**Case Report**

**Systemic Lupus Erythematous with Severe Nephritis That Mimicked Henoch-Schoenlein Purpura**

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**Abstract**

**Introduction**: Systemic lupus erythematous (SLE) belongs to a family of related autoimmune rheumatic disorders that are capable of affecting multiple organs, and they are all associated with a variety of autoantibodies. Henoch Schoenlein purpura (HSP) is a sort of systemic vasculitis that is not associated with auto-antibodies and can affect different organs including the kidneys.

**Case report**: A 12 year-old girl presented with abdominal pain, low grade fever, swollen and tender feet and left hand, skin rash on the lower extremities, and high blood pressure. Initial laboratory tests revealed severe proteinuria, microscopic hematuria and low C3 level. Renal biopsy showed diffuse proliferative glomerulonephritis with IgA, fibrinogen and C3 deposits. The case was accordingly diagnosed as HSP with severe IgA nephropathy. Treatment was started with mycophenolate mofetil (MMF) and pulse methylprednisolone followed by prednisolone. The patient improved and treatment was discontinued after 5.5 months. One month after withdrawal of her medications, the patient presented again with serositis and recurrent proteinuria. Both antinuclear antibodies (ANA) and anti dsDNA were positive. At this point she was diagnosed to have SLE disease and immunosuppressive treatment was restarted. Following this, symptoms disappeared, proteinuria regressed and anti-dsDNA titer dropped.

**Conclusion**: This case presented with features of HSP and was later-on diagnosed to have SLE. This kind of clinical overlapping has not been reported in the literature to the best of our knowledge.

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**Introduction**

Systemic lupus erythematous (SLE) belongs to a family of related autoimmune rheumatic disorders, including dermatomyositis, scleroderma, polymyositis, mixed connective tissue disease, and rheumatoid arthritis. All of these disorders are capable of affecting multiple organs, and they are all associated with a variety of autoantibodies that rise and fall depending on the clinical disease pattern and/or the treatment given. Because symptoms in systemic rheumatic diseases often overlap, the diagnosis of SLE is not always clear-cut and true overlapping has been frequently reported in the literature. Henoch Schoenlein purpura (HSP) is a sort of systemic vasculitis that is not associated with auto-antibodies and can affect different organs including the kidneys. Its diagnosis is mainly clinical. We report on a case that presented with "typical" clinical features of Henoch-Schoenlein purpura with renal biopsy features of IgA nephropathy. At a later stage the full blown features of systemic lupus erythematosus became apparent.

**Case report**

A 12 year-old previously healthy girl, was referred to our hospital with a history of recent symptoms including abdominal pain, low grade fever, swollen and tender feet and left hand, skin rash on the lower extremities, left-sided neck pain and vomiting. On presentation her general condition was stable but her blood pressure was 138/100 mmHg and her temperature 37.8 C. Her body weight was 61 kg and her body surface area (BSA) was 1.6 m². On admission she had purpuric, purple-colored
skin rash on the lower extremities, no pitting edema, no hepatosplenomegaly, no lymphadenopathy or joint swelling. Other systems were unremarkable.

Initial laboratory tests revealed severe proteinuria (6–7 g/day) and microscopic hematuria (50 RBCs/µl). Serum creatinine was 1.0 mg/dl, albumin 24 g/l, ESR 17 mm/hour, hemoglobin 10.8 g/dl, Platelets 170,000/µl, WBC 7500/µl; normal coagulation profile, cholesterol 328 mg/dl (high), antistreptolysin-O (ASO) titer 400 units, C3 0.29 g/l (low) and C4 0.16 g/l (lower normal range).

Our primary clinical diagnosis was Henoch-Schoenlein purpura (HSP) with significant renal involvement. Renal biopsy was performed on the third admission day. The examined 29 glomeruli showed diffuse increase in the mesangial cells and matrix, accentuation of glomerular size and increased lobulation of some glomeruli. Immunofluorescent study revealed mesangial and membranous granular immunoreactivity to IgA, C3 and fibrin, with no immunoreactivity to IgG, IgM or C4. The conclusion of the renal histology was a diffuse proliferative glomerulonephritis with IgA, fibrinogen and C3 deposits.

According to the clinical presentation, laboratory results and histopathology findings, the case was diagnosed as HSP with severe glomerulonephritis and nephrotic syndrome. This was based on the pathognomonic clinical symptoms and signs of HSP, and the fact that 10% of HSP cases can have low C3 complement. Moreover, the IgA deposits in the renal biopsy and the absence of IgG were also supportive of this diagnosis.

Treatment was started with pulse methylprednisolone at a dose of 1g/m²/dose, a total of three doses, followed by oral prednisolone at a dose of 80 mg daily and MMF 750 mg twice daily. Hydroxychloroquine was also added. After starting this treatment the clinical and biochemical status improved, reflected by the disappearance of symptoms and the regression of proteinuria as well as normalization of C3, C4 and decrease of anti-dsDNA titer. Ten months later the laboratory tests showed fairly well controlled SLE including 24 hour urine protein of 0.2 g/day, normal C3 and C4 complement, anti dsDNA 42 IU/ml and ESR 18 mm/hr. The urine sediment was almost normal.

**Discussion**

To the best of our knowledge this is the first reported case of SLE that clinically presented as HSP with severe renal involvement as nephrotic-nephritic syndrome and later-on developed clinical and laboratory manifestations of SLE with markedly positive serology. Systemic lupus erythematosus (SLE) belongs to a family of related autoimmune rheumatic disorders, including dermatomyositis, scleroderma, polymyositis, mixed connective tissue disease, and rheumatoid arthritis. All of these disorders are capable of affecting multiple organs, and the symptoms in systemic rheumatic diseases often overlap as has been repeatedly reported in the literature [1-9].

In our case, the initial clinical diagnosis of Henoch-Schoenlein Purpura was clinically confirmed by three different skilled pediatricians. The renal histology was a diffuse proliferative glomerulonephritis with fibrinogen and C3 deposits which can be a feature of either HSP or SLE nephritis. However, the positive immunostaining for IgA in renal biopsy gave more support to the clinical diagnosis of HSP. The low level of complement C3 has been described occasionally in HSP cases in the literature [10] and deposition of IgA in the kidney are typical finding in HSP nephritis [11].

The initial clinical and laboratory response after starting the intensified immunosuppressive treatment with steroids and MMF was first regarded as one of two
SLE that mimicked Henoch-Schoenlein Purpura

possibilities: first, the glomerulonephritis responded well to this treatment; second, this could have been a spontaneous improvement of HSP nephritis. Because data from randomized clinical trials are sparse, there is no clear path for treatment recommended for the established renal disease in HSP [12, 13].

However, as the patient developed frank clinical and laboratory manifestation of SLE after discontinuation of this treatment, we retrospectively proposed that this could have been a good response of SLE nephritis to immunosuppressive therapy [14, 15].

Conclusion

In conclusion, this case presented with clinical features of Henoch-Schoenlein purpura and later-on presented with findings supporting the diagnosis of systemic lupus erythematosus. To the best of our knowledge, this kind of clinical overlapping has not been reported in the literature so far.

References


