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Research Article

Clinico-Epidemological Profile of Children with Henoch-Schonlein Purpura at Tripoli Children's Hospital

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Abstract

Background: Henoch schonlein purpura (HSP) is one of the most common vasculitides of unknown etiology in childhood. It is a non-Granulomatous vasculitis that is characterized by deposition of immunoglobulin A, complement and immune complex in blood vessel wall as well as renal mesangium.

Aim of the study: To describe the demographic, epidemiologic, clinical characteristics and possible etiology of HSP patients.

Design: Retrospective descriptive observational study.

Patients and Methods: Medical records of all patients who diagnosed as HSP according to the European league against rheumatism and pediatric rheumatology European society (EULAR\ PReS) criteria and followed up at Tripoli Children Hospital rheumatology and nephrology departments from January 2005 to June 2017 were reviewed.

Results: 75 Children were included in the study, of these 40 were boys giving a male to female ratio of 1.14: 1. Their ages ranged from 2 to 12 years (mean age 6.5 ± 1.5 years). At the time of diagnosis, 50.7% of patients were ≤ 5 years and 93.4% were less than 10 years, with peak age onset of 2-9 years. Upper respiratory tract infection (URTI) preceded the onset of the disease in 57.3% of the patients. 100% of the patients had rash either at presentation or during the disease course. Joint involvement observed in 80%, with the ankles the most frequently affected joints. Gastrointestinal (GIT) involvement occurred in 65% of patients, the dominant digestive clinical features were abdominal pain and vomiting. Renal involvement documented in 40% with various degrees of severity, none of our patients had acute renal failure.73.3% of patients treated as in-patient and steroid was used in 53.3% with a mean duration of 10 days. None of our patients developed chronic renal failure or hypertension as long-term sequels. Conclusion: HSP is a mild disease, etiology, epidemiological, clinical findings and etiological factors of HSP patients in our region were found to be similar to those reported in the national and international studies, more research is warranted to study the prevalence and complications of HSP in Libyan based cohort.

Introduction

Henoch schonlein purpura (HSP) is one of the most common vasculitides of unknown etiology in childhood [1]. It is a non-Granulomatous vasculitis that is characterized by deposition of immunoglobulin A, complement and immune complex in blood vessel wall as well as renal mesangium [2-4].

Clinically HSP is characterized by non thrombocytopenic purpura, arthritis and arthralgia, abdominal pain and gastrointestinal hemorrhage, and glomerulonephritis [1]. A diagnostic triad of purpuric rash, arthritis, and abnormalities of the urinary sediment was proposed for the first time by Schonlein in 1837, and Henoch described the association of purpuric rash, abdominal pain with bloody diarrhea, and proteinuria in 1874 [4,5,8].

HSP is predominantly a disease of childhood, although a similar syndrome has been reported in adults [9] it occurs most frequently between 3-15 years of age with half of all cases occurred in children aged < 5 years, it is more common in male than females 1.5:1 and frequently encountered in winter and spring. The annual incidence has been reported as 10-20 per 100,000 annually [6,7].

Currently, HSP diagnosis is mainly clinical, and the first criteria that used to diagnose HSP is the American College of Rheumatology (ACR) criteria which were set in 1990 [2]. In 2010 the vasculitis working group of the Pediatric Rheumatology European Society (PReS) proposed new classification criteria for pediatric vasculitides, endorsed by the European League Against Rheumatism (EULAR), which required palpable purpura with lower limb predominance (as mandatory criteria) plus at least one among the following four features: (1) diffuse abdominal pain, (2) biopsy showing typical

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leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant immunoglobulin A (IgA) deposition, (3) arthritis or arthralgia, and (4) renal involvement (any hematuria and/or proteinuria) [3] the sensitivity and specificity of this criteria was 100% and 87% respectivly.

In 2012, a new classification was proposed as part of an overall updating of nomenclatures of systemic vasculitides, using that nomenclatures, HSP is now referred as IgA vasculitis and defined as vasculitis with IgA1 dominant immune deposit affecting small blood vessels and often involve the skin and gastrointestinal tract and frequently causes arthritis, glomerulonephritis that is indistinguishable from IgA nephropathy [11].

HSP, in general, is considered benign and self-limited disease with an excellent prognosis for most of the children. Significant morbidity and mortality is associated with gastrointestinal lesions in the short-term and with nephritis in long-term [1-7]. In two-thirds of patients, HSP runs its entire course within 4 weeks of onset, one-third to one-half of cases have at least one recurrence that commonly take place within the first 4 weeks period but may occur as late as 2 years after onset [7].

Although several reports dealt with this disease internationally, reports from Libya are few, to the best of our knowledge only one study described the HSP presentation in Benghazi was reported [16].

Patients and Methods

The medical records of all children who diagnosed with HSP from January 2005 to June 2017 and followed up in rheumatology and nephrology clinics at Tripoli Children Hospital were reviewed. Tripoli Children Hospital is the only referral centre in the west region of Libya and the only center that include pediatric rheumatology and nephrology service. The inclusion criteria used were the European league against rheumatism and pediatric rheumatology European society (EULAR\PReS) endorsed consensus criteria for HSP proposed by Ozen et al. [3]. For all patients included in the study, we collect the following data: Age, gender, season of presentation, clinical presentation, triggering factors, clinical examination at presentation, laboratory data including complete blood count, C reactive protein, ESR, and complete urinalysis. Type of treatment includes steroid, immunosuppressive drugs, angiotensine-converting enzyme inhibitors, etc and follow-up period. Data were analyzed using SPSS 16 program, descriptive statistics expressed as the mean ± standard deviation (minimum-maximum). Univariant analysis and Chi-square test were used for categorical variables.

Results

75 children with a diagnosis of HSP according to the EULAR\PReS criteria and following in rheumatology and nephrology clinics, Tripoli children hospital from January 2005 to June 2017 were included. Of these 40 were boys giving a male to female ratio of 1.14:1. Their ages ranged 2 -12 years (mean age 6.5 ± 1.5 years). 50.7% of patients were ≤ 5 years and 93.4% were less than 10 years, with peak age onset between 2-9 years. 30.7% of our patient presented during winter months, followed by 26.7% during spring (Figures 1 and 2). 78.6% of patients had a potential trigger event before HSP onset; upper respiratory tract infection (URTI) preceded HSP in 43(57.3%) patients. Fever alone in 13(17.3%) and one patient had a skin infection. epidemiological and etiological data are summarized in table 1.

Rash (mandatory diagnostic criterion) in the form of palpable non-thrombocytopenic purpura was developed in all cases (100%) during the course of the disease. And was distributed mainly over the extensor surface of the legs and buttocks, also, 3 cases had involvement of upper extremities and face. In 10 (13.3%) patients, the rash was associated with edema of hands and feet. Joint involvement (arthralgia & arthritis) occurred in 60(80%) patients, the most affected joints were the ankles in 32(41.3%) None of our patients had small joints involvement.

Gastrointestinal (GIT) involvement was reported in 49(65%), of these cases, 24(32%) had only abdominal pain, 9 (12%) had abdominal pain with vomiting and melena, and 3(4%) had occult blood in stool in addition to abdominal pain. Renal involvement was documented in 30 (40%) patients of which 8(10.7%) patients manifested as microscopic hematuria and proteinuria, 5 (6.7%) had only microscopic hematuria, 4(5.3%) had isolated hypertension while 4 patients their hypertension was accompanied with gross hematuria, testicular swelling developed in one patient. 28\30(93.3%) patients with renal involvement had their manifestations at disease onset. One patient has neurological manifestations in form of a headache, convulsion and intracranial hemorrhage which was confirmed by Brain MRI. Clinical features are summarized in table 2 and figure 3.

Laboratory finding included total blood count, shows; WBC was normal in 51\64 (79.6%) while leuckocytosis confirmed in

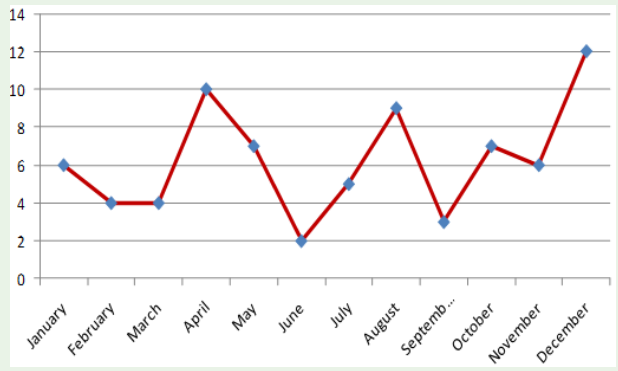

Figure 1: Monthly occurrence of Henoch-Schonlein purpura in 75 children.

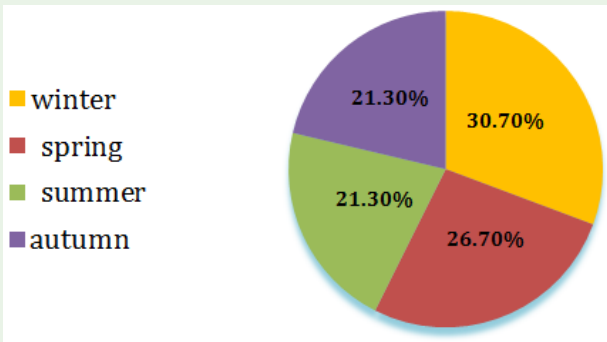

Figure 2: Seasonal distribution of disease onset showing maximum cases with onset in winter & spring.

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Table 1: Epidemiological and etiologic factors in 75 patients with HSP.

Variable	Number (%)
Age at onset(years)	
Range	2 to12 years
Mean	6.5 ± 1.5 years
Gender	
Male	40
Female	35
M:F ratio	1.14:1
Age distribution	
≤ 5 years	38(50.7%)
>5years to≤ 10 year	32(42.7%)
> 10 rears	5(6.6%)
Etiologic factor	
URTI	27 (36%)
Fever alone	13 (17.3%)
URTI and fever	16 (21.3)
Diarrhea	2 (2.7%)
Skin infection	1 (1.3%)
unknown	16 (21.4%)

13\64(20.3%), hemoglobin was normal in the majority of patients, anemia confirmed in one patient (1.56%), thrombocytosis in 5(7.8%). ESR was elevated in 23 out of the 46 tested patients(50%), C-reactive protein done in only 35 patient and was high in 16 (45.7%). complement: C3, C4 done in 23 patients and was normal in all. Throat swab detects B hemolytic streptococci in 2 out of the 7 cases tested, 2\7 (28.57%), while anti-streptolysin O titer was high in 2 out of 3 tested patients (66.6%). Skin biopsy was done in 2 patients. Laboratory finding are summarized in table 3. Regarding treatment 55(73.3%) patients needs hospitalization while 20(26.7%) treated as an outpatient. steroid either intravenously or orally was given to 40(53.3%) patient with a mean duration of 10 days, 21(28%) patient received angiotensine-converting enzyme inhibitor. 4(5.3%) patients received mycophenolate mofetil as immunosuppressive therapy for nephritis.

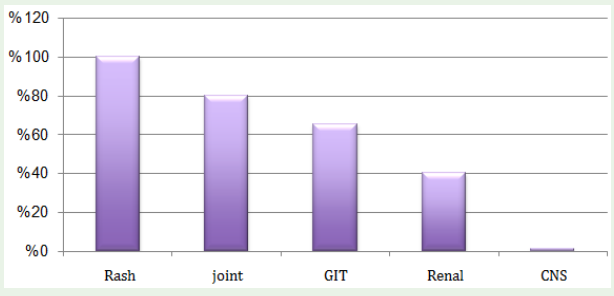

Figure 3: Clinical manifestations of HSP patients.

Table 2: The spectrum of HSP manifestations.

Variable	Number (%)
Skin, joint & GIT	24 (32.0%)
Skin and joint	18 (24.0%)
Skin, GIT , joint& renal	15(20.0%)
Skin & renal	8(10.7%)
Skin, GIT, renal	3 (4.0%)
Skin & GIT	5 (6.7%)
Skin manifestations	75 (100%)
Joint manifestations	60 (80%)
Arthralgia	15 (20%)
Arthritis	45 (60%)
GIT manifestations	49 (65%)
Abdominal pain	24(32%)
Abdominal pain+ vomiting	11(14.7%)
Abdominal pain+ vomiting+ melena	9(12%)
Abdominal pain +occult blood	3(4%)
Renal manifestations	30 (40%)
Microscopic hematuria + proteinuria	8 (10.7%)
Microscopic hematuria	5 (6.7%)
Gross hematuria + hypertension	4 (5.3%)
Gross hematuria +proteinuria	4 (5.3%)
Gross hematuria	3 (4%)
Hypertension	4 (5.3%)
Proteinuria	1 (1.3%)
Testicular swelling	1 (1.3%)
CNS manifestations	1 (1.3%)

Table 3: Laboratory finding in 75 patients with HSP.

Variable	% (NO positive\ NO tested)
Leuckocytosis	20.3% (13\64)
Anemia	1.56% (1\64)
Thrombocytosis	7.8% (5\64)
Elevated ESR	50% (23\64)
Positive C-reactive protein	45.7% (16\35)
Low C3or\and C4 serum level	0% (0\23)
Positive throat culture for B-hemolytic streptococci	28.7% (2\7)
High antistreptolysin O titer	66.6% (2\3)
Microscopic or\and macroscopic hematuria	32% (24\75)
Proteinuria	17.3% (13\75)

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Table 4: Comparison of the finding of the present study with other studies.

Variable	Our study	Libya(Benghazi) [16]	Saudi Arabia1 [12]	Saudi Arabia2 [13]	Turkey [11]
No. of Patients	75	117	29	78	151
Age (mean)	6.5 ± 1.5 yrs	4.7 yrs	7.5±3.8 yrs	6.3yrs	7.3±3.4 yrs
Gender M:F	1.14:1	1.2:1	1.1:1	1.4:1	1.47:1
URTI as triggering	43(57.3%)	-----	12(41.4%)	41(52%)	33(22%)
Season	Winter , Spring	Autumn	Winter	Autumn	Winter
Rash	75(100%)	117(100%)	29(100%)	78(100%)	151(100%)
GIT involvement	49(65%)	66(56%)	21(72.4%)	37(47%)	111(73%)
Joint involvement	60(80%)	77(66%)	24(82.8%)	52(66.7%)	91(60%)
Renal involvement	30(40%)	33(28%)	7(24.1%)	19(24%)	41(27%)
Hematuria	24(32%)	------	3(10.3%)	19(24%)	38(25%)
Proteinuria	13(17.3%)	------	----	10(12.8%)	24(16%)
Others					
CNS	1(1.3%)	0%	----	2(2.5%)	1(0.6%)
Cardiac	----	0%	----	------	1(0.6%)
Testicular	1(1.3%)	6(5%)	2(6.9%)	7(15%)	----

URTI: Upper Respiratory Tract Infection; GIT: Gastrointestinal System; CNS: Central Nervous System.

Discussion

Henoch Schonlein purpura is the most common childhood vasculitis [1]. In the present study, 75 patients were evaluated in terms of their epidemiologic, clinic and possible etiologic characteristics and finding were compared with national and international studies. The mean age was 6.5 ± 1.5 years, in terms of the mean age the present study was observed to be consistent with those from East of Libya, Saudi Arabia and turkey (Table 4). Comparable studies reported that most patients with HSP presented during winter and autumn months (Table 4), most patients in our study reported symptoms in winter months, 30.70% with 2nd peak in spring months. Of the study patients 40 were boys and 35 were girls with M:F ratio of 1.14:1 and this ratio was similar to those in the literature (Table 4), however there are also reports indicating that HSP is more frequent in females [10]. In our study 57.3% of patients had symptoms of URTI before disease onset compared with 41.4% and 52% in Saudi studies; in turkey study only 22% of their patients had URTI before disease onset. URTI ranked first as a triggering factor in a prospective study from Finland, streptococci infection was reported in 36% of patients [15].

24(32%) of our patients had the classic triad of HSP, rash, joint involvement and gastrointestinal involvement. Classic skin purpura present in all patients, and the rash was distributed over the lower limbs and buttocks in 84% of patients. Joint involvement observed in 80% of our patients, either as arthritis or arthralgia, similar results was reported by other authors who found that 60-82.8% of patients with HSP had articular manifestations. Ankles were the most commonly involved joints; none of our patients had small joint involvement. In comparable studies GIT involvement has been reported in approximately two-thirds of HSP patients (47%-73%), which is consistent with our report as GIT involvement occurred in 65% of our patients. Abdominal pain with or without vomiting was the most frequent symptom. Both upper and lower GIT bleeding can occur in HSP patients, in our study melena was reported in 9(12%) patients and 3(4%) had positive occult blood in stool examination. No significant complications as gut perforation or intussusceptions were reported. Renal involvement is the predicting and affecting factor of long term prognosis in HSP. The rate of renal involvement in HSP varies between 24 to 28% in the comparable studies (Table 4). In current study 30 (40%) of our patients had renal involvement during their disease onset, microscopic and macroscopic hematuria was the commonest as it was documented in32% of patients. The high incidence of renal involvement in our study may be related to the causative and environmental factors. Although renal involvement was high comparable with other studies (Table 4) none of our patient progressed to acute renal failure, nephrotic syndrome or chronic hypertension. Scrotal swelling is a well-known manifestation of HSP, in current study only one boy reported to have testicular swelling compared with 5-15% of patients in comparable studies. Also, central nervous system reported in one case who presented with convulsion due to CNS bleeding which confirmed by MRI brain. There are no diagnostic laboratory tests for HSP and most laboratory studies are utilized to exclude other conditions that resemble HSP. In the current study, CBC and ESR done in 64 (85.3%) patient elevated ESR found in 50% of patients. Conservative treatment in the form of hydration, analgesics and non-steroidal anti-inflammatory drugs are effective in majority of cases, in this study; 73.3% of patients treated as inpatient compared with 26.7% treated as an outpatient and this reflect the

high rate of hospitalization. More than half of cases 53.3% treated by steroids either intravenously or orally with a mean duration of 10 days.

Retrospective nature of the study caused limitations in the evaluation of the data, and also small number of the cases which may be due to nature of the study as we include patients following in rheumatology and nephrology clinics, mild cases with HSP treated in the outpatient department with- out a referral to follow-up. Present study was similar to the other HSP studies in East region of Libya, Saudi Arabia and Turkey in terms of clinical features of patients and outcome.

Conclusion

HSP is a self-limited disease; it can lead to long-term morbidity especially in the patient with renal involvement. More research is needed to study HSP in Libya to find the prevalence and out- come of the disease.

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