

Henoch-Schönlein purpura in an adult patient, presenting with severe nephrotic syndrome

Ađır nefrotik sendrom ile bařvuran eriřkin bir hastada Henoch-Schönlein purpurası

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ABSTRACT

Henoch-Schonlein Purpura (HSP) in adults is a rare clinical finding. Even though HSP nephritis frequently leads a course with asymptomatic urinary findings, presentation with severe nephrotic syndrome is quiet rare and associated with poor renal outcome. In this article, a patient who was admitted to the hospital with severe nephrotic syndrome and diagnosed with HSP nephritis is presented. He also achieved full remission with steroids and cyclophosphamide treatment.

Keywords: Henoch-Schonlein purpura, nephrotic syndrome, adult

ÖZ

Eriřkinlerde Henoch-Schönlein Purpurası (HSP), ender görölen klinik bir tablodur. HSP nefriti sıklıkla asemptomatik idrar bulgularıyla seyretmekle birlikte, ađır nefrotik sendromla bařvuru oldukça enderdir ve kötü renal sonlanımla ilişkilidir. Burada ađır nefrotik sendromla bařvurup HSP nefriti tanısı alan, steroid ve siklofosfamid tedavisiyle tam remisyona sađladığımız bir olgu sunulmaktadır.

Anahtar kelimeler: Henoch-Schönlein purpurası, nefrotik sendrom, eriřkin

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INTRODUCTION

HSP is a disease characterized by leukocytoclastic vasculitis of small vessels with resultant deposition of immune complexes containing IgA antibody ^(1,2). Ninety percent of the cases occur between the ages of 3 and 15 in the pediatric group, and the incidence of the disease in the adults above age 20 is between 0.1 to 1.2 in one million people ⁽³⁾. Patients frequently exhibit purpuric rash, arthritis and/or arthralgia, abdominal pain and renal involvement. Renal involvement in adults, apart from the children, is more common and indicates poor prognosis. Renal involvement is generally correlated with the severity of the systemic disease. Renal findings exhibit a wide spec-

trum from hematuria and proteinuria to nephritic syndrome with severe kidney failure. However, it must be noted that presentation with nephrotic syndrome is still infrequent. Here in we report on a patient who presented himself at the emergency room with generalized rash, abdominal pain and nephrotic syndrome, and subsequently diagnosed as HSP.

CASE REPORT

A 42-year-old male patient came to the emergency room, complaining of abdominal pain, edema in both legs and rash. Patient's history revealed nonsteroidal anti-inflammatory drug use for the symptomatic treatment of the possible upper respiratory tract

infection he had 10 days prior to the admission. After that, he developed rash and generalized swelling in both lower extremities. Skin biopsy was performed at the outpatient clinic because of these skin rashes, but the patient came to the emergency room before the biopsy results were obtained. In his anamnesis, there was no history of chronic disease or regular drug use. His blood pressure was 130/80 mmHg and apart from local tenderness at both legs during palpation, there was no other pathological finding at the physical examination of the cardiovascular and respiratory system and abdomen. On the legs, +2 grade pitting edema and non-blanching rashes with irregular borders were present. Urinalysis performed at the emergency room revealed 3+ proteinuria and 7-8 erythrocytes. Abdominal ultrasound did not show any pathology. The patient was hospitalized in the clinic of internal medicine with initial diagnosis of vasculitis and nephrotic syndrome.

Detailed laboratory test results were as follows: Hgb 12.6 g/dL, serum urea 67 mg/dL, serum creatinine 1-2 mg/dL, LDL 304 mg/dL, albumin 2.9 g/dL and 24-hour urine protein: 11.1 g. ANA, c-ANCA, p-ANCA tests all yielded negative results. Patient priorly received intravenous hydration which faded his purpuric lesions. Abdominal ultrasound performed because of his abdominal pain revealed peripheral millimetric outpouchings at the wall of the colon in the left lower quadrant which were interpreted as signs of diverticulitis. To further support the diagnosis, abdominal CT scan was performed. Non-contrast CT scan was preferred due to the renal function test results nearing to upper limit of normal (ULN). and massive proteinuria which did not collaborate the diagnosis of diverticulitis. However, the reason for this preference was thought to be not being able to administer contrast agent to this patient.

Due to the massive proteinuria of the patient and increased renal function test values close to ULN, nephrology consultation was requested, and nephrologists strongly suggested a kidney biopsy. Histopathological examination of the biopsy specimen showed diffuse proliferative glomerulonephritis

pattern with endocapillary hypercellularity. Histopathological findings were consistent with IgA nephropathy. On the basis of these findings and biopsy results, the patient was given 500 mg methylprednisolone for 3 days and a single loading dose of (750 mg) cyclophosphamide. Daily urinary clearance of protein decreased to 3.3 g after the treatment. The patient was discharged with prescription of daily oral doses of 48 mg methylprednisolone and once monthly intravenous injection of 1 g cyclophosphamide. At each follow-up visit, methylprednisolone dose was gradually decreased to the point where treatment was maintained only with once monthly doses of 1 gram cyclophosphamide. Approximately one year after the diagnosis, the last 24-hour urine test revealed 0.3 g protein in the urine. The patient is still visiting our clinic for regular nephrology check-ups. The patient's laboratory parameters at the first admission and last follow-up visit are shown in Table 1.

Table 1. Laboratory parameters of the patient on first and last admissions.

	First Admission	Last Admission
Blood pressure (mmHg)	130/70	120/70
P. Urea (mg/dL)	67	26
P. Cre (mg/dL)	1.2	1.3
P. Albumin (g/dL)	2.9	4.3
T. cholesterol (mg/dL)	467	182
TG (mg/dL)	171	142
HDL (mg/dL)	113	33
LDL (mg/dL)	304	128
Hemoglobin (g/dL)	12.6	12.5
Proteinuria (mg/day)	11190	795
Urinalysis Examinations	15-20 erythrocytes	Normal

DISCUSSION

Although HSP is a common disease of childhood, it may be also rarely seen in adult patients with different clinical manifestations. In adults, renal involvement is more frequent and associated with poor renal prognosis. Renal involvement manifests itself with diverse clinical and morphological features. Presentation with severe nephrotic syndrome is very rare. We discussed our patient, because he was in the adult age group, and presented with severe nephrotic syndrome, and responded well to the immunosup-

pressive treatment.

HSP, also known as the IgA vasculitis, is supposed to be triggered by some infections and chemical substances. However, exact underlying cause of the disease is still unknown. Immunologic, genetic and environmental factors all contribute to the development of the disease⁽⁴⁻⁶⁾. While all HSP patients have purpura, up to 84% of the patients show signs of arthritis/artralgia⁽⁷⁾. Despite the high percentage of occurrence of this symptom, our patient did not exhibit joint involvement. This was one of the reasons why we thought the case was worth sharing.

Renal involvement is seen nearly in 21-54% of the children diagnosed as HSP⁽⁸⁾. Possibility of renal involvement that leads to end-stage renal failure is much higher in adults than in children^(9,10). Hematuria and/or nephritic level proteinuria are frequent clinical findings of the HSP nephritis which are present in 70-80% of the patients. Presentation with nephrotic syndrome is rarely seen, and in a study of pediatric age group, only 4% of the 261 patients with HSP presented with nephrotic syndrome⁽¹¹⁾. In the adult age group, there is not enough data on the relationship between HSP nephritis and nephrotic syndrome, but some studies demonstrated that incidence is higher in adults^(10,12-14). In a retrospective study of 250 HSP diagnosed patients with average age of 50, renal failure (GFR<50%) within 4 months of after onset of symptoms developed in 32% of the patients and almost all of them was accompanied with hematuria (93%) and proteinuria (99%)⁽¹⁴⁾. In another study of 136 adult patients, who were followed up for an average of 5.5 years, 13% of the patients needed dialysis, and in 25% of them serum creatinine values doubled, while 16% of the patients had proteinuria with normal serum creatinine levels⁽¹⁵⁾.

Histopathologic findings in adult age groups can also be different than those detected in the pediatric age group. Various studies have shown that chronic findings stand out in adults. In a study⁽¹⁶⁾, Shan Lu and his associates asserted the presence of a correlation between age and chronic findings. Indeed when compared to the pediatric age group, chronic symp-

toms were statistically found at higher frequencies in adults. For the pathologic categorization of the HSP nephritis, ISKDC classification is used and most frequently patients are in the 3a-3b group (focal and/or diffuse mesangial proliferation with <50% crescents). Frequency of 3a-3b varies between 40 and 50%. Our patient was classified in group 6, with an incidence of less than 3-4% in all pediatric cases with HSP nephritis. There is no adequate information pertaining to the same topic in adult age groups.

In the treatment of HSP, most of the time, supportive treatments such as hydration, resting and analgesics are applied. In cases of major organ system involvements, such as GIS and kidney, mainly and firstly use of glucocorticoids and immunosuppressive agents such as cyclophosphamide is recommended. In medical literature, there is no sufficient data on the treatment of a patient with group 6 HSP nephritis. Even though our patient had not crescentic findings, he had serious active diffuse endocapillary proliferation, which is why he was given cyclophosphamide in addition to steroids. Clinical response to therapy was outstanding during the follow-up period. In these patients, steroids and cyclophosphamide can be an efficient treatment option.

In conclusion, HSP nephritis, even though rare, may present with manifestations of severe nephrotic syndrome. In patients with rash, HSP nephritis should be considered in the differential diagnosis of severe nephrotic syndrome.

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