MINI REVIEW

When children Rheumatoid Purpura complicated by nephropathy? What therapies?®

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The prevalence of renal involvement varies widely according to the pattern of referral and diagnostic criteria used; however because of its high prevalence, it is considered as a feature of the disease. Clinical expression of Henoch-Schönlein nephritis (HSN) ranges from transient, isolated microscopic haematuria to rapidly progressive glomerulonephritis and uraemia. The commonest renal manifestation is haematuria with variable degree of proteinuria. The long term outcome in HSN is uncertain though majority of children does not have serious renal disease.

Key Words: *Lymphatic fistula; Vein glue; Lymphatic fistula*

INTRODUCTION

Henoch Schonlein Purpura (HSP), was first recognized in 1801 by Heberden and first described as an association arthritis by Schonlein in 1837[1]. It is a systematic vasculitic disease, and mainly affects the small vessels of skin, joints, gastrointestinal tract, and kidneys. In some series, boys are affected more than girls [2]. The mean age of HSP patients is 6 years; 75% of the patients are under 8 years of age and 90% are less than 10 years of age [3,4]. Although many antigens, such as foods, infective agents, drugs, vaccinations, and insect bites have been reported to be related to HSP, the etiology of this disease remains unclear [5,6].

The pathogenesis also remains unknown. At present, HSP is regarded as an inflammation and immune-mediated disease. IgA and some proinflammatory cytokines have the pivotal role in the typical clinical characteristics involve the triad of palpable purpura, abdominal pain and arthritis. Progressive renal function impairment, bowel perforation, central nerve system involvement is rare [7]. The occurrence of purpura, which is nonthrombocytopenic, is the essential element for the diagnosis of HSP. The purpura is often located on some parts just as lower extremities and buttocks. HSP, in general, is considered benign and self-limited and the treatment is supportive [8]. The prevalence of renal involvement varies widely according to the pattern of referral and diagnostic criteria used; however because of its high prevalence, it is considered as a feature of the disease. Clinical expression of Henoch-Schönlein nephritis (HSN) ranges from transient, isolated microscopic haematuria to rapidly progressive glomerulonephritis and uraemia. The commonest renal manifestation is haematuria with variable degree of proteinuria. The long term outcome in HSN is uncertain though majority of children does not have serious renal disease [9,10].

CASE PRESENTATION

She is a three-year-old girl who was admitted for the management of rheumatoid purpura with edematous syndrome in the face and lower limbs. On admission, eutrophic child was found according to the WHO curves, nonpyretic at 37.2° with correct blood pressure in relation to age. The clinical examination found a puffy child with edem of the lower limbs, a purpura of vascular appearance made of raised petechial elements. The urinary strip found no blood or proteins. During the evolution and on day six 6 of his hospitalization, the child developed moderate abdominal pain with this times a hematuria with two crosses and a proteinuria with three crosses. Diuresis was preserved a renal assessment was carried out with Urea=0.26 g/creat=05.The blood pressure profile remained normal. The albumin level was 38 g/l. The 24-hour proteinuria assay found a rate of 3,111.36 mg/24 hours, or 190 mg/kg, and the Spot urine protein-creatinine ratio (UPCR) at 2,031.98 mg/mmol, which is very high, greater than 200 mg/mmol.

We therefore decided to perform a renal biopsy puncture in order to classify this nephroapthia that this reveals a mesomeric glomerulo nephritis with focal and segmental non-necrotizing crescent interesting 01 glomerulus/10 including 00 PAC, very good conservation of the interstitium and the tubes apart from 02 very moderate infiltrates of mononuclear inflammatory cells, one under the capsular and the other cortico-median. The arteriolar and arterial vascular network is normal. Immunofluorescence objectified granular mesangial positivity with IgA and .C3 and negativity of Glomeruli with IgG IgM and C1q and fibrinogen, this nephropathy was classified stage III according to ISKDC histologic classification of HSP nephritis. The child received three boluses of methyl prednisolone over three days with maintenance of corticosteroid therapy in decreasing doses. Clinical surveillance focused on oedematous syndrome and the urine strip and blood pressure, this experienced a slight increase and the child was put on converting enzyme inhibitors. Corticosteroid treatment was maintained for 16 months, at decreasing doses. The disappearance of proteinuria was noted from the third month, and the hematuria had persisted macroscopically until the 13th month then microspique; and was negated at the end of the 16th month. The antihypertensive treatment was maintained at weak dose for 13 months then stopped. We have a two-year follow-up in our patient, who is doing well, presents no clinical or biological relapse, his growth is normal [7].

DISCUSSION

HSP nephritis frequently presents as acute nephritic and/ or nephrotic syndrome, reflecting a rapidly developing pathophysiological mechanism [11]. The histologic lesions of HSP nephritis are categorized as ISKDC grades I, II, III, IV and V according to the presence and percentage of glomeruli with crescents [12]. The literature showed that there significantly more boys than girls, with a ratio of 1.40:1 [13], in our observation the patient was a girl. According to Trnka [14]. The mean age at presentation of nephritis was 87.4 ± 30.9 months, which was significantly higher than the age of those without nephritis, he was thirty six months for our patient. Renal involvement in the risk of HSPN increases with age; in particular, cases of children over 10 years of age were more likely to progress to HSPN. [15] There are worldwide data supporting the development Thirty-eight patients in a total of 319 children with renal involvement reported in three major series developed

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chronic renal failure and this accounted for 12% of their patients. A further 14 patients (7%) of the remaining 206 patients still had active disease. This made up 19% of patients with significant renal disease [16-18]. More than 25-30 years ago, correlation between the severity of clinical manifestation and kidney histology were derived [19]. There were several reports, which addressed the issue of risk factors for renal involvement in children. Older age, abdominal symptom, persisting purpura and low factor XIII activity 29 19 had been reported to be associated with a higher risk of renal involvement. Henoch-Schönlein [20,21]. Levy et al. [22], found that proteinuria exceeding 1 g/day was associated with less favorable outcome. The question we asked, ourselves for our patient was whether should receive treatment and if it was the case that it would be this treatment?

To answer this question, renal biopsy is seldom necessary for our patient, also for a purpose to assess severity of the disease. We have seen that our treatment did not use immunosuppressants, we only used a bolus of corticosteroids and oral corticosteroids in decreasing doses, that were maintained until clinical and biological remission; and ACE inhibitors. Our therapeutic attitude was identical to that published Jen et al. [23], one patient with hypertension >95th percentile at diagnosis required antihypertensive treatment. Corticosteroids were dont required antihypertensive treatment. Corticosteroids were administered orally in 43 patients (47%) for a median of 135 days (135–180), 31 received three methylprednisolone pulses followed by corticosteroids orally for a median of 150 days (120–210), and 18 (20%) none. No patient took steroids before renal biopsy; treatment was initiated at the time of biopsy. This multicenter study showed that 25% of patients with HPN class 2 develop chronic kidney disease with persistent proteinuria, which justifies long-term follow-up and treatment. For our patient the cutaneous involvement began to fade from the 6th day of evolution; his age is not in favor of a poor prognosis, only perhaps the mild abdominal attack in this case can be correlated with a risk of recurrence or long-term damage, as well as the results of the renal biopsy puncture which objectified a Grade3 damage according to ISKDC [9].

CONCLUSION

Henoch-Schönlein nephritis varies in severity. In this article we present the evidence showing of using corticosteroids on nephritic involvement. Therefore, HSP children who have the risk factors for renal involvement, especially severe kidney disease, should be closely observed. There is a need for large, well-designed clinical studies to determine the best regimens for the treatment of HSP nephritis in children. Prospective randomized controlled treatment studies are needed; research should be supported by online registry data coupled to all prospective clinical trials in patients with HSP nephritis.

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