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Letter to Editor

Factor 13 Activity in Children with Henoch-Schonlein Purpura in Iranian Children

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Dear Editor

Henoch-Schonlein purpura (HSP) or immunoglobulin A (IgA) vasculitis is the most common small-vessel vasculitis in children, characterized by non-thrombocytopenic palpable purpura, arthritis, subcutaneous edema, gastrointestinal (GI) and renal involvement. The disease has usually a self-limited course with the major potential renal complications.[1]

This retrospective observational study was performed by reviewing medical records of 24 children with HSP who were admitted to Mofid Children's hospital (a tertiary referral center in Tehran, Iran). The clinical presentations, laboratory findings, and severity of the disease were evaluated. Plasma factor 13 activity was also measured by latex agglutination immunoturbidity method. The Diagnosis was made according to the European League Against Rheumatism/Pediatric Rheumatology International Trials Organization/ Pediatric Rheumatology European Society (EULAR/ PRINTO/PRES) criteria for HSP.[2] In addition, a severity scoring system was used for determining the disease severity. This scoring system is based on a 4-point scale (0-3) according to the distribution of purpura, joint symptoms (arthralgia, arthritis), abdominal problems (abdominal pain, stool occult blood), and renal complications (proteinuria, hematuria). The severity was classified to mild, moderate, and severe regarding the total scores of 1-5, 6-10, and >10, respectively.[3]

In this study, cutaneous (100%), GI (66.7%), renal (37%), and articular (29.2%) involvement were the most common manifestations of the disease. The most common laboratory findings were as follow: elevated C-reactive protein (CRP) (45.8%), elevated erythrocyte sedimentation rate (ESR) (37.5%), thrombocytosis (29.2%), and leukocytosis (16.7%). The frequency of mild, moderate, and severe disease was 45.85%, 29.15%, and 25%, respectively. Factor 13 activity in all of the 24 patients with each each score of severity and organ involvement including GI and renal was normal.

A correlation between the severity of the HSP and GI involvement, and rapid decline of factor 13 has been reported.[4, 5] Kamitsuji and colleagues demonstrated a significant decrease in factor 13 activity and its antigenic determinants with a further decrease in gastrointestinal complications. They prescribed factor 13 concentrate which led to relief of pain and gastrointestinal bleeding.[4] Furthermore, Henriksson and colleagues reported a decreased level of factor 13 in 13 out of 17 children with HSP. Treatment with factor 13 concentrate combined with the antifibrinolytic drug in one of them controlled severe GI bleeding.[5] The reduced factor 13 activity may be used as a prognostic or diagnostic marker. There are a few reports of successful treatment of severe GI involvement in patients with HSP with factor 13 replacement therapy.[6, 7] It is noted that decreased factor 13 activity is reported to be associated with increased risk of nephritis.[8, 9]

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We evaluated the activity of coagulation factor 13 in 24 children with HSP with a variety of disease severity in terms of GI, renal and other systemic manifestations. Despite previous studies, we did not find any abnormality in activity of factor 13 and there was not any association between factor 13 and severity of the disease according the severity score previously mentioned.[3]

It could be hypothesized that the race may be a determining factor in creating this difference. Also, the sample size of this study was too small to reach a solid conclusion.

Abbreviations: HSP: Henoch-Schonlein purpura, IgA: immunoglobulin A, GI: gastrointestinal, EULAR/PRINTO/PRES: European League Against Rheumatism/ Pediatric Rheumatology International Trials Organization/Pediatric Rheumatology European Society, SPSS: Statistical Package for the Social Sciences.

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Ethics and Consent to Participate

The study protocol was approved by the Ethics Committee of Shahid Beheshti University of Medical Sciences and registered as IR.SBMU.MSP.REC.1398.252 and in accordance with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.All patients' data were regarded as confidential.

Author's contribution

VJP study concepts; study design; definition of intellectual content; literature research; clinical studies; supervision of statistical analysis; manuscript writing. AK acquisition of medical records and clinical data; literature research. RS guarantor of integrity of the entire study; study concepts; manuscript editing. All authors read and approved the final manuscript.

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Conflict of Interest: None declared.

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