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Strongyloides Stercoralis Infection Associated Henoch-Schönlein Purpura : A Case Report

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Abstract

Introduction: Henoch-Schönlein purpura (HSP) is a small blood vessel vasculitis. The exact cause of the disease is unknown. *Case report:* We reported a case of 40-year-old male who presented with the appearance of red macules on his extremities and face, along with vomiting, and pain in the abdomen and joints. The patient was initially diagnosed with Henoch-Schönlein purpura. Renal biopsy was suggestive of IgA nephropathy. One months later, he developed chronic diahorrea and duodenal biopsy shows strongyloides stercoralis (s. stercoralis) larvae and was treated with ivermectin following which his diahorrea subsided and later there was complete resolution of active urine sediments. *Conclusion:* The time course of the disease and present knowledge concerning the pathogenic mechanisms of HSP suggest that s. stercoralis infection could have caused HSP in the presented patient, which was complicated by nephritis.

Keywords: Henoch-Schönlein Purpura; Strongyloides Stercoralis.

Introduction

Henoch-Schönlein purpura (HSP) is a small blood vessel vasculitis, which usually manifests during childhood and is characterized by the presence of immunoglobulin A1 (IgA1) deposit [1]. HSP is a self-limiting, systemic, non-granulomatous vasculitis with multiorgan manifestations. The exact cause of the disease is unknown.

Case Report

A 40 year-old male presented to us with mild diffuse abdominal pain, red macules on his extremities and face, vomiting and large joint pain of one month duration. He did not have any history of hypertension or diabetes mellitus. Skin macules did not blanch on compression and there was no associated itching. Physical examination revealed mild diffuse abdominal tenderness and tenderness of large joints in the lower limbs. His blood pressure was 140/90 mm of hg with heart rate of 98/minute. Laboratory tests showed hemoglobin of 10g/dl, total count of 9600 cells/cumm, platelet count 2.2 lakhs / cumm. Blood urea was 36 mg/dl, creatinine 1.5 mg/dl, sodium 137mmol/dl, potassium 3.7mmol/dl. Liver function test were normal. Urine analysis showed 3+ proteinuria, RBC's 10-15/high power field (hpf), WBC's 5-8/hpf. Urine culture was sterile. HIV, HbsAG and anti HCV were negative. Complement levels were within normal limits. Anti nuclear factor, anti-ds DNA and ANCA were negative. 24 hour urinary protein estimation was 2.4 gms/day. Ultrasound showed normal sized kidneys.

Skin biopsy (Fig. 1) was done which showed perivascular and interstitial infiltrates of lymphocytes with many neutrophils infiltrating the wall in the dermis, along with extravasated RBCs and karyorrhectic debris, which was suggestive of leucocytoclastic vasculitis.

Renal biopsy (Fig. 2) showed 12 glomeruli of which one was globally sclerosed. Two of the glomeruli showed segmental increase in mesangial matrix and cellularity. Interstitium and blood vessels were normal. Immunofluorescence (IF) showed focal and global mesangial granular deposits of IgA which was consistent with IgA nephropathy. Based on skin and renal biopsy HSP was diagnosed.

He was started on telmisartan 40 mg/day and wysolone 1 mg/kg/day as antiproteinuric

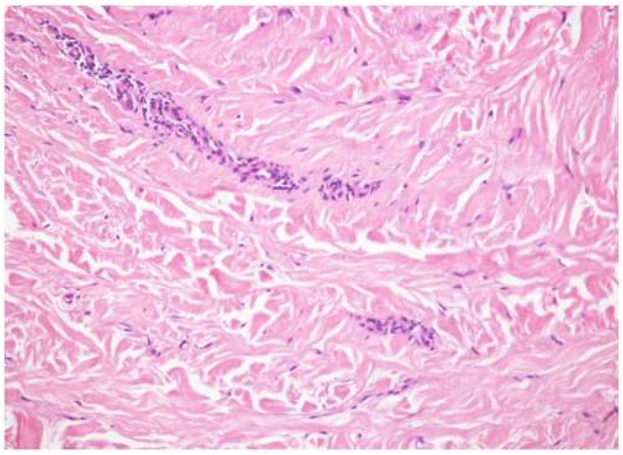

Fig.1: Skin biopsy showing leucocytoclastic vasculitis.

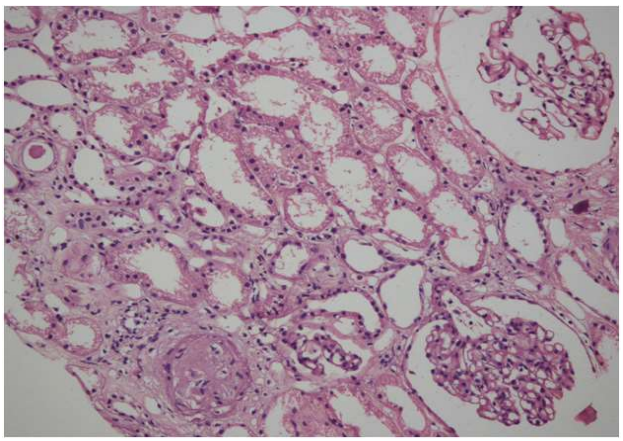

Fig.2: Renal biopsy showing features suggestive of IgA Nephropathy.

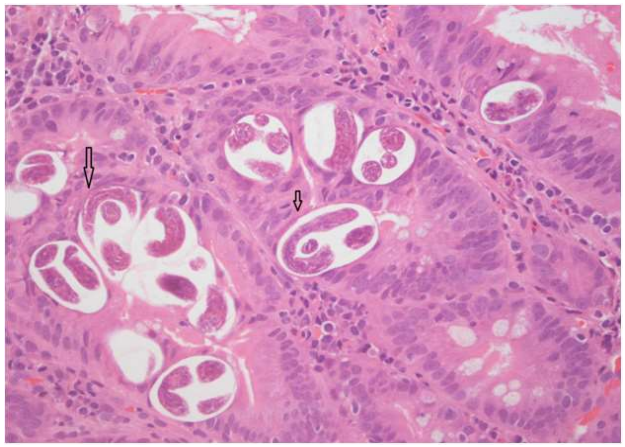

Fig. 3: Duodenal biopsy showing strongyloides larvae.

measure. He again presented to us one month later with watery loose stools mixed with blood about 6-7 episodes per day, associated with low grade fever, vomiting, reduced oral intake and weight loss (about 4 kilograms) since 20 days. On further laboratory evaluation, his serum creatinine had worsened to 2.8 mg/dl. Urine analysis showed persistence of proteinuria and microscopic hematuria but skin lesions had disappeared He received intravenous fluids and antibiotics, but diarrhea continued to persist. Wysolone and telmisartan was stopped. As a part of his workup, typhidot, weil felix, VDRL were nonreactive. Upper gatroduodenoscopy showed a gastroduodenal ulcer and biopsy from the same area showed (Fig. 3) presence of many larval forms of strongyloides in the gastric glands along with dense inflammatory cells infiltrates composed of plasma cells, lymphocytes and many eosinophils in the lamina propria. However, stool routine and microscopy could not demonstrate any larval forms of strongyloides.

He received oral ivermectin for 7 days following which he improved symptomatically and apetite improved and renal functions recovered to 1.2 mg/dl. 15 days later, there was complete resolution of proteinuria and absence of active urine sediments.

Discussion

The presented patient was initially diagnosed with Henoch-Schönlein purpura according to the 2010 EU-LAR/PRINTO/PRES (EULAR– The European League Against Rheumatism; PRINTO–Paediatric Rheumatology International Trials Organisation; PRES–Paediatric Rheumatology European Society) criteria [2], based on characteristic palpable purpuric rash without thrombocytopenia, associated with pain in the abdomen and joints and signs of kidney damage. Our patient had rash associated with abdominal pain and a brief period of intermittent appearance of fresh blood in the stool. After detection of s.stercoralis larvae in duodenal biopsy samples and upon administering ivermectin, there were no further bursts of rash nor gastrointestinal complaints. Based on the present knowledge, s. stercoralis may be considered as the initiator of HSP in our patient.

HSP aetiology is unclear, but possible causes might be bacterial, viral and parasitic infections [3], alterations in secretion of interleukins (interleukin 1 and 6) [4] or growth factors (platelet derived growth factor, transforming growth factor β) [5]. The key role in the pathogenesis of HSP most probably belongs to abnormal glycosylated IgA1 [6] from the mucosal tissues and bone marrow [7]. Therefore, we suggest that infection caused by s. stercoralis provoked the production of IgA1 in the intestinal mucosa, which preceded the forming and precipitation of immune complexes containing aberrantly glycosylated IgA1 leading to the development of vasculitis and clinical manifestations of HSP. However, our patient's urine examination showed microhematuria and significant proteinuria in the 24-hour urine collection test.

Recent papers describe cases with s.stercoralis infection leading to renal damage and the development of nephrotic syndrome [9,10], but direct infection of renal parenchyma has not been demonstrated in neither of those cases. Pathohistological findings of our patient's kidney biopsy matches those seen in HSP nephritis [11], thus damage by direct infection is unlikely. During the course of his illness, the patient never had loose stools, maldigestion, nor any other symptom of infection with an intestinal parasite except abdominal pain, so the abdominal pain might be considered as the only symptom of s.stercoralis infection. Abdominal pain is one of the most common manifestations of HSP and about two-thirds of patients complain of this symptom [12], so if we attribute this symptom to HSP, the infection itself could be considered asymptomatic. Our patient complained of abdominal pain in spite of early introduction of corticosteroids, and the complete withdrawal of symptoms was achieved only after treatment with ivermectin, indicating the significance of strongyloidiasis for the development of HSP in the presented case. Corticosteroids provide for quick resolution of symptoms in HSP patients [3], however, patients with strongyloides infection receiving immunosuppressive drugs, and especially corticosteroids, may develop severe hyperinfection or disseminated infection syndrome [14].

Corticosteroids contribution to the propagation of the autoinfection process could not be ruled out in the presented patient. It also needs to be emphasized that the use of corticosteroids did not lead to clinical presentation of severe hyperinfection or disseminated infection syndrome, but it is probable that corticosteroid treatment created the conditions for s.stercoralis reproduction and increasing their number, which made possible microbiological detection of larvae in duodenal biopsy samples.

S. stercoralis can persist for more than 30 years without developing a clinically notable disease [15]. The risk for severe strongyloidiasis exists for all patients treated with corticosteroids that come in contact with this nematode, while the duration of corticosteroid use is highly variable (4 days to 20 years) [14], and is therefore not a major determinant. Having in mind the life cycle and the lack of symptoms in most infected people, we could not know when the presented patient became infected. Laboratory examinations of the presented patient demonstrated biochemical signs of renal damage during the first week of illness, and since strongyloidiasis clinical course could be with no symptoms, it is possible that pre-existing strongyloides infection could have caused nephritis, although there are no known mechanisms for parasitic infection to influence the early onset of nephritis in HSP. Some published reports demonstrate remission of nephrotic syndrome after strongyloides eradication [9,16,17], implying a causal link between strongyloidiasis and glomerulonephritis.

Conclusion

Strongyloidiasis is not a severe disease in immunocompetent people and its association with HSP has not been previously described. Resolution of clinical signs of HSP in our patient was achieved after treatment with ivermectin. The time course of the disease and present knowledge concerning the pathogenic mechanisms of HSP, suggest that s.stercoralis infection could have caused HSP in the presented patient, which was further complicated by nephritis.

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