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Henoch-Schönlein Purpura in Chinese Children and Adults

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SUMMARY **From 1987 to 1996, we retrospectively analyzed 84 children and 38 adults admitted to Chang Gung Memorial Hospital with the diagnosis of Henoch-Schönlein purpura (HSP). All of the adult patients had skin biopsy finding showing leukocytoclastic vasculitis. Male predominance was noted in children, but not in adults. Preceding infection was noted in 40.5% of children and 31.6% of the adults (P = 0.46). 8.3% of children and 13.2% of adults had medication intake at disease onset (P = 0.62). Children had more frequent abdominal pain than the adults (70.2% vs 28.9%, P < 0.01). Renal involvement was more common and severe in adults, manifested as more frequent hypertension (P < 0.05) and heavy proteinuria (P < 0.01). During acute attack, leukocytosis, thrombocytosis, elevation of serum C-reactive protein levels were more frequently observed in children, while elevated serum IgA and cryoglobulin levels were more common in adults. The overall prognosis was good in both age groups, although two adults developed end stage renal disease. Our study demonstrated the different expression of HSP in Chinese children and adults.**

Henoch-Schönlein purpura (HSP) is a vasculitis syndrome characterized by cutaneous purpura, arthritis, abdominal pain, and nephritis.[1,2] This illness occurs more frequently in childhood than in adults. The pediatric form of the disease has been studied extensively, and is generally considered a self-limited disorder.[3,4] However, there is relatively little information about the clinical features and disease course in adults,[5,6] as the definition of HSP in adults is more problematic than for children,[7] and the frequency of non-HSP vaculitides is much higher than in children.[8]

Though HSP in adults is in many ways similar to that seen in older children, there may be different clinical features between childhood and adult presentations. Only a few studies have focused on the comparison of clinical features between children and adults, yet yielding contradictory results.[9,10] In the present study, we compare the clinical and laboratory features of Chinese children and adults with HSP. All of our adult patients had skin biopsy-proven leukocytoclastic vasculitis. We clearly demonstrate the different expression of the same syndrome in Chinese children and adults.

MATERIALS AND METHODS

This was a retrospective study of 84 children (< 20 years, 54 males and 30 females) and 38 adults (> 20 years, 18 males and 20 females), with acute HSP admitted to our hospital from January 1987 to December 1996. A diagnosis of HSP in children required the presence of a purpuric rash concentrated on the buttocks and lower extremities, with or without one or more of the characteristic manifes-

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tations of the syndrome, such as hematuria, arthritis, or abdominal symptoms. The diagnosis of HSP in adults was made according to American College of Rheumatology (ACR) 1990 criteria for diagnosis of HSP,[7] which requires the presence of 2 or more of the following characteristics: age of onset 20 years or younger; palpable purpura; acute abdominal pain; and vessel wall granulocytes on biopsy. Three adults with rheumatoid arthritis, two with ankylosing spondylitis, and one with Sjögren syndrome were excluded. Patients with recurrent diseases were also excluded. A skin biopsy was performed in all of our adult patients with HSP showing leukocytoclastic vasculitis.

HSP nephritis was considered present if a patient had gross hematuria or microscopic hematuria(> 8 red blood cells per high power field) in conjunction with proteinuria of 30 mg/dl or greater. Mild nephritis was considered if the patient had proteinuria and/or hematuria not in nephrotic range. Severe nephritis was considered severe if the patients had heavy proteinuria (> 1 gm proteinuria/ day/m^2 of body surface area), hypertension, or increased plasma urea nitrogen and creatinine. Gastrointestinal (GI) bleeding was defined as the passage of grossly bloody stools or guaiac-positive stools.

Data from the medical records of these patients were reviewed and analyzed to compare the following aspects in the children and the adults: demographic and etiologic factors, clinical and laboratory features, and renal outcome. Group comparison was carried out by using Chi-square tests and Fisher exact tests when appropriate. A P value of < 0.05 was considered statistically significant.

RESULTS

The demographic and etiologic data

In children, HSP was more frequent in males and less frequent during the summer (Table 1). In adults, males and females were equally affected, without any seasonal variation. 40.5% of children and 31.6% of adults (P = 0.46) had preceding infection, mostly upper respiratory tract infections (URIs). One child had mycoplasma infection, while one adult had tuberculosis. 8.3% of children and 13.2% of adults (P = 0.62) were taking medicine at disease onset.

Clinical features

The clinical features were present at disease onset or observed during the course of admission (Tables 2 and 3). All of our

Table 1. Demographic data and etiologic factors in children and adults with Henoch-Schönlein purpura

	Children (n=84)	Adults (n=38)
Age, mean ± SD	6.6 ± 2.7	40.5 ± 17.4
Sex, M/F	54/30	18/20
Seasonal pattern; no. (%)		
Spring	25 (29.8)	12 (31.6)
Summer	9 (10.7)	12 (31.6)*
Fall	21 (25.0)	3 (3.6)
Winter	29 (34.5)	11 (28.9)
Antecedent events; no. (%)		
Unknown	50 (59.5)	26 (68.4)
Infection	34 (40.5)	12 (31.6)
Drugs at disease onset	7 (8.3)	5 (13.2)

*P< 0.05, compared to children

Table 2. Clinical manifestations of 84 children and 38 adults with Henoch-Schönlein purpura during the course of admission

	Children no. (%)	Adults no. (%)
Purpura	84(100.0)	38(100.0)
Abdominal pain	59(70.2)	11(28.9)**
Gastrointestinal bleeding	28(33.3)	11(28.9)
Arthritis	44(52.4)	22(57.9)
Nephritis	25(29.8)	20(52.6)*

*P < 0.05/**P <0.01, compared to children

Table 3. Renal involvement in 84 children and 38 adults with Henoch-Schönlein purpura

	Children no. (%)	Adults no. (%)
Mild	20 (23.8%)	9 (23.7%)
Severe		
Hypertension	2 (2.4%)	6 (15.8%)*
Elevation of BUN, Cr[1]	2 (2.4%)	5 (13.2%)
Heavy proteinuria[2]	5 (6.0%)	11 (28.9%)**

*P<0.05/ **P<0.01, compared to children
[1]BUN, blood urea nitrogen; Cr, creatinine
[2]Urine protein excretion > 1g/m^2/day

Table 4. Laboratory features in 84 children and 38 adults with Henoch-Schönlein purpura

	Children	Adults
Leukocytosis[1]	65/84 (77%)	13/38 (34%)**
Thrombocytosis[2]	44/84 (52%)	3/38 (7%)**
Increased CRP[3]	29/45 (64%)	8/32 (25%)**
Increased IgA	14/38 (37%)	19/25 (76%)**
Increased ASLO[4]	18/39 (46%)	8/21 (38%)
Cryoglobulinemia	2/12 (17%)	9/15 (60%)*
Low C_3 and/or C_4	1/40 (2%)	2/20 (10%)
Positive ANA[5]	1/14 (7%)	2/16 (12%)

*P<0.05/ **P<0.01, compared to children
[1]white blood cell count > 10^4/mm^3
[2]platelet count > 40 x 10^4/mm^3
[3]C-reactive protein > 10mg/l
[4]anti-streptolysin O > 200 Todd unit
[5]antinuclear antibody

patients, including children and adults, exhibited a characteristic rash. Abdominal pain was more frequent in children than in adults (70.2% *vs* 28.9 %, P < 0.001). GI bleeding was observed in 33% of children and 28.9% of adults (P = 0.79). Joint symptoms, usually oligoarthritis involving knees or ankles, were observed in 52.4% of children and 57.9% of adults (P = 0.71). Renal involvement was more frequent in adults than in children (52.6% *vs* 29.8%, P < 0.02). More adult patients presented with hypertension (P < 0.05) and heavy proteinuria (P < 0.01), compared to pediatric patients.

Laboratory features

White blood cell counts, and platelet counts were carried out on all pediatric and adult patients on admission, while other serological tests such as C-reactive protein (CRP), serum immunoglobulin A levels (IgA), anti-streptolysin O titer (ASLO), cryoglobulin, complements, and antinuclear antibody (ANA) were carried out only with some of the patients (Table 4). Leukocytosis, thrombocytosis, and increased serum CRP levels were more frequent in children than in adults (P < 0.01). Elevated serum IgA levels (P < 0.01) and cryoglobulinemia (P < 0.05) were more common in adults (P < 0.05). Most patients, children and adults alike, had normal C_3, C_4, and ANA when tested.

Treatment and outcome

Corticosteroid treatment was given to 62 (72.9%) children and 34 (88.9%) adults because of persistent skin lesions or severe abdominal pain, with good response in both groups. Three adult patients required additional cytotoxic drugs because of severe nephritis. Recurrences (reappearance of symptoms after the first attack subsided for at least 1 month) were noted in 30.9% of children and 26.3% of adults (P = 0.36). The overall prognosis was good for both children and adults: only eight children and five adults had persistent hematuria/proteinuria beyond 1 year (P = 0.78). Two of the five adults developed end-stage renal disease within 3 years.

DISCUSSION

In the present study, we sought to determine whether HSP vasculitis in adults had different clinical characteristics from those of children. Different criteria have

been proposed to define HSP in adults, with varying success.[7,8] There is, however, no "gold standard for the diagnosis of HSP at present. All of our adult patients presented with characteristic rash and positive skin biopsy results, thus fulfilling at least the two more essential criteria of the four proposed by ACR.[7] Patients with underlying disease, found in study population of previous reports,[5] were also excluded from this study.

We found that the peak incidence of HSP in children was in winter, while significantly more adult cases were found in summer. This might be explained by the different infectious diseases affecting children and adults. Preceding infections, mostly URIs 1-2 weeks before the illness, were equally found in children and adults. Although medications have been implicated as a frequent trigger of HSP in adults,[5] relatively few children and adults were taking medication at disease onset in this study. The etiologic factors remained unclear in the majority of patients.

Similar to Ilan, *et al.*[9] abdominal pain was more common in our pediatric patients. In contrast to those reported by Michel *et al.*[8] and Blanco *et al.*[10] that HSP in adults was associated with a significantly higher proportion of melena, the frequency of GI bleeding was comparable in children and adults in this series. All of our adult patients with abdominal pain had GI bleeding, while only about half of the children with abdominal pain did, suggesting that children were more sensitive to pain than adults. In agreement with Blanco *et al.*,[10] renal involvement was more common and severe in adult patients with HSP at initial presentation.

As for the laboratory features, children with acute HSP had more frequent leukocytosis, thrombocytosis, and elevations of serum CRP than adult patients did, indicating a more intense inflammatory response to HSP vasculitis in children. Elevated serum IgA levels, which may play a pathogenic role by formation of IgA immune deposits,[11,12] were more frequently observed in adult patients. We found higher incidence of cryoglobulinemia in adult patients (60%) as compared to 17% in children. Cryoglobulins have been shown to be composed of immune complexes with inflammatory properties,[13] occurring in various infections and autoimmune diseases with an immune complex pathogenesis, including HSP.[14] We may speculate that the different etiological agents of HSP between children and adults, such as variations in infecting organism, may result in stronger immune complex responses, and therefore, higher frequency of cryoglobulinemia observed in adult patients.

We found that the courses of renal disease in children and adults are similar, confirming previous reports.[9,10] The overall prognosis was good in both groups. Several reports, however, indicated that adult HSP nephritis is a potentially catastrophic disease, requiring long-term follow-up.[15,16] Two of our adult patients developed chronic renal failure, both presenting as acute nephrotic syndrome at disease onset. Further studies are needed to determine the prognosis of adult HSP nephritis.

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DATE	:	8-11 December 1998
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