

**HENOCH-SCHONLEIN PURPURA - CLINICAL AND INVESTIGATIVE STUDY OF 20 PATIENTS**

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**ABSTRACT****BACKGROUND**

Henoch-Schonlein Purpura (HSP) is a systemic small vessel vasculitis with multiorgan involvement characterised by palpable purpura, arthritis, abdominal pain and renal disease. The aetiology and clinical features are varied but histopathology is characterised by leucocytoclasia with deposition of IgA immune complexes.

Aims and Objectives- The present study was done to know the spectrum of various cutaneous manifestations, systemic involvement and aetiology of Henoch-Schonlein purpura.

**MATERIALS AND METHODS**

The study was carried out on patients who were clinically diagnosed as Henoch-Schonlein purpura fulfilling EULAR criteria, attending the Outpatient Department of Dermatology, Venereology and Leprosy, King George Hospital, affiliated to Andhra Medical College, Visakhapatnam from January 2014 to December 2015. It is a cross-sectional type of study and a total of 20 cases were studied.

Inclusion Criteria- All patients attending to OPD, clinically diagnosed as Henoch-Schonlein purpura fulfilling EULAR criteria, irrespective of age and sex were included in study.

Exclusion Criteria- Patients with thrombocytopenic purpura, disorders of coagulation and on warfarin/heparin treatment.

**RESULTS**

Out of 20 patients enrolled in study, most common age group is in between 0-20 years, female preponderance, all patients presented acutely with lesions less than 6 weeks duration, 60% of patients had symptoms of burning and itching in lesions, 30% had pain in lesions, 30% had history of low grade fever, 80% of patients had arthralgia, 65% had gastrointestinal symptoms, 30% had history of sore throat, none had history of significant drug exposure within 6 weeks of onset of lesions. Majority of patients had elevated ESR (70%). 30% of patients had ASO titres positive, of which only 15% had throat swab positive for beta-haemolytic streptococci, ANA titres were negative in all patients, renal involvement in the form of albuminuria was seen in 30% and haematuria in 20%. Out of 30% of patients with renal involvement only 5% had significant renal impairment with elevated blood urea and serum creatinine levels. Histopathology of all patients showed leucocytoclastic vasculitis with deposition of IgA.

**CONCLUSION**

In the present study, aetiology in most of patients is idiopathic but upper respiratory tract infection was found to be the commonest aetiology among others. So, infections like sore throat should be ruled out as prompt treatment. All presented with arthralgia but gastrointestinal tract involvement was also high with presentation as acute abdomen in one patient. Renal involvement was present in 30% but there is renal impairment in 5% of patients.

**KEYWORDS**

Henoch-Schonlein Purpura, Arthritis, Pain Abdomen, Renal Involvement.

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**BACKGROUND**

Henoch-Schonlein purpura (HSP) is a systemic small vessel vasculitis with multiorgan involvement characterised by palpable purpura, arthritis, abdominal pain and renal disease. The aetiology and clinical features are varied but histopathology is characterised by leucocytoclasia with deposition of IgA immune complexes.<sup>[1,2]</sup>

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**MATERIALS AND METHODS**

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**Inclusion Criteria**

All patients attending to OPD, clinically diagnosed as Henoch-Schonlein purpura fulfilling EULAR criteria,<sup>[3]</sup> irrespective of age and sex were included in study. The criteria to diagnose HSP include Palpable purpura, (mandatory) +≥ one of the following: 1. Diffuse abdominal pain. 2. Histopathology: typical LCV with predominant IgA deposits or proliferative glomerulonephritis with predominant IgA deposits. 3.

Arthritis or arthralgia. 4. Renal involvement (Proteinuria and/or haematuria). All patients who are fulfilling EULAR criteria were included for clinical, aetiological and laboratory correlation.

**Exclusion Criteria**

Patients with thrombocytopenic purpura, disorders of coagulation and on warfarin/heparin treatment.

The study protocol included detailed history and clinical examination. The following investigations were done: complete haemogram, renal and liver function tests, urine analysis, antinuclear antibody (ANA), antineutrophilic cytoplasmic antibody (ANCA), Rheumatoid factor (RF), C-reactive protein (CRP), cryoglobulins, lupus anticoagulant, HBsAg, anti-HCV antibody, antistreptolysin O (ASO) titre, throat swab for culture and sensitivity, Chest X-ray and Mantoux test. Other tests carried out depending upon the clinical indications included stool for occult blood, 24-hour urinary protein, sputum for acid-fast bacilli (AFB), HBV RNA, Anti-HBc IgM, dsDNA, anti-Ro/anti-La antibody, IgA, IgG, IgM, C3, HIV serology, X-ray of wrist and feet, arterial and venous Doppler, ultrasonography (USG) abdomen, echocardiography, renal biopsy, contrast-enhanced computed tomography of chest, digital subtraction angiography (DSA) and contrast-enhanced magnetic resonance angiography. Two skin biopsies (punch biopsy, 4 mm) were taken in all cases, one each for routine histopathology and direct immunofluorescence (DIF).

**RESULTS**

20 patients who were clinically diagnosed as Henoch-Schonlein purpura and histologically confirmed as leucocytoclastic vasculitis with deposition of IgA immune complexes were included in the study and all parameters were evaluated. Out of 20 patients enrolled in study, most common age group is in between 0-20 years around 70% remaining patients belong to age group of >40 years [Table 1]. Out of 70% of patients who were of age group less than 20 years, majority of patients belong to age group of 11-15 years. There was female preponderance with male to female ratio of 1:1.8 [Table 2]. All patients presented acutely with lesions less than 6 weeks duration, 60% of patients had symptoms of burning and itching in lesions, 30% had pain in lesions, 30% had history of low grade fever, 80% of patients had arthralgia, 65% had gastrointestinal symptoms, 30% had history of sore throat, none had history of significant drug exposure within 6 weeks of onset of lesions [Table 3]. Majority of patients had elevated ESR (70%). 30% of patients had ASO titres positive, of which only 15% had throat swab positive for beta-haemolytic streptococci, ANA titres were negative in all patients, renal involvement in the form of albuminuria was seen in 30% and haematuria in 20%. Out of 30% of patients with renal involvement only 5% had significant renal impairment with elevated blood urea and serum creatinine levels [Table 4]. Histopathology of all patients showed leucocytoclastic vasculitis with deposition of IgA.

Age Distribution	Number of Patients
0-20 years	14
>20 years	6
<b>Table 1. Age Distribution in Present Study</b>	

Sex Distribution	Number of patients
Males	13
Females	7
<b>Table 2. Sex Distribution in Present Study</b>	

Systemic symptoms	
Fever	
Sore throat	
Upper respiratory infection other than sore throat	
Pain abdomen	
Arthralgia	
Haematuria	
<b>Table 3. Systemic Symptoms</b>	

Laboratory investigations	
Elevated total leucocyte count	
Elevated ESR	
Haematuria	
Albuminuria	
Elevated blood urea and serum creatinine	
<b>Table 4. Investigations</b>	

**DISCUSSION**

Henoch-Schonlein purpura is a systemic self-limited small vessel vasculitis with multiorgan involvement characterised by palpable purpura, arthritis, abdominal pain and renal disease. The aetiology and clinical features are varied but histopathology is characterised by leucocytoclasia with deposition of IgA immune complexes. It is the most common childhood vasculitis, with an annual incidence of about 10 cases per 100,000.<sup>[1]</sup> HSP was announced in association with infections, medications, vaccination. The HSP children are mainly between the ages of 5 and 15 years.<sup>[2]</sup> In the present study, most common age group presented was 0-20 years around 70% followed by 20-40 years (30%) which was consistent with studies done by Salusbury et al. Out of 70% of patients presented between 0-20 years, most of the patients belong to age group of 11-15 years. In the present study, there is a female preponderance of 65% which was in consistence with studies done by Calvino et al,<sup>[4]</sup> but contrast with studies done by Salusbury and Chen et al.<sup>[2,5]</sup>

The aetiology was idiopathic in most cases around 60% followed by infections in around 40%. Out of 40% of patients, 30% had preceding sore throat having ASO titre positivity. Out of 30% of patients only 15% of patients' beta-haemolytic streptococci was isolated on culture and sensitivity and in remaining 15% Streptococcus pneumonia was isolated. Remaining 10% of patients with infectious aetiology had preceding upper respiratory tract infection as viral rhinitis. This was in consistence with the studies done by Salusbury et al.<sup>[2]</sup> Some authors had reported the trigger events including vaccinations, insect bites, and drugs have been connected with HSP<sup>[6,7]</sup>

In the present study, all patients presented with purpura. This was in consistence with the previous studies. Lesions extended above waist in 20%. Out of 20 patients, two had superficial ulcers and one patient who was a female child presented with ulcerations and purpura over perineum. The most common symptoms at the site of lesion was itching and burning which was seen in 60% and 30% had pain at the site of lesion. Majority of the patients presented with arthralgia around 80%, knee and ankle joints were commonly involved which was in consistence with the previous studies.<sup>[8]</sup> 30% of

patients had haematuria and 65% of patients had abdominal pain.

In routine laboratory investigations, 40% had elevated total leucocyte count, 70% had elevated ESR. On routine urine analysis, 20% had haematuria and 30% had albuminuria. Out of 30% of patients with renal involvement, only 5% had significant renal impairment with elevated blood urea and serum creatinine. This is not correlating with advanced age and lesions above the waist as the study sample size was less. All the patients showed leucocytoclastic vasculitis on histopathology H&E stain of skin biopsy specimen from the lesion. On direct immunofluorescence examination, all showed IgA deposits in the vessel walls. Out of twenty patients, four showed additional deposits of IgG and complement.

#### CONCLUSION

In the present study, aetiology in most of the patients is idiopathic, but upper respiratory tract infection was found to be the commonest aetiology among others. So, infections like sore throat should be ruled out for prompt treatment. Systemic manifestations like arthralgia, abdominal pain and haematuria was present in present study. But significant renal impairment was found in one patient and this was not corresponding to site of lesion and advanced age as the study sample was less.

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