ABSTRACT

Objective: Henoch Schönlein Purpura (HSP) is the most common benign vasculitis occurring during childhood. Morbidity and mortality rates rise when there is organ system involvement. We aimed to retrospectively evaluate HSP cases in our clinic according to organ involvement, clinical and laboratory findings.

Material and Methods: Twenty-eight HSP cases were followed in our clinic from January 1st 1997 to June 1st 2000. The cases were retrospectively evaluated.

Results: The mean age of the patients was 7.96 years (interval: 3-14 yrs.). 17 of them were males, 11 were females. The mean hospitalization period was 6.36 days (interval: 1-16 days). Two patients had been hospitalized twice due to recurrence and two others had each been hospitalized once before in another clinic due to the same disease. 9 patients (32.1 %) had a history of an upper respiratory tract infection (URTI) of whom 3 (10.7 %) had positive throat cultures for beta hemolytic streptococcus. Skin lesions were seen in all patients. Arthritis/arthralgia developed in 12 (42.9 %) patients. Gastrointestinal system tract (GIT) involvement developed in 19 patients (67.9 %), 7 (25 %) of them presented with abdominal pain and vomiting, 12 (42.9 %) with occult fetal blood. In 4 patients (14.3 %) with GIT involvement, hematochezia and melena developed later. Abdominal ultrasonography was performed on 17 cases (60.7 %). No surgical intervention was required for any of the patients. 11 patients (39.3 %) had renal involvement. In 8 of the cases (28.6 %) renal disease presented with microscopic hematuria with or without proteinuria and in the other 3 (10.7 %) with macroscopic hematuria. In 1 (3.6 %) case acute renal failure developed and hemodialysis was performed. Central nervous system (CNS) involvement occurred in one patient (3.6 %) who had convulsions. In 2 cases (7.1%) scrotal involvement was observed. 13 patients were given steroids.

Conclusions: In HSP cases, renal and GIT involvement should be searched for meticulously and patients with renal involvement should be followed up regularly.

Key Words: Henoch Schönlein Purpura, HSP nephritis, Vasculitis, Gastrointestinal bleeding.
INTRODUCTION

Henoch Schönlein Purpura (HSP) is the most common vasculitis in childhood. It is characterized by non-thrombocytopenic purpura, arthritis/arthralgia, abdominal pain, gastrointestinal bleeding and glomerulonephritis (1,2).

Although it is known as a benign vasculitis, morbidity and mortality increase when there is organ system involvement. It is important to follow a patient with renal, GIT and CNS involvement regularly. This study was performed to evaluate retrospectively the HSP patients hospitalized in our clinic within a certain period.

MATERIALS AND METHODS

Patients diagnosed as and treated for HSP between January 1st 1997 and June 1st 2000 and later followed up in the outpatient clinic were chosen for the study. The number of patients was 28 (17 males, 11 females). Records of their age, sex, medical history, hospitalization period, number of recurrence episodes, clinical findings, laboratory tests, radiological and histopathological findings were taken and analyzed in the light of literature.

RESULTS

The age and sex distribution of the cases are presented in Table-I.

Table I.: The age and sex distribution of the cases

<table>
<thead>
<tr>
<th>Number</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age Range (Yr)</td>
<td>17</td>
<td>11</td>
</tr>
<tr>
<td>Mean Age (Yr)</td>
<td>3-13</td>
<td>3-14</td>
</tr>
<tr>
<td>Male / female ratio</td>
<td>1.5 / 1</td>
<td></td>
</tr>
</tbody>
</table>

Male / female ratio was 1.5 / 1. The youngest patient was 3, the eldest was 14 years of age. The overall mean age was 7.96.

Distribution of the cases according to months and years is given in Table-II.

Table II.: Distribution of cases according to months and years.

<table>
<thead>
<tr>
<th>Months/Years</th>
<th>1997</th>
<th>1998</th>
<th>1999</th>
<th>2000</th>
</tr>
</thead>
<tbody>
<tr>
<td>January</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>-</td>
</tr>
<tr>
<td>February</td>
<td>-</td>
<td>1</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>March</td>
<td>2</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>April</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>May</td>
<td>-</td>
<td>-</td>
<td>4</td>
<td>-</td>
</tr>
<tr>
<td>June</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>July</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>1</td>
</tr>
<tr>
<td>August</td>
<td>1</td>
<td>-</td>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>September</td>
<td>1</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>October</td>
<td>4</td>
<td>1</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>November</td>
<td>2</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>December</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
<td>4</td>
<td>9</td>
<td>3</td>
</tr>
</tbody>
</table>

The hospitalization period varied between 1 to 16 days (mean: 6.36 days). Two patients were hospitalized twice in our clinic due to recurrence and 2 others were each hospitalized once before in different clinics.

The medical histories revealed upper respiratory infections in 9 patients (32.1 %), of whom 3 (10.7 %) had positive throat cultures for beta hemolytic streptococcus; one had otitis media (3.6 %), one had urinary tract infection (3.6 %), and one had bronchopneumonia (3.6 %). 2 patients had a history of acetyl salicylic acid (80 mg) use, 1 had furosemide intoxication, and 1 patient a recent trauma history.

Non-specific findings were as follows: fever between 38.0-38.5°C in 5 cases (17.9 %). Between 37.5-37.9°C in 7 patients (25.0 %), less than 37.0°C in 16 (57.1 %); hypertension in 4 patients (14.3 %). The findings are given in Table-III.

Table III.: The clinical findings and organ involvements of the cases.

<table>
<thead>
<tr>
<th>Clinical Findings</th>
<th>Number of Cases</th>
<th>Percentage (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Purpura</td>
<td>28</td>
<td>100.0</td>
</tr>
<tr>
<td>Arthralgia / Arthritis</td>
<td>12</td>
<td>42.9</td>
</tr>
<tr>
<td>Gastrointestinal System Involvement</td>
<td>19</td>
<td>67.9</td>
</tr>
<tr>
<td>Renal Involvement</td>
<td>11</td>
<td>39.3</td>
</tr>
<tr>
<td>Scrotal Involvement</td>
<td>2</td>
<td>7.1</td>
</tr>
<tr>
<td>CNS Involvement</td>
<td>1</td>
<td>3.6</td>
</tr>
</tbody>
</table>

Distribution of the cases according to months and years is given in Table-II.
Purpuric skin lesions were seen in all patients (100.0%). In some patients they occurred after arthralgia/arthritis and in some following abdominal pain and/or vomiting.

Arthralgia/arthritis was found in 12 cases (42.9%). The most involved joints were ankles and knees. In 1 patient (3.6%) elbow pain was recorded.

Data relating to GIT and renal involvement is given in Table-IV:

In 19 cases (67.9%) there was GIT involvement. In 7 of them (25.0%) presentation was with abdominal pain and vomiting. 12 patients (42.9%) had occult fecal blood in addition to abdominal pain. In 4 of them (14.3%) hematochezia and melena developed.

Renal involvement was evident in 11 cases (39.3%). 8 of them (28.6%) also had GIT involvement. 8 of the patients with renal disease (28.6%) presented with microscopic hematuria with or without proteinuria and the other 3 (10.7%) with macroscopic hematuria. In 1 (3.6%) case acute renal failure developed and hemodialysis was performed. CNS involvement occurred in 1 patient (3.6%) presenting with convulsions. The only finding in cerebro-spinal fluid was xanthochromia. EEG did not reveal any specific findings for epilepsy.

Two patients (7.1%) had scrotal involvement. One of them was diagnosed as having orchio-epididymitis by ultrasonography. The other, who had an orchiectomy before due to abdominal orchiopexy, was found to suffer only from testicular pain.

Complete blood count results are given in Table-V:

C3/C4 were measured in 9 cases. C3 was found to be lowered in 1 patient and increased in 2 patients. C4 was normal in all cases.

Bronchopulmonary infiltration was observed on the chest X-ray of 1 patient.

On 17 patients ultrasonography (USG) was taken. In 11 patients no pathological findings were found. In 1 patient (3.6%) bowel dilatation and in 2 patients (7.1%) suspected invagination with fluid collection was observed. In the control USG analyses the findings returned to normal. No patient required surgical intervention.

Two patients (7.1%) were found to have grade 1-2 increases in renal echogenicity. In 1 patient (3.6%) minimal fluid collection in the right pleural space was observed.

Seven patients required skin biopsies to ascertain the diagnosis after the consultation with the Dermatology Clinic. All the biopsy results were congenial with leukocytoclastic vasculitis.

Renal biopsies were taken in 2 patients with acute renal failure and massive proteinuria. The findings were typical of HSP nephritis with crescent formation.

<table>
<thead>
<tr>
<th>Table IV.: Gastrointestinal and renal involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gastrointestinal Involvement</td>
</tr>
<tr>
<td>-------------------------------</td>
</tr>
<tr>
<td>Number</td>
</tr>
<tr>
<td>Abdominal Pain and Vomiting</td>
</tr>
<tr>
<td>Occult Fecal Blood</td>
</tr>
<tr>
<td>Melena+Hematochezia</td>
</tr>
</tbody>
</table>

Table V.: Complete blood count results.

<table>
<thead>
<tr>
<th></th>
<th>Ranges</th>
<th>Mean and SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>5.6-14.5g/dl</td>
<td>11.98g/dl±1.91</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>16-43 %</td>
<td>35.94 %±5.93</td>
</tr>
<tr>
<td>Erythrocyte</td>
<td>3.7-5.3 million/mm³</td>
<td>4.62 million/mm³±0.45</td>
</tr>
<tr>
<td>Leukocyte</td>
<td>5600-21000/mm³</td>
<td>12600/mm³±3563</td>
</tr>
<tr>
<td>Thrombocyte</td>
<td>219000-572000/mm</td>
<td>389153/mm³±123754</td>
</tr>
<tr>
<td>Sedimentation Rate</td>
<td>12-120 mm/h</td>
<td>44.25 mm/h±29.40</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Gastrointestinal Involvement</th>
<th>Renal Involvement</th>
<th>Gastrointestinal + Renal Involvement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>%</td>
<td>Number</td>
</tr>
<tr>
<td>Abdominal Pain and Vomiting</td>
<td>7</td>
<td>25.0</td>
</tr>
<tr>
<td>Occult Fecal Blood</td>
<td>12</td>
<td>42.9</td>
</tr>
<tr>
<td>Melena+Hematochezia</td>
<td>4</td>
<td>14.3</td>
</tr>
</tbody>
</table>
Nurdan Erol, et al

Thirteen cases (46.4%) were treated with steroids. Steroids were given in a dose of 1-2 mg/kg/day for 3-14 days to patients with G1T involvement. Patients having undergone renal biopsy were treated with a dose of 30 mg/kg/day for the first 5 days and then with 1 mg/kg/day for the next month after consultation with the Pediatric Nephrology Department of the Istanbul School of Medicine.

Of the 28 patients only 6 regularly attended follow-up visits in the outpatient clinic. Therefore no reliable data could be obtained for the long term period.

**DISCUSSION**

HSP is a leukoclastic vasculitis according to the Chapel Hill classification (1993) and is the most common vasculitis of the childhood period (3). Its incidence is about 13.5-18.0 / 100,000 (1-3). Although it can occur at any age after the first 4-6 months of life, 50% are seen at under 5, and 75% are seen at under 10 years of age. The youngest of our cases was 3, and the oldest was 14 years of age. The disease is more common in males where the male/female ratio in the literature is reported to be around 1.2/1 - 2.6/1 (1,2,4). The ratio in our series was 1.5 / 1 and was similar to the study of Dicle University Medical School (4).

HSP is a disease which recurs. It recurs mostly within the first 3-4 months of the first attack, or sometimes in a year or rarely in a few years (1,2). Four patients in our study had to be hospitalized twice because of recurrence and complications of HSP.

Although the etiology is unknown, an upper respiratory infection due to streptococci is thought to be the cause. The disease is mostly seen in winter and autumn (2). But in our cases there was no significant tendency of distribution in a specific season. Also adenovirus, mycoplasma and parvovirus infections are seen among the causes of the disease, and the pseudomonas which are accepted as another triggering factor (5). In one of our recurrent cases with G1T and renal involvement the urine culture was positive for pseudomonas aeruginosa. Trauma, some drugs and vaccines are also seen among the causative factors (2,6). Nine of our patients had URTI, where 3 had positive throat cultures, and three had other infections. One patient had been hospitalized due to furosemide intoxication and two had a history of being treated with 80 milligrams salicylic acid the week before the HSP symptoms occurred. Another patient had a trauma history of an arm fracture.

Among the clinical findings of HSP the purpuric and/or petechial skin lesions involving mostly the buttocks and legs and sometimes the arms and face are almost always seen (7-10). One of the diagnostic criteria by the American College of Rheumatology (1990) is leukoclastic vasculitis with polymorphonuclear leukocytes at small vessel walls (2). All of our patients had these lesions. However, when there is organ system involvement other than skin beforehand, it can be difficult to make a diagnosis. Histologically, changes occur in the arterioles, capillaries and venules due to IgA, C3 and fibrin deposition. Complement abnormalities (like C2 deficiency, homozygote null C4 phenotype, C4B deficiency) are held to be responsible for IgA deposition (2).

In recent studies the abnormal glycosylation of IgA subclass A1 has been given a main role in pathogenesis (8,11). Skin biopsies were performed on 7 of our cases. In one of them the skin lesions were pruritic. In another patient the lesions started as purpura and then turned to ecchymoses. All of the results were congenial with vasculitis. To differentiate HSP from other vasculitic diseases IgA ANCA can be measured (1,12).

Arthralgia or arthritis is more frequently seen at the knees, ankles and elbows. The rate of occurrence is between 60-84% (6-8). Arthritis or arthralgia was positive in 12 (42.9%) of our patients. In some cases arthritis preceded skin lesions.

Gastrointestinal system involvement is about 50-80% (1-4, 6-9). In half of the cases occult fecal blood is present (1). The clinical findings include abdominal pain, vomiting, hematemesis, melena (7, 13). Rarely intussusception, bowel perforation, pancreatitis, appendicitis or hydropic gall bladder can occur. The rate of such a major complication is 4.6% (1.3 - 13.6) (13). G1T involvement occurred in 19 (67.9%) of our patients. 12 (42.9%) patients had occult fecal
Henoch Schönlein purpura

blood. In 4 patients (14.3%) rectal bleeding was apparent. None of the cases required surgical intervention. During the control ultrasonographic analysis, cases with suspected invagination were found to have recovered. Ultrasonography is not always sufficient to show GIT involvement but it is useful in following cases with suspected invagination and bowel dilatation (13).

Renal involvement is reported to occur in between 20 and 100% of the cases (2). In 89% of the cases it becomes evident in the first 4 weeks, although in the rest it can take more than 2 months. Renal involvement is more common in patients with GIT involvement (4). 11 of our cases (39.3%) had renal disease where 8 of them (28.6%) had it together with GIT disease. The clinical presentation is mostly as microscopic hematuria +/- with or without proteinuria. In 3 patients we observed macroscopic hematuria, with a decrease in glomerular filtration rate. One patient needed to undergo hemodialysis because of acute renal failure.

Since renal involvement can be a serious challenge during hospitalization or in the follow-up period some methods of monitoring have been suggested like measuring the tubular proteins N-acetyl beta-D-glucosaminidase (NAG) and alpha-1-microglobulin (a 1 MG) (14).

Renal findings include primary lesion, and endocapillary proliferative glomerulonephritis involving the endothelial and mesanchymal cells. Extracapillary cells can cause crescent formation. With immunofluorescence deposits of IgA, IgG, C3 and fibrin can be noticed (2). Renal biopsies were performed in 2 of our patients revealing findings of HSP nephritis including crescent formation.

Central nervous system involvement presents with convulsions, paresis and coma. It is not common (1,2). One patient of our series had convulsions during the hospitalization period.

Scrotal involvement is reported not to be uncommon, and it sometimes presents as testicular torsion (1,2). A study reported that 15% of affected boys complained of significant pain. Ultrasonographic findings include dilated and torsioned epididym, scrotal thickening, hydrocele without involvement of testicles (15). We had scrotal involvement in 2 cases.

The other findings of HSP are due to very rarely seen cholecystitis, myocardial infarction and interstitial lung disease (2, 16). A patient in our clinic had bronchopulmonary infiltration but it has not been possible to ascertain whether it was due to HSP or not.

There are no specific laboratory findings for HSP. Thrombocytosis may be related with the severity of disease increased Von Willebrand factor and serum thrombomodulin levels which show endothelial damage. In severe GIT involvement factor XIII may increase. In 50% of cases serum IgA levels have been reported to be higher than normal. IgA nephropathy and HSP are diseases with similar renal findings. They can be found within the same family. Therefore the ACE gene deletion found in IgA nephropathy patients was searched for also in HSP cases but no significant relation was found (17). Complement levels may be normal or increased. The mechanism for increased complement in renal disease was thought to be associated with lectin levels (18). No specific laboratory findings were present in our cases.

In treatment, steroid use relieves the complaints of GIT involvement rapidly but is not effective in preventing renal disease (19,20). In serious renal involvement pulse steroid treatment alone or with immunosuppressants is suggested (1,21). In our clinic, cases with GIT disease were treated with 1-2 mg/kg/day steroids whereas 2 patients who had renal biopsy were given pulse treatment.

Only a small part of our patients came to regular follow-up visits due to economic and educational problems and because some of them were living in other towns. In the dialysis patient the low GFR rate continued for a while. In the other patient with macroscopic hematuria the purpura recurred once while the GFR returned to normal. The rest had no further problems.

Although it is a benign disease, the morbidity and mortality of HSP increases with multiorgan involvement. Special attention must be paid to complications in cases of gastrointestinal, renal and central nervous system involvement. End stage renal failure has been reported to develop...
in 5-15% of cases with renal involvement. It has been reported to occur more frequently especially in cases with glomerular filtration rate below 70 ml/min., 1 gr. of protein excretion in urine and sclerotic crescent formation. In renal transplant patients, HSP nephritis can recur (19). Therefore long term follow up is recommended for all who develop nephritis.

REFERENCES


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- Duyarlı patojenlerin neden olduğu abdomen, kemik-eklem-cilt ve yumuşak doku, ürogenital sistem, solunum sisteminde gelişen infeksyonlar, sepsis, menenjit, cerrahi profilaksi
- Doz ve Uygulama:
  - Genel olarak 24 saatte 1-2 g (yeni doğanlar: 20-50 mg/kg/gün) maksimum 4 g/gün, tek dozda parenteral olarak uygulanır.
- Kontrendikasyonlar:
  - Sefalosporin duyarlılığı.
- Uyarılar:
  - Penisilinler ile çapraz alerjik reaksiyon görülebilir.
  - Kesin endikasyon olmadıkça gebelikte kullanılmamalıdır.
- Yan Etkiler:
- Ticari Şekiller:
  - Parenteral uygulama için 0.5 g IM, 0.5 g IV, 1 g IM, 1 g IV flakon.

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- Tek moleküle yüksek beta-laktamaz stabilitesi
- Geniş spektrum
- Pratik tedavi
- Tedaviye uyum kolaylığı
- Orta kulak sıvısına iyi penetrasyon

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**Özellikleri:**
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**Endikasyonlar:**
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**Doz ve Uygulama:**
Günde 2 kez kullanılır. Dose 250-1000 mg arasında değişebilir. Çocuklarda 2 kez 125 mg (2 kez 50 mg) uygulanır. Doz 250-1000 mg arasında değişebilir. Çocuklarda 2 kez 50 mg (2 kez 25 mg) uygulanır.

**Kontrendikasyonlar:**
- Sefalosporin duyarlılığı.
- Uyanlar, Penisilinler ile çapraz alerjik reaksiyon görülebilir. Kesin endikasyon olmadıkça gebelikte kullanılmamalıdır.

**Yan Etkiler:**
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**Ticari Şekiller:**
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Roche

Moklobemid Özellikleri, etkileri: Moklobemid, tip monoaminoksidaz enziminin reversibly inhibitördür. Psikolojik durumu düzeltici ve psikomotor aktiviteyi artırıcı etkisinin bir sonucu olarak AURORIX, depresyonun şiddetine göre günde 300-600 mg, iki veya üç eksi bölünerek verilir. AURORIX, depresif hastaların yaşam isteğinin kaybolduğu, bitkinliği ve dikkati yoğunlaştırma yeteneğinin zayıflaması gibi semptomların giderilmesinde etkili olur. AURORIX, uyanıklık durumunu bozma, iyileştirici etkisi olup, karaciğer ve kalbe toksik bir etkisi gözlenmemiştir.

Endikasyonlar: Depresif sendromlar ve sosyal fobi tedavisi için standart doz: Depresyon: Depresyonun şiddetine göre günde 300-600 mg, iki veya üç eksi bölünerek verilir. Sosyal Fobi: Önerilen doz, 600 mg günde iki doz olarak verilir. Bu剂量, depresyonun şiddetine göre günde 300-600 mg, iki veya üç eksi bölünerek verilir. Sosyal Fobi için önerilen doz, 600 mg günde iki doz olarak verilir. Eğer ilacın etkisini belirtmek için yeterli klinik deneyim olmaması durumda, 300 mg günde 1 doz olarak verilebilir.

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70.996 Hastada 7 yıllık çalışmalarla ispatlanmış(*)
GÜVENİLİRLIK

Günde tek doz ile her hasta grubu için uygun form seçeneği,
TABLET, SUPÖZİTUAR, AMPUL
