteinuria; therefore, she was referred to the nephrology and romatology departments. Her complaints regressed without medical intervention, and she was discharged from the hospital.

During follow-up, the results of combined chromosomal abnormality screening test, ultrasonographic fetal structural anomaly screening, and 50-g glucose challenge test were normal. The patient had no complaints until the 27th gestational week when she was admitted to the hospital due to nausea and vomiting. Her C-reactive protein (CRP) level was elevated at 1.37 mg/L, whereas IgA, anti-nuclear antibody, and anti-neutrophil cytoplasmic antibody levels were within the reference range. No medical treatment was administered, and she was discharged from the hospital after romatology and infectious disease consultations.

During the course of pregnancy, her blood pressure and kidney function were monitored closely, and she consulted with the perinatology, romatology, and nephrology departments at close intervals.

She was referred to the delivery room when she had regular uterine contractions at 40 weeks and 5 days of gestation. A 3380-g female neonate was delivered through spontaneous vaginal birth without any complications. The APGAR score was 9/10/10. After the delivery, her hemoglobin was 9.3 mg/dL, and her blood biochemistry values including sedimentation, CRP, complement C3–C4, IgA, and urine analysis results were all within the normal range.

## Discussion

HSP is an IgA-mediated systemic vasculitis and the most common type of systemic vasculitis in the pediatric population (1). HSP is rarely observed in adulthood; therefore, only case reports exist in scientific literature (10-12). Although the symptoms are generally non-specific, lower limb palpable purpura, abdominal pain, and arthralgia are the most common symptoms (10). Diagnosis may be challenging, particularly in cases presenting without purpura, because other symptoms are vague and can be often observed in pregnant women, depending on various reasons (10). Renal involvement (glomerulonephritis), with proteinuria and hematuria, together with headache and convulsions, may be present in some cases; therefore, establishing a differential diagnosis for preeclampsia may be difficult for physicians (12). Physicians should be cautious because renal complications are more prominent in adults, which may lead to end-stage renal disease (8).

Based on previous case reports, no consensus has been established on the prognosis or management of HSP during pregnancy. Corticosteroids may be beneficial for symptom relief; however, some severe cases required plasmapheresis (10-12). Most cases had good obstetric outcomes in the absence of renal involvement, and in some cases, only supportive care was sufficient for the management (10-12). HSP symptoms may progress or resolve during pregnancy, and a new onset of the disease may be observed (10-12). Furthermore, IgA cannot cross the placenta; therefore, the fetus is protected from the immune deposits (10-12). Notably, we did not use corticosteroids in our case.

To the best of our knowledge, a limited number of case reports have been published in this field, most probably because of the disease nature (childhood disease). In our report, the patient did not necessitate active intervention during pregnancy, and we did not observe severe complications. We believe that patients undergoing careful follow-up may deliver successfully; however, severe obstetrical complications have been reported previously (10-12). To the best of our knowledge, this report will present the 22nd case of HSP in the literature.

In conclusion, the diagnosis of HSP should be kept in mind, particularly in patients with palpable lower limb purpuras during pregnancy. Corticosteroids may be beneficial for symptomatic treatment, but supportive care may be sufficient in most cases. However, the differential diagnosis of preeclampsia should be established, and severe renal complications of the disease must be considered in the management of HSP during pregnancy.

## References

- Jennette JC, Falk R, Bacon P, Basu N, Cid M, Ferrario F, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. *Arthritis & Rheumatology*. 2013;65(1):1-11.
- Gardner-Medwin JM, Dolezalova P, Cummins C, Southwood TR. Incidence of Henoch–Schönlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. *The Lancet*. 2002;360(9341):1197-202.
- Blanco R, Martínez-Taboada VM, Rodríguez-Valverde V, García-Fuentes M, Gonzalez-Gay MA. Henoch–Schönlein purpura in adulthood and childhood. Two different expressions of the same syndrome. *Arthritis & Rheumatology*. 1997;40(5):859-64.
- Arend WP, Michel BA, Bloch DA, Hunder GG, Calabrese LH, Edworthy SM, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. *Arthritis & Rheumatology*. 1990;33(8):1129-34.
- Ozen S, Pistorio A, Iusan SM, Bakaloglu A, Herlin T, Brik R, et al. EULAR/PRINTO/PRES criteria for Henoch–Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: final classification criteria. *Annals of the Rheumatic Diseases*. 2010;69(5):798-806.
- Jennette JC, Falk RJ. Small-vessel vasculitis. *New England Journal of Medicine*. 1997;337(21):1512-23.
- Yang Y-H, Yu H-H, Chiang B-L. The diagnosis and classification of Henoch–Schönlein purpura: an updated review. *Autoimmunity Reviews*. 2014;13(4):355-8.
- Kang Y, Park J-S, Ha Y-J, Kang M-I, Park H-J, Lee S-W, et al. Differences in clinical manifestations and outcomes between adult and child patients with Henoch–Schönlein purpura. *Journal of Korean Medical Science*. 2014;29(2):198-203.
- Audemard-Verger A, Terrier B, Dechartres A, Chanal J, Amoura Z, Le Gouellec N, et al. Characteristics and management of IgA vasculitis (Henoch–Schönlein purpura) in adults: data from the 260 patients included in the IGAVAS survey. *Arthritis & Rheumatology*. 2017.
- Tayabali S, Andersen K, Yoong W. Diagnosis and management of Henoch–Schönlein purpura in pregnancy: a review of the literature. *Archives of Gynecology and Obstetrics*. 2012;286(4):825-9.

11. Kalmantis K, Daskalakis G, Iavazzo C, Vranos A, Mesogitis S, Antsaklis A. Henoch–Schonlein purpura in pregnancy. *Journal of Obstetrics and Gynaecology*. 2008;28(4):403-5.
12. Chakrabarti SB, Tigga MP, Ray J, Debbarma A. Henoch–Schonlein purpura in pregnancy: a lesser known phenomenon. *International Journal of Reproduction, Contraception, Obstetrics and Gynecology*. 2017;4(4):1227-30.