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Henoch-Schonlein Purpura in childhood: a tertiary care hospital experience in Turkey

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ABSTRACT

Background: This study examines the clinical and laboratory features of children with Henoch-Schönlein purpura. **Methods:** A retrospective study was applied for the 32 Henoch-Schönlein purpura patients who were monitored between January 2014 and June 2017 at the Pediatric clinic of a tertiary care hospital. The diagnoses were made based on the HSP criteria of the American College of Rheumatology. Age, gender, clinical and laboratory findings of the patients were recorded.

Results: The mean age of the 32 Henoch-Schönlein purpura patients was 9.29±3.29 years. Complaints at the time of admission were as follows: 93.8% of the patients (n=30) had rash; 40.6% (n=13) had abdominal pain; and 59.4% (n=19) had joint involvement and inability to walk. The results of fecal occult blood test examined during the admission were found as negative for the 75% (n=25) of patients and positive for the 19% (n=6) of patients. According to the urinary specimens, which were considered as the evidence of renal involvement at the applications, proteinuria was found negative for the 27 (8.4%) patients and positive for the 5 (1.6%) patients; hematuria was negative for the 29 (90.6%) children and positive for the 3 (%9.4) children. Antihistamine and non-steroidal anti-inflammatory were started as the treatment to the patients. During the follow-up, the 15.5% of patients had positive fecal blood tests although their test results were negative at the admission. During the clinical follow-up, the presence of proteinuria was found positive for the 6.3% of patients who had negative proteinuria test at their admissions.

Conclusions: Henoch-Schönlein purpura is a childhood vasculitis that manifests itself with rash and joint findings. It can cause significant complications due to gastrointestinal and genitourinary system involvement.

Keywords: Gastrointestinal tract, Henoch-Schönlein purpura, Renal involvement, Vasculitis

INTRODUCTION

Henoch-Schönlein purpura (HSP), which was defined at the beginning of the 19th century, is the most common vasculitis of childhood observed with arthritis, nontrombocytopenic purpura, abdominal pain and kidney involvement.¹ The etiology of this disease, which mostly holds and damages small veins, is not fully known. It is

often a self-limiting disease that affects the skin, gastrointestinal tract (GIT), joints, kidneys, and rarely other organs. Testicular involvement or kidney involvement, which leads to nephrotic syndrome, can be seen.^{2,3} Clinically, most patients with HSP have non-thrombocytopenic palpable purpuras on their hips and lower extremity skins. Arthritis and edema at the joints; abdominal pain due to gastrointestinal tract involvement; and hematuria and proteinuria due to kidney involvement

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can also be observed.³ The HSP criteria of the American collage of Rheumatology used in the diagnosis of the disease were defined as follows:

- the purpura, which is palpable and is not with thrombocytopenia;
- emergence of initial symptoms under the age of twenty;
- abdominal pain (common abdominal pain, ischemia in the intestines, bloody diarrhea);
- granulocytes on the vein wall in the biopsy. If a patient has at least two of these four criteria, the HSP diagnosis can be reported.⁴

In addition, the HSP diagnosis can also be made if diffused abdominal pain, IgA deposits in biopsy, or arthritis and renal involvement is observed together with the palpable purpura, which is the diagnostic criteria of the European Rheumatology Union.⁵

Patients with HSP usually benefit from symptomatic supportive treatment, but those who had gastrointestinal involvement recover with steroid therapy without any sequela. Renal and gastrointestinal involvements can rarely cause morbidity and mortality.^{6,7}

In this study, we aim to compare the HSP patients' demographic features; clinical and laboratory findings at the time of their applications; and laboratory findings during their follow-ups.

METHODS

This cross sectional study was initiated after obtaining the research permission from the Clinical Research Ethics Committee of Afyon Kocatepe University (Number: 304 Date: 01.12.2017). A total amount of 36 patients with HSP who were monitored between January 2014 and June 2017 at the Pediatric Clinic of Afyon Kocatepe University Faculty of Medicine were evaluated retrospectively. The diagnosis was made based on the HSP criteria of the American College of Rheumatology. American Collage of Rhematology's HSP diagnostic criteria are:

- Palpable, purpura not associated with thrombocytopenia,
- Appearance of the first symptoms before 20 years of age,
- Abdominal pain (common abdominal pain, ischemia in the bones, bloody diarrhea),
- Granulocytes in the biopsied vessel wall, At least two of them were diagnosed with HSP.

Four patients were excluded owing to incomplete medical charts. The remaining 32 patients who met the American Collage of Rhematology's HSP diagnostic criteria were included. According to the hospital records of the patients examined retrospectively, we considered the complaints at the time of admission, gastrointestinal, kidney and joint

involvements, durations of hospitalization and the course of the disease. The age, sex, clinical findings, laboratory findings of the patients were recorded.

Statistical analysis

Percentage and frequency values were calculated as descriptive statistics for categorical data. For quantitative data, mean±standard deviation values were calculated. SPSS (SPSS V 20.0 IBM, NY, USA) package program was used for statistical analysis.

RESULTS

A total amount of 32 children with HSP were included in this study; the 21 were female (65.6%) and 11 were male (34.4%). The mean age of the patients was 9.29 ± 3.29 (6-12.5 years) years. Complaints at the time of admission were as follows: the 93.8% of patients (n=30) had rash; 40.6% (n=13) had abdominal pain; and 59.4% (n=19) had joint involvement and inability to walk. After the classification of complaints, there were 5 (15.62%) patients who had only rash; 2 (6.25%) patients complained for abdominal pain alone; 13 (40.62%) patients who had rash and swelling on their joints; 5 (15.62%) patients complained for rash and abdominal pain; and 7 (21.87%) patients who had rash, swelling on joints and abdominal pain. The results of fecal occult blood test examined (Figure 1) during the admission were found as negative for the 75% (n=25) of patients and positive for the 19% (n=6) of patients.



Figure 1: Non-thrombocytopenic palpable purpuras on right lower extremity skin.

During the applications, proteinuria was found negative for the 27 (84.4%) patients and positive for the 5 (15.6%) patients; hematuria was negative for the 29 (90.6%) children and positive for the 3 (9.4%) children. The mean duration of hospitalization was 3.16±2.02 days. Antihistamine and non-steroidal anti-inflammatory were applied as the treatment to the patients. During the follow-up, fecal blood tests constantly stayed negative for the 37.5% of patients. However, the 15.5% of patients

had positive fecal blood tests although their test results were negative at the admission. In addition, the 47% of patients had negative fecal blood tests although their test results were positive at the admission. During the clinical follow-up, the presence of proteinuria perpetually stayed negative for the 71.9% of patients. However, it changed into positive for the 6.3% of patients who had negative proteinuria test at their admissions. The presence of proteinuria stayed constantly positive for the 3.1% of patients. It turned into negative for the 9.4% of patients who had positive at their admissions.

DISCUSSION

HSP is the most commonly seen vasculitis among children. Although its reasons are mostly unknown, infections, medications, immunizations, insect bites and some foods are proposed as responsible in the etiology. The HSP is generally seen more frequently among males, on the contrary, some studies indicate that it occurs more frequently among females. 8-11 In present study, the 65% of patients were females.

In Turkey, the most common age range of HSP was found as 5-15 years old and the average age of onset was reported as 7-8.5 years old.^{8,12} In this study, the mean age of the patients was 6-12.5 years old.

The first finding of HSP is the skin involvement, usually in the form of a palpable purpuric rash.¹ In current study, the application complaint of the almost all patients was detected as rash. The joints are the most frequently held area after the skin involvement. The incidence percentage of joint involvement among the children with HSP was reported as 50%-80%.^{2,13} In present study, more than half of the patients (59.4%) had joint involvement and they recovered without any sequela.

The gastrointestinal tract involvement is observed among the two-thirds of the HSP patients. Vomiting, severe abdominal pain, hematuria, hematocycosis, melena, massive hemorrhage, obstruction or perforations are the clinical signs of HSP. This situation is thought as the result of edema and hemorrhage on the vein wall resulting from mesenteric vasculitis. 1,2,14 In the international literature, the gastrointestinal involvement rate is reported as 50%-75%, and intestinal perforation or obstruction rate is found as 4%-5%. In Turkey, the gastrointestinal involvement rate is 38%-60%, fecal occult blood rate is reported as 9.6%-40.3%, and intestinal perforation or obstruction rate is 2.6%. 13,15 In present study, the abdominal pain rate was 40% and the fecal occult blood test was positive for the 20% of patients. In our series, patients with gastrointestinal track involvement were also followed by the pediatric surgery and no urgent surgical intervention was required. Kidney investments among the patients with HSP can be seen in a wide range such as fastly developed and end-stage renal failures due to microscopic hematuria and mild proteinuria. Renal involvement rate is reported between 10%-60% in various publications.¹⁶ In our country, the mean rate of renal involvement is reported as 20%, the incidence of hematuria rate is 20%-38.6% and the presence of proteinuria rate is 18%-68%.^{8,10,12}

In the study of Bayrakci et al, renal involvement was observed among the 18.7% of 96 patients, who had normal urine examination and did not receive corticosteroid treatment.¹⁷ In our study, proteinuria was positive among the 15.6% of patients and hematuria was positive for the 9.4% of patients at their admissions. During the follow-up, the presence of proteinuria changed into positive for the 6.3% of patients who had negative proteinuria test at their admissions.

CONCLUSION

In present study, the most frequent complaint of children with HSP was rash and joint findings. Considering the patient applications, it was observed that at least two systems of patients were affected. During the follow-up of inpatients, fecal occult blood tests and the presence of proteinuria in the urine changed into positive from negative. Considering this situation, patients with HSP should be followed up regularly by pediatric gastroenterology and pediatric nephrology in terms of positivities of fecal occult blood and proteinuria in the urine.

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