Henoch-Schönlein purpura in children: A cross sectional study

Fatih Karaaslan¹, Betul Gemici Karaaslan¹, Huseyin Dag¹, Soner Sazak¹, Emine Turkkan¹, Bilal Yılmaz¹, Gulsen Kose Midillioglu²

From Department of Pediatrics, ¹University of Health Sciences, Istanbul Okmeydani Training and Research Hospital, ²Şişli Hamidiye Etfal Training and Research Hospital, İstanbul, Turkey

Correspondence to: Dr. Huseyin Dag, Okmeydanı Egitim Ve Araştırma Hastanesi, Pediatri Kliniği Darülaceze cad. No: 27 Şişli/Istanbul, İstanbul, Turkey. E-mail: huseyindag2003@gmail.com

Received - 11 February 2019 Initial Review - 02 March 2019 Accepted - 14 March 2019

ABSTRACT

Background: Henoch-Schönlein purpura (HSP) is a leukocytoclastic vasculitis with small vessel involvement and mainly affects the skin as well as joints, the gastrointestinal system (GIS), kidneys, and, more rarely, other organs. Objective: The objective of this study was to evaluate the sociodemographic characteristics, and clinical and laboratory findings of patients diagnosed with HSP.

Materials and Methods: This was a retrospective study done to find out the sociodemographic data, clinical, laboratory findings, and treatment information of patients diagnosed with HSP and was admitted to the Pediatric Clinic of a tertiary care hospital between January 1, 2008, and August 31, 2013. The data were obtained from the hospital’s data processing system. HSP cases were validated according to EULAR/PRINTO/PRES criteria. Mean standard deviation, median, lowest and highest, frequency, and ratio values were used in the descriptive statistics of the data. Results: The study included 85 patients between the ages of 2 and 16 years, wherein 49 patients (57.6%) were male and 36 (42.4%) were female. The mean age was 9.9±3.3 years and 53 patients (62.4%) were under 10 years of age. The most common precipitating factor was upper respiratory tract infections. Purpura was the only symptom observed in all the patients and joint involvement was the second most common symptom (60%). GIS involvement was observed in 46 patients (58.8%) and intussusception was observed in one patient. Nine patients (10.6%) had renal involvement with mild nephropathy. The most frequently observed laboratory findings were increased C-reactive protein (47%) and leukocytosis (31%). Conclusion: HSP is commonly seen in children and leads to life-threatening complications in a minority of patients. Whole patients with GIS and renal involvement should be examined and monitored to assess the severity of the disease and any complications.

Key words: Children, Complications, Henoch-Schönlein, Purpura

Henoch-Schönlein purpura (HSP) was first described by Heberden, in 1801, and its relationship with arthritis was highlighted by Schönlein, in 1837 [1]. HSP is a leukocytoclastic vasculitis with small vessel involvement; it mainly affects the skin as well as joints, the gastrointestinal system (GIS), kidneys, and, more rarely, other organs. Its etiology is not fully known [2] and it is the most widespread reason of non-thrombocytopenic purpura in children. It is a disease that is generally seen in children between the ages of 5 and 15 years. The estimated incidence of the disease in children <17 is 13–20/100,000 [3].

Although the etiology of HSP is unknown, it has been reported that various factors such as infections, vaccines, drugs, foods, and insect bites trigger the disease [4]. The fact that two-thirds of the patients had a history of upper respiratory tract infection (URTI) and the disease was more commonly seen in winter and spring, when URTIs are very frequent, could be incidental, but it is more likely that HSP is linked to infectious diseases. The most important microorganisms are streptococci; in addition, many viral and other bacterial agents have also been reported to precipitating the disease [3]. IgA plays an important role in the pathogenesis of HSP. Studies have shown renal mesenchymal IgA accumulation, increased serum IgA, and the presence of IgA-containing immune complexes in patients with HSP [5].

The most common and distinct sign of the disease is a skin rash in the form of non-thrombocytopenic palpable purpura that appears, especially on lower extremities. Non-thrombocytopenic purpura, arthritis, and abdominal pain in the early phase of the disease constitute the classic triad of symptoms of HSP and facilitate a clinical diagnosis [5]. The skin rash can be accompanied by soft tissue swelling, especially on the scalp, on the tibial aspect of the lower extremities, and on the back of the hands. Less frequently, localized edema on the eyelids, lips, ears, and perineum can be seen especially in younger children [6]. This edema is not associated with proteinuria, but it generally stems from angioedema and does not leave an indented mark when pressed [7]. Joint involvement in HSP manifests as arthralgia or arthritis. Knee, elbow, wrist, and ankle involvement are frequently seen. There is no intra-articular hemorrhage or effusion, nor permanent deformity [8].
GIS involvement generally manifests as abdominal pain. If abdominal pain occurs before the rash, it can be confused with acute abdomen. Acute abdomen can be accompanied by vomiting and blood in stools [8,9]. In GIS involvement, serious life-threatening complications other than bleeding include intussusception and perforation. The diagnosis should always be considered in the case of very severe and long-term pain [8].

Long-term prognosis of HSP is determined by the severity of kidney involvement. The most common sign of kidney involvement is hematuria, which generally develops within the first 4 weeks. Proteinuria can be seen in various degrees of severity, ranging from mild proteinuria to severe proteinuria, as in nephrotic syndrome. Hypertension can generally be seen in the early stages. Kidney function tests are usually normal; however, some patients may develop severe kidney involvement and progressive glomerulonephritis [10].

Neurologic involvement is rarely seen in HSP. Central nervous system damage occurs either due to the direct effect of vasculitis or the indirect effect of systemic inflammation. Severe neurologic involvement is rare in HSP, whereas mild effects have been reported more frequently. In one study, 244 cases were retrospectively screened and it was found that 17 (6.9%) patients experienced convulsions and confusion. Recent studies have reported a much lower frequency of such symptoms [11].

HSP is the most common vasculitis of childhood and can cause serious complications. The aim of this study was to investigate the epidemiologic, clinical, and laboratory characteristics of patients with HSP who were admitted to our clinic and to review the treatment approaches for these patients.

RESULTS

A total of 85 patients with HSP were included in the study, of which, 49 patients (57.6%) were male and 36 (42.4%) were female, with male:female ratio of 1:36. The ages of the patients were between 2 and 16 years, and the mean age was 9.9±3.3 years. 53 patients (62.4%) were under 10 and 32 patients (37.6%) were over 10. Considering the distribution of patients by the season of admission, 32 patients (37.6%) were presented in winter, 22 (25.9%) in autumn, 19 (22.4%) in spring, and 12 (14.1%) in summer. Of the total study patients, 23 (27%) had a history of a potentially triggering factor before the onset of HSP. As a triggering factor, 20 patients (23.5%) had a history of URTI (rhinitis, otitis, tonsillitis, and pharyngitis) and three patients had a history of insect bites.

It was found that all patients had non-thrombocytopenic purpura ranging from petechiae to ecchymosis, and these lesions were localized most frequently on the lower extremities and hips, and less frequently on the upper extremities. Joint involvement was observed in 51 patients (60%). Joint involvement manifested as arthritis in 33 (38.8%) patients and arthralgia in 18 (21.2%) patients.

Gastrointestinal involvement was found in 50 patients (58.8%) and all these 50 patients had abdominal pain and 36 (42.3%) had gastrointestinal bleeding. The bleeding manifested as fecal occult blood in 33 patients (38.8%) and as gross bleeding in three patients. Abdominal ultrasonography (USG) revealed intussusceptions in one patient with severe abdominal pain. It was found that abdominal pain was an early symptom in 15 patients (17.6%) followed by purpuric rash. Renal involvement was detected in 9 patients (10.6%). These patients had mild nephropathy along with microscopic hematuria. None of the patients had proteinuria or severe nephropathy.

The main laboratory findings in the acute phase of the disease were as follows: Mean leukocyte count was 11.5±3.4 (1000×/μl) and 26 patients (30.6%) had leukocytosis. The mean hemoglobin count was 13.3±1.1 g/dl and 10 patients (11.7%) were anemic. The mean platelet count was 364.2±117.9 (1000×/μl) and 19 patients (22.4%) had thrombocytosis. CRP was positive (>8 mg/dl) in 40 patients (47%). The mean urea value was 24.3±8.5 mg/dL, and the mean creatinine value was 0.5±0.2 mg/dL. Creatinine values were within normal limits in all patients, whereas urea was higher than normal in four patients. The CRP-positive group had a significantly higher rate of arthritis as compared to the CRP-negative group (p<0.05). There was no significant difference between CRP positivity and other clinical findings.

The rate of renal involvement with hematuria was significantly higher in the group with leukocytosis in comparison to the group without leukocytosis [Table 1].

It was observed that nonsteroidal anti-inflammatory drugs (ibuprofen, 30 mg/kg/day) were administered to 29 patients (34.1%), corticosteroids were administered to 38 patients (44.7%), and 18 patients (21.2%) were followed up without any treatment. Among the patients that received treatment, 44 (65.6%) had only GIS involvement, 2 (2.9%) had only renal involvement, and 7 (10.4%)...
had both GIS and renal involvement. It was found that one patient developed intussusception as a complication and the other patients did not have any complications during the hospitalization period.

**DISCUSSION**

The findings of our study, i.e. age of the patients and the male-to-female ratio (1.36:1) were similar to the data reported in other studies [13,14]. Although the disease is seen throughout the year, it is known that the disease is more frequent in winter as seen in our study; furthermore, it is followed by autumn. There are studies reporting that HSP is associated with respiratory tract infections, and the fact that the disease is more frequently seen in cold weather, especially in autumn and winter, implies that infections could be the triggering factor in the pathogenesis of this disease. However, this may be a coincidence on account of the increased incidence of URTI in children during winter [15].

Clinical findings from the patients revealed that all patients had a rash in the form of purpura. Lesions were localized generally on the lower extremities and rarely on the upper extremities and torso. Sometimes, skin findings are not present as the first symptom and can appear after arthritis or GIS symptoms. The occurrence of skin findings following joint and GIS involvement ranges between 25 and 50% [15,16]. Two different studies reported that joint involvement was seen in 15% and 25% of patients, respectively, and abdominal pain in 12% and 20% of patients, respectively, as initial symptoms [17,18]. Among our patients, 17.6% had abdominal pain and 18.8% had joint pain as initial symptoms [15,16,19]. In our study, 60% of the patients had joint involvement. The most common joint involvement was seen in the ankles and knees, which is consistent with literature.

Studies reported a GIS participation rate of 38–75% in HSP. In one study, it was obtained that the rate of GIS involvement was 62.3% and incident of fecal occult blood was 37.7% [3]. In our study, 58.8% of the patients had GIS involvement. All patients had abdominal pain, 38.8% of the patients had fecal occult blood and 3.5% had fecal fresh red blood, all consistent with the data reported in literature. The most common surgical complication of HSP is intussusception. Radiologic, USG, and endoscopic assessment of patients presenting to the clinic with GI symptoms significantly reduced the number of unnecessary laparotomies. Studies that carried out gastrointestinal USG examinations in children with HSP, who also had abdominal pain, revealed increased duodenum wall thickness and echogenicity in 82%, increased duodenum diameter in 64%, and presence of gallbladder hydrops in 36% of these children. Some studies showed intestinal wall thickening, segmental dilatation of the intestine, and intestinal hypomotility in patients with HSP who were evaluated using abdominal USG [20,21].

In another study conducted by Connolly et al. [22], repeated USG of 12 patients with HSP showed decreased bowel wall thickening, a reduced amount of peritoneal fluid, return of peristalsis, and improved loose invaginations, and the improvement in sonographic findings were consistent with the clinical improvement. Therefore, it was reported that the severity and extent of gastrointestinal involvement could be monitored in HSP with repeat USG. In our study, an abdominal USG examination of a patient with abdominal pain showed requiring surgery.

The rate of renal involvement in HSP was reported to have a range between 10 and 60%, and the first signs of renal pathology are generally seen within 2 weeks after purpuric lesions are observed [23]. HSP nephritis generally manifests as mild proteinuria and/or microscopic hematuria that resolves spontaneously. However, renal involvement can be the primary cause of chronic kidney disease (CKD) in HSP. Therefore, kidney functions in HSP patients should be closely monitored, and treatment provided in the case of severe involvement [24]. A study by Sano et al. found that at the age of >4 years, presence of severe gastrointestinal involvement, and purpura tending to last more than 1 month, was reported as risk factors for renal disease, and the most significant factor determining prognosis was reported the severity of renal symptoms at disease onset [25]. In our study, it was found that 10.6% of patients had renal involvement with microscopic hematuria, and renal symptoms appeared along with the rash or within the first few days. None of the patients had proteinuria and their blood pressure was within normal limits throughout hospitalization. In a study by Hamdan and Barquari, the effect of age distribution on systemic involvement was investigated in 68 patients with HSP, and it was found that 19%
of the patients younger than 5 years and 67% of the patients older than 10 years had developed nephritis. Hence, it was highlighted that the development of nephritis was age dependent and the incidence of nephritis was higher in older children [26]. Similarly, our study has also demonstrated that 89% of the patients with renal involvement were older than 7 years. Consequently, urinalysis results and blood pressure should be monitored more carefully in the presence of renal involvement, especially in older children. Laboratory tests are generally supplementary rather than diagnostic in HSP. In our study, the mean values of hemoglobin, leukocyte, platelet, urea, and creatinine parameters were within normal limits. Urea levels were higher than normal in four patients, but a relationship between elevated urea level and HSP could not be established in these patients as they did not have renal involvement. In all, 30.6% of the patients had leukocytosis, 22.4% had thrombocytosis, and 47% had CRP positivity. The detected high levels of acute phase reactants such as CRP, platelets, and leukocytes were consistent with other studies [19,27] and show that inflammation plays a role in the pathogenesis of HSP. The absence of other diseases and symptoms that may cause arthritis, URTI, or leukocytosis in patients with hematuria means that the risk of kidney involvement may increase in patients with leukocytosis. Even so, this field requires further prospective studies.

HSP is generally a self-limiting disease that does not require any special treatment other than support. Joint pain can usually be managed using analgesics such as ibuprofen. In patients with active GIS bleeding, nonsteroidal anti-inflammatory treatment for joint symptoms is undesirable. Hypertension is managed with suitable antihypertensive drugs. If there is an infection, it is treated with suitable antibiotics depending on the infectious agent and severity of infection. Surgery may be required for serious GIS complications. Severe skin symptoms, abdominal pain, and protein loss respond dramatically to oral corticosteroid treatment within a few days [16,27]. Corticosteroid treatment can lead to remission of arthritis and abdominal pain; however, it does not shorten the duration of the disease or prevent relapse. Starting steroid treatment in a patient with abdominal pain but no bleeding is considered undesirable as it may mask more severe complications. GIS symptoms exhibit remission without treatment in cases other than those requiring surgery. Starting treatment in the initial phase does not prevent the symptoms from developing. A few studies asserted that starting steroid treatment in the initial phase in patients (older than 7, with severe GIS involvement, and long-term purpura) who are suspected to have GIS symptoms exhibit remission without secretion. Starting steroid treatment in patients with severe GIS complications is higher in older children (especially older than 7 years of age), patients who have more severe and long-term skin symptoms, leukocytosis, early renal findings, and serious abdominal symptoms. Therefore, these patients should be closely monitored.

**REFERENCES**

Karaaslan et al. Purpura, children


Funding: None; Conflict of Interest: None Stated.


Doi: 10.32677/IJCH.2019.v06.i03.001