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Analysis of Henoch-Schonlein Purpura in Adults and Children: Experience at King Abdulaziz University Hospital

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ABSTRACT. This is a retrospective study of all patients admitted to King Abdulaziz University Hospital between 1983 and 1999 with Henoch-Schonlein Purpura. Twenty-four patients (18 children and 6 adults with the ratio of 3:1) were analyzed. Seventy-one percent were Saudis and 29% non-Saudis. The mean age of the children was 5.9 years of age and 32 years of age in adults. Preceding upper respiratory tract infection was found in 50% of patients. Skin lesions were the most common presentation in both groups, followed by abdominal pain which occurred in children only, and joint pain in 5 (6%) children and 16 (7%) adults. Supportive treatment was used in the majority of children while all adults required steroid administration. Outcome was good in all patients except 1 adult who died of gastrointestinal bleeding and jejunal perforation. Results of this study were comparable with other reports. No significant relation between URTI and severity of skin involvement with the development of complications and mortality was found.

Keywords: Henoch-Schonlein purpura, Complications, Age, Saudi Arabia.

Introduction

Henoch-Schonlein Purpura (HSP) is a vasculitic disease characterized clinically by palpable purpura, abdominal pain, arthralgia and hematuria. The outcome is usually benign. Renal involvement may sometimes lead to acute or chronic renal failure. Upper respiratory tract infection (URTI) is the main triggering factor in children, while med-

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ications were implicated as a frequent triggering factor in adults[1]. Published series from Riyadh, Asir, Qatar, and Kuwait indicated were all on children[2-5] . However, no series were found published from the Jeddah area either on children and adults. The study retrospectively studied 24 cases of adults and children patients with HSP. The purpose of this work was to identify the precipitating factors, to document the presenting symptoms and signs, and to report the prognosis of the cases seen in King Abdulaziz University Hospital (KAUH).

Methods

The study retrospectively analyzed all patients admitted to KAUH, Jeddah (the western part of Saudi Arabia), with a diagnosis of HSP in the period from October 1983 to March 1999. Patients who fulfilled the American College of Rheumatology criteria were included. The patient's: age, sex, nationality, presentation, preceding use of medication or URTI, renal involvement (diagnosed by presence of hematuria, proteinurea, red cell cast, or renal biopsy), investigations like complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein, hepatitis serology, complications, and treatment were reported as well as recurrences and outcome.

Statistical Analysis: Analysis of data was conducted using the Statistical Package for Social Sciences (SPSS 7.5). A two-tailed student's t-test and Chi-square were used as appropriate. Results were considered significant if the P-value was less than 0.05.

Results

Sixty two records were analyzed. Thirty-eight cases were excluded as they didn't fulfill the American College of Rheumatology criteria and a total of 24 cases were analyzed, 18 children and 6 adults. All adult patients had skin biopsies showing Leukocytoclastic vasulitis with IgA deposition in the vessel wall. The overall children: adult ratio was 18:6 (3:1). Of these 17/24 (71%) were Saudi and 7/24 (29%) were non-Saudi (3 Palestinian, 2 Ethiopian, 2 Yemenese). Most of the cases, 20/24 (83%) presented between December and March. Table 1 shows comparison between adults and children. No significant relation was found between history of preceding URTI and renal involvement, intestinal involvement, development of cerebritis, scrotal swelling, or mortality. The observation of the development of a generalized skin rash was more in those with a history of preceding URTI, which was not statistically significant. Drug intake was documented only in 1 adult who gave a history of Acetyl Salicylic Acid intake.

Presentation: the most common presentation in both groups was a skin rash that was in the form of purpura in 94% and maculopapular eruption in 5.6% in children, while in adults it was 67% and 12% respectively, 17% of the adult patients had erythema of the skin and 4% had hemorrhagic bullae. No relation was found between the severity of skin involvement and involvement of other organs, or the development of complications, and mortality.

Renal involvement developed in 1 female child whose biopsy showed cresentic glomerulonephritis, and the patient progressed to chronic renal failure over a period of one year. Two adults had renal involvement, where their renal biopsies showed mesengioproliferative glomerulonephritis and focal proliferate glomerulonephritis. None of them progressed to renal failure over a follow-up period of 3 months and 1 year, respectively.

TABLE 1. Comparison between adults and children according to some variables.

Variables	Adults (N=6)	Children (N=18)	P-Value
Age (mean +/- SD)	32.17 +/-12.46	5.85 +/-3.35	-
Preceding URTI N(%)	3(50)	9(50)	-
Drugs Intake N(%)	1(17)	-	-
Sex (M:F)	1:5	1:1	0.15
Presentation			
Skin Rash	5(83)	14(78)	0.7
Abdominal Pain	-	3(17)	0.2
Joint Pain	1(17)	1(6)	0.3
Clinical Manifestation			
Skin Rash*	6(100)	18(100)	-
Lower Limbs' Rash	2(33)	11(61)	-
Generalized Rash	4(67)	7(39)	-
GIT*			
Abdominal Pain	6(100)	13(72)	0.15
Vomiting	2(33)	1(6)	0.07
Hemaemesis	1(17)	1(6)	0.72
Diarrhea	2(33)	3(17)	0.07
Hematochezia	2(33)	-	0.01
Joints	5(83)	13(72)	0.8
Lower Limb Arthritis	4(67)	11(61)	-
Generalized Arthritis	1(17)	2(11)	-
Renal Involvement	2(33)	1(6)	0.07
Fever	1(17)	9(50)	0.15
Scrotal Swelling	-	2(11)	0.3
Cerebritis	1(17)	1(6)	0.4
		-	

*URTI=Upper Respiratory Tract Infection, *GIT=Gastrointestinal Tract

Complications: Intestinal perforation developed in 2 adults, one of them had illial and the other had jejunal perforation.

Investigations: Elevated white cell count, ESR, and C-reactive protein were noticed more in children, 12/18 (67%), 5/18 (28%), 17/18 (94%) versus 2/6 (33%), 1/6 (17%), 2/6 (33%) in adults. Hepatitis B-serology was positive in 1 adult patient with normal liver function tests.

Treatment: Supportive measures of treatment in the form of simple analgesic and non-steroidal anti-inflammatory drugs (acetaminophine, aspirin, naproxin, and mefenamic acid) were the mode of treatment in the majority of children, 12/18 (67%) and 3/6 (50%) of adults. Steroids were used in all adults and in only 10/18 (56%) children. Prednisolone tablets in doses from 20-60 mg/day were given to adults, tapered over 2 weeks to 8 months. One adult required intravenous methylprednisolone for severe intestinal involvement for 3-weeks. In children, prednisolone was given to 2-children with scrotal swelling tapered over 6-weeks, while it was given for 1-year in 1 child with renal involvement. Immunoglobulin and immunosuppressive medications were used in 1 child with severe progressive renal involvement and in 1 adult with severe gastrointestinal involvement.

Recurrence and Outcome: While none of the adults had recurrences, 2/18 (11%) of children had recurrence of skin rash and arthritis during a follow-up period of 1-year. Generally, the outcome was good in both children and adults except for 1 adult who died of severe intestinal involvement and jejunal perforation.

Discussion

HSP is a vasculitic disease predominantly occurring in children, with a peak incidence at 5 years of age, but it does occur in adults[6]. It is characterized clinically by palpable purpura, abdominal pain, arthralgia, and hematuria. The American College of Rheumatology criteria for the identification of HSP is mainly clinical, relying on the presence of 2 or more of the following characteristics: age (20 years or younger), palpable purpura, acute abdominal pain, and granulocytic infiltration of arteriolar or venular walls with a sensitivity and specificity of 87%[7].

In our adult patients, the age criteria could not be used but the diagnosis of HSP was suggested by the other criteria. In addition, all of them had IgA deposition in the vessel walls of the dermis. Preceding upper respiratory tract infection (URTI) was common in our cases as in other studies[2-5]. But, it did not predict kidney involvement, as suggested by Trancrede-Bohin[6], or other organ involvement, or the development of complications or mortality. Also, the presence of bullous or necrotic cutaneous lesions did not predict other organ involvement, complications or mortality. These lesions were present in adults only, which is in agreement with previous observation[8].

When we compared our children series with published series from Riyadh, Asir, Qa-

tar, and Kuwait[2-5], we found that we have similar results in almost all aspects, although our series were the smallest in number. We cannot explain why we only have 24 cases in 16 years, although, we are a referral hospital in Jeddah. The differences we found were in the lower renal involvement and the male to female ratio that was equal in our study while it showed male predominance in the others. This may be due to the small number of cases in our study.

It is worth noting that in our children series is the development of scrotal swelling in 2 children; none of them required surgical treatment and were resolved with medical treatment only. This is a very rare manifestation and there is little mention of it in the textbooks. Lee *et al* described 7-cases of acute scrotum due to HSP over 20-years. In Korea, 2 children underwent operations and 5-children received conservative treatment only[9]. Evaluation of the cases with testicular scanning allowed proper management and avoided unnecessary exploratory surgery to role out testicular torsion[10].

Comparison of our children series with our adult series with HSP showed that preceding upper respiratory tract infections to be similar in both groups and that the cutaneous lesions were the main clinical manifestations in both groups. However, adults had a lower frequency of fever and a higher frequency of joint involvement at presentation. This was similar to the finding of Blanco *et al*[11]. There is a lack of information about the characteristics of HSP and its course in adults worldwide.

To the best of our knowledge, no adult series with HSP have been published in Saudi Arabia or in our area. Thus, more studies about adult HSP are needed.

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تحليل حالات مرض هينوخ شونلين عند الكباروالصغار في مستشفى جامعة الملك عبدالعزيز

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المستخلص. إن مرض هينوخ شونلين هو أحد أمراض الأوعية الذي يصيب الأطفال والكبار، وقد هدفت هذه الدراسة إلى معرفة العوامل المؤدية لظهور هذا المرض وتدوين أعراضه وعلاماته عند ظهور الشكوى، ولوصف صفات المرضى المصابين به وعرض نتائج حدوث هذا المرض في مستشفانا. كما نود معرفة العلاقة بين الإصابة بالتهاب الجهاز التنفسي العلوي وشدة ظهور الطفح الجلدي مع ظهور المضاعفات وإصابة الأعضاء الأخرى في الجسم واستخدامها كعامل تنبؤي لحدوثها. تم تجميع الحالات المنومة في المستشفى وتحليلها، وأظهرت النتائج وجود ٢٤ مريض (١٨ طفل و ٦ كبار) بنسبة ١:٣ وكان ٧١٪ منهم سعوديين و ٢٩٪ غير سعوديين. وكان متوسط العمر في الأطفال ٥,٨٥ سنة وفي الكبار ٣٢,١٧ سنة. خمسون في المائة من المرضى أصيب بالتهاب الجهاز التنفسي العلوي قبل ظهور المرض وواحدمن المرضى كان حاملاً لفيروس التهاب الكبدالوبائي والآخر تناول عقار حامض السلسيلات. وكان الطفح الجلدي أكثر الأعراض ظهورا وتبعته آلام البطن والتي حدثت في الأطفال فقط ثم آلام المفاصل في ٦,٥٪ من الأطفال و١٦,٧٪ من الكبار. واحتاج جميع الأطفال إلى العلاجات المساندة بينما احتاج جميع الكبار إلى استخدام الكورتيزون للعلاج، وكانت نتيجة العلاج جيدة في معظم الحالات إلا في حالة واحدة توفي فيها المصاب بسبب حدوث انثقاب في الأمعاء. من ذلك نستنتج أن نتائجنا تشبه النتائج الأخرى

المنشورة، ولم نجد أي علاقة بين وجود التهاب في الجهاز التنفسي قبل ظهور المرض ولا لشدة الطفح الجلدي مع حدوث مضاعفات في الأعضاء الأخرى. ولذا فمن الصعب التنبؤ بحدوث المضاعفات ونتائج العلاج في هؤلاء المرضى.