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ORIGINAL ARTICLE

Epidemiological Study of Henoch-Schonlein Purpura and its Characteristics in Infected Children During 2009-2016

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ABSTRACT

Introduction: Due to the different complications caused by Henoch-Schonlein purpura and in order to prevent additional treatment costs for patients, we decided to investigate the clinical features of cases of Henoch-Schonlein purpura.

Materials and Methods: In this descriptive cross-sectional study, 52 children in whom the diagnosis of Henoch-Schonlein purpura was confirmed were enrolled in the study from 2009 to 2016. The instruments of this research include a researcher-made questionnaire with two parts. The first part is related to demographic characteristics including age, sex and season of referral and the second part was related to morbidity such as various symptoms. Data were collected from patients' records and interviews with families and were analyzed using SPSS V.22.

Results: Out of 52 patients, 30 (57.7%) were boys and 22 (43.3%) were girls. The mean age of patients was 3.02 to 6.58 years. Autumn and winter had the most clients of Henoch-Schonlein purpura. One of the most common manifestations seen in most patients was cutaneous manifestations. There was no significant relationship between gender and clinical manifestations and seasons of the year with these manifestations ($p > 0.05$). The results showed that age was significantly associated with renal manifestations ($P < 0.05$).

Conclusion: The results of this study showed that the prevalence of Henoch-Schonlein purpura disease is higher in boys. Cutaneous manifestations were seen in most patients and in patients with renal manifestations, the mean age was higher and significant.

Keywords: Henoch-Schonlein purpura, clinical manifestations, complications, gender, age

INTRODUCTION AND STATEMENT OF THE PROBLEM

Henoch-Schonlein purpura (HSP) is an acute, pervasive, non-purulent vascular inflammation (1). In 1800, Henoch described the syndrome as incomplete. In 1830, Schonlein described typical rashes and the articular features of the disease. In 1870, Henoch recognized its renal and gastrointestinal manifestations, and the disease became known as Henoch-Schonlein purpura Syndrome (2).

Henoch-Schonlein purpura is a condition in which small blood vessels (capillaries) become inflamed, called vasculitis, and usually affects the small blood vessels, skin, intestines, and kidneys (3). The cause of this disease is unknown and its characteristic feature is inflammation of small blood vessels. The disease may be caused by allergies, although this has not been proven (4). In more than 75% of cases, there is an upper respiratory infection or (52-35%) gastrointestinal infection before the onset of the disease (5). Occasionally, Henoch-Schonlein purpura has been reported also after insect bites, the use of certain drugs such as chitin, ampicillin, erythromycin, and penicillin, infectious agents such as hemolytic streptococcus B, varicella, measles, rubella, hepatitis B, parvovirus B12, and food allergies (6). Henoch-Schonlein purpura is mainly a childhood disease and its incidence is about 9 per thousand and often occurs between the ages of 3 to 15 years and is seen in boys 1.5 times more than girls (8, 7 and 9). Different epidemiological studies have considered seasonal differences in the incidence of HSP. In most studies, the highest prevalence of the disease has been reported in autumn and winter and the lowest in summer (11,10). The main symptom of HSP is cutaneous manifestations in the form of non-cytopenic purpura (palpable) (12). Sore throat occurs about two weeks before the onset of purpura in 40% of children and mild fever, headache, joint pain and abdominal pain occur. Arthritis (joint pain and swelling) occurs in 80% of patients with HSP and it can occur in any joint, but tends to involve the lower limbs and most of all the ankle and knee joints (13). Gastrointestinal involvement also occurs in about half of children and one-third of children with HSP show renal involvement (14). Diagnosis is mainly based on clinical symptoms and there is no specific laboratory test for it (15). According to the modified criteria of the American College of Rheumatology, Henoch-Schonlein purpura is diagnosed with criteria of tangible purpura, Angina pectoris, gastrointestinal bleeding, hematuria, onset of disease under the age of 17 and rejection of drug reactions as a cause of lesions. Diagnosis of this disease requires the presence of 2 of the 4 cases listed in Table 1, which there is 87.1% sensitivity and 87.7% specificity for this disease (16,17).

The short-term and long-term prognosis of children with this disease is usually excellent (18). The initial onset

of the disease in cases where there is no obvious renal involvement, and in about two-thirds of cases, resolves within 4 weeks. In the remaining one-third (33%), we observe a recurrence of the disease, which is usually resolved after 4-6 months (21, 19, 20). Central nervous system involvement and severe renal involvement are associated with a poor prognosis, and long-term prognosis of this disease is mainly determined by renal involvement (increased urea nitrogen, persistent proteinuria, hypertension and renal insufficiency) (22 and 23) which leads to negative behaviors and attitudes towards oneself, non-productive work and absenteeism from activities, low behavior and dissatisfaction(24).

Table 1.1. Diagnostic criteria of Henoch-Schonlein purpura

Tangible purpura	Prominent, tangible, bleeding skin lesions in the absence of thrombocytopenia
Abdominal angina	Diffuse abdominal pain or diagnosis of abdominal ischemia
Diagnostic biopsy	Histological changes indicating granulocytes in the walls of arterioles or venules
Children's age group	Age less than 17 years at the onset of symptoms

Work and clients, non-productive work and absenteeism from activities, low behavior and dissatisfaction. . Treatment of this disease includes supportive, symptomatic and other treatments to reduce the complications of the disease in each case (25). Symptomatic treatment includes pain control, hydration, soft diet, reduced activity and preventing keeping the lower limbs (legs) stable (26, 27). Due to the different clinical manifestations in patients with Henoch-Schonlein purpura and its unique characteristics, the present study was conducted to investigate the characteristics of this disease in patients between 2009 and 2016.

MATERIALS AND METHODS

This study is a descriptive cross-sectional study that was performed to evaluate the epidemic and clinical characteristics of patients with Henoch-Schonlein purpura between 2009 and 2016. In this study, all patients who were definitively diagnosed with the disease within a specified time interval were studied. The study included 52 people. The research tool included an author-made questionnaire composed of two parts. The first part includes demographic characteristics such as age, sex, season (spring, summer, autumn and winter) and place of residence and the second part is related to clinical findings including cutaneous manifestations (including petechiae, purpura, ecchymosis), gastrointestinal symptoms (abdominal pain, gastrointestinal bleeding), renal symptoms (overt hematuria, microscopic hematuria, edema), joint symptoms (arthritis, arthralgia), laboratory findings (inflammatory markers, urine analysis, kidney biopsy) and treatments for each patient were entered into the checklist by referring to patient records or interviewing the child's parents. Cases whose records were not available in the hospital archives for any reason or those that were incomplete were excluded. First, written consent was obtained from the parents of all patients. Then, the researcher filled the questionnaire using the patients' records or interviewing with the child's parents if needed. Data were analyzed using SPSS V.22 software and P value less than 0.05 was considered significant.

Findings: Out of 52 patients under study, 30 (57.7%) were males and 22 (42.3%) were females. The mean age of patients was 6.58+_3.02 years with a 2-13 years interval. Patients were equally residing in villages and cities, 26 per each group (50%). In the present study, the most prevalent manifestations observed in patients included cutaneous manifestations, available in 50 (98.1%). Table 1 gives types of manifestations observed in subjects.

Table 1. Types of clinical manifestations in children with purpura

Variable	Frequency	Percentage of frequency
Previous history of respiratory infection	30	57/6
History of allergies	6	11/5
Cutaneous manifestations	50	98/1
Gastrointestinal manifestations	34	65/38
Renal manifestations	12	23/07
Articular manifestations	42	80/76

In investigating the seasons of referral of patients using t-test it was found that the highest rate of referral was in the cold seasons of year, i.e., winter and autumn, and November with a frequency of 12 cases (23.07%) had the highest frequency and June with a frequency of 2 cases (3.84%) had the lowest frequency of referrals. Table 2 presents distribution of clinical manifestations in terms of the year seasons. Findings of this research showed that although different clinical manifestations are more observed in the cold seasons, no significant relationship was found between clinical manifestations and different seasons (p>0.05).

Table 2. Distribution of clinical manifestations in terms of seasons

Seasons of the year	Spring N(%)	Summer N(%)	Autumn N(%)	Winter N(%)	P Value
Clinical manifestations	(19/23)10	(6/96)4	(30/76)16	(38/46)20	0/78
cutaneous manifestations	(11/53)6	(3/74)2	(30/76)16	(19/23)10	0/43
Gastrointestinal manifestations	(3/74)2	(3/74)2	(3/74)2	(11/53)6	0/12
Renal manifestations	(15/38)8	(3/74)2	(30/76)16	(30/76)16	0/23

Also, distribution of clinical manifestations in terms of gender was investigated in Table 3 using t-test. Results indicate that there was no significant relationship between none of clinical manifestations and gender (p<0.05).

Table 3. Distribution of types of clinical manifestations in terms of gender

Gender Clinical manifestations	Male N(%)	Female N(%)	P Value
cutaneous manifestations	30(57/6)	20(38/46)	0/87
Gastrointestinal manifestations	20(38/46)	14(26/)92	0/54
Renal manifestations	4(6/96)	8(15/)38	0/22
Articular manifestations	24(46/15)	18(34/61)	0/69

Using t-test, the relationship between patients' age and clinical manifestations was examined. In this study, only a significant correlation was observed between age and renal manifestations, and renal manifestations increase with increasing the age (p=0.03), but no significant relationship was observed between clinical manifestations and age (table 4).

Table 4: The relationship between the mean age and the incidence and absence of clinical manifestations

Age Clinical manifestations		Mean age	SD	P value
Skin	Presence Absence	6/55 8/00	3/04 00/00	0/63
Gastrointestinal	Presence Absence	6/99 5/74	3/11 2/71	0/16
Renal	Presence Absence	8/59 4/06	3/30 2/74	0/01
Articular	Presence Absence	6/58 6/55	2/58 3/83	0/97

DISCUSSION

Henoch-Schonlein purpura is usually a self-limiting disease. However, in some cases it can be associated with long-term complications such as intussusception, intestinal infarction and intestinal perforation and even death (28). The disease may affect several organs, and the main organs vulnerable to this disease include the skin, kidneys, joints, and digestive system. Kidney damage can have serious consequences for such patients (29).

In this study, the number of male patients was more than female patients. However, statistically, there was no difference between the sexes in terms of clinical manifestations. Also, the mean age of patients was 6.58 years. Cutaneous manifestations included the most and renal manifestations the least. However, patients with renal manifestations were older than other manifestations and this relationship was statistically significant. In cold seasons such as autumn and winter, the disease was more common.

According to the results of the present study, the ratio of males to females is 1.4 to 1. In the Matini's study, this ratio was 1.6 to 1, in the study of Assar et al. it was 1.9 to 1, in the Mortazavi's study, this ratio was 2.6 to 1, and in the study of Chen et al. (2013), it was 1.9 to 1 (10,5,30,31). In the study of Fertzaias et al., the incidence ratio was 1.3 to 1 (31). In a study by Panjips et al. (2011), this ratio was reported to be 1.4 to 1 (33). It seems that the research population in the above studies has a similar distribution to our study. The reason for this similarity is due to the type of statistical population and climatic conditions as well as other variables discussed regarding the higher prevalence of this disorder among boys than girls. Therefore, in several studies, the priority of prevalence of Henoch-Schonlein purpura has been reported in boys (34,35).

Based on the results, the age range of the participants in the present study is 2-13 years with an average age of 6.58 years, which corresponds to the average age of patients in the study by Assar, which was 6.4 years (10). Also, in the study of Mortazavi and Chen, the average age was 6.8 and 6.6 years (31,30). In the study by Fertzaias, the mean age of patients was 5.2 years (32). The average age of children in the study of Panjips was 7.2 years (33). In various studies, there is no definite age for infection, and it has been more or less than average age in our work compared to the above studies due to different environments and age-related factors. Various studies have shown that children aged 5-15 years and the average age of 5 to 6 years are at risk for Henoch-Schonlein purpura (36, 37). In this regard, the results of our research are supported.

The most clinical manifestations of the studied patients are cutaneous, articular, gastrointestinal and renal symptoms, respectively. In the study of Heydarian et al. after cutaneous manifestations, articular manifestations appeared in the next rank (1 and 38), which are consistent with our results. However, studies conducted by Matini, Chen, Mortazavi and Assar (10,5,20,31) found a different order of manifestations from the present study, so that following the cutaneous manifestations, gastrointestinal manifestations have been proposed that is not consistent with our results. Differences in results can be attributed to the age of infection, duration and severity of infection, demographic or geographical characteristics of the region. Several studies have been conducted on stress and adaption, only recently researchers have developed an interest in the realm of religion and spirituality as a possible source for individuals to apply when dealing with stress-inducing events in their lives(39).

According to the findings of our study, it was observed that the highest incidence of Henoch-Schonlein purpura is in cold seasons (autumn and winter). Various sources have also stated that the incidence of the disease decreases significantly in the warm season of the year, i.e., summer, for unknown reasons (40,41), which are in line with the results of our research. The family-centered empowerment

model (FCEM) is one of the effective ways to empower the chronic patients and their families(42).

CONCLUSION

According to the results of the study, the prevalence of the disease is higher in boys with an average of 6.58 years. Cutaneous manifestations are seen in most patients. In patients with renal manifestations, the mean age is significantly higher. Therefore, in patients with Henoch-Schonlein purpura, more attention can be paid to skin complaints and renal complications so that early diagnosis and treatment can be achieved.

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