CASE REPORTS

A case series of Henoch–Schönleinpurpura in a street in Northwest of China

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ABSTRACT

Henoch-Schönleinpurpura (HSP), a self-limiting systematic vasculitic disease, occurs almost sporadically, and spatial-clustering series cases have rarely been reported. Herein, we report a case series of HSP that is strictly confined to a street of Zheng village in Xianyang City of Shaanxi Province, northwest of China.

Key Words: Henoch-Schönleinpurpura, Spatial-clustering, Environmental factors

1. INTRODUCTION

Henoch-Schönleinpurpura (HSP) is a self-limiting systematic vasculitic disease characterized by a cutaneous purpuric rash, as well as joint, abdominal and renal manifestations. HSP occurs at any age with a higher incidence in childhood[1,2] and the annual incidence is an estimated 3-26.7/100,000 for children.[3] Asian children which is estimated 33.86/100,000 in China[4] have a higher annual incidence than the white or black.[5] In addition, HSP occurs almost sporadically, and spatial-clustering series cases have rarely been reported. Herein, we report a case series of HSP that is strictly confined to a street.

Epidemiological investigation and deduction

In this report, all five cases from five families presented as shown in Figure 1, in a street of Zheng village in Wugong County of Shaanxi Province, northwest of China. Although there are several other street districts parallel with this street with similar density of youth population, none of them suffered from HSP. Furthermore, all five patients studied in one primary school, which is only 35 meters away in the southeast of the district, alongside about 270 primary school students, but none of them suffered from this disorder either, except these 5 cases.

An air-raid shelter cave had been built in 1950's right under the house 2 and 3 shown as Orange Square in Figure 1. The shelter cave was filled with plaster figures and straws which may release harmful gases like sulfur dioxide and methane. The cave had been filled in 1972 and the ground had been authorized for public buildings. As shown as Blue Square in Figure 2, the ground of the house 3 had been authorized as a medical station at that time; and there had been a dry well in the east-north corner for storage of medical waste as Green Square which may release harmful microbes and gases such as methane and hydrogen sulfide. In 1976, medi-

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cal station had moved out and this street was authorized to be personal housing till now. Based on all the results of field investigations, the association between the patients and the spatial distribution of the air-raid shelter cave as well as the medical waste well suggested that a component of the soil played an important role in HSP occurrence.

Samples of water and soil from the reservoir, village clinic and patients' houses were collected and analyzed according to the Standard Determination Methods for Drinking Water Quality (GB 5750-1985) and Soil Quality (GB15618-1995) and were proven qualified for drinking and living. All the environmental tests were performed at Xi'an Center for Disease Control and Prevention.

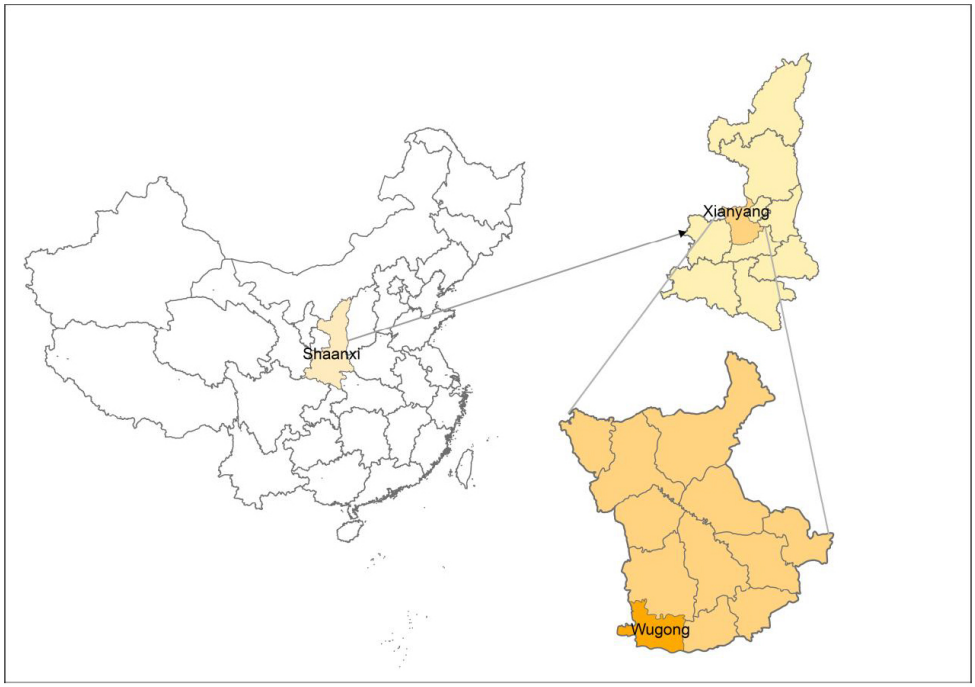

Figure 1. Geographic Location of HSP in Zheng village. It shows a map of Wugong County, Xianyang City, Shannxi Province, China, where all the patients in this case report acquired HSP in a street (34°13'28"N, 108°13'30"E)

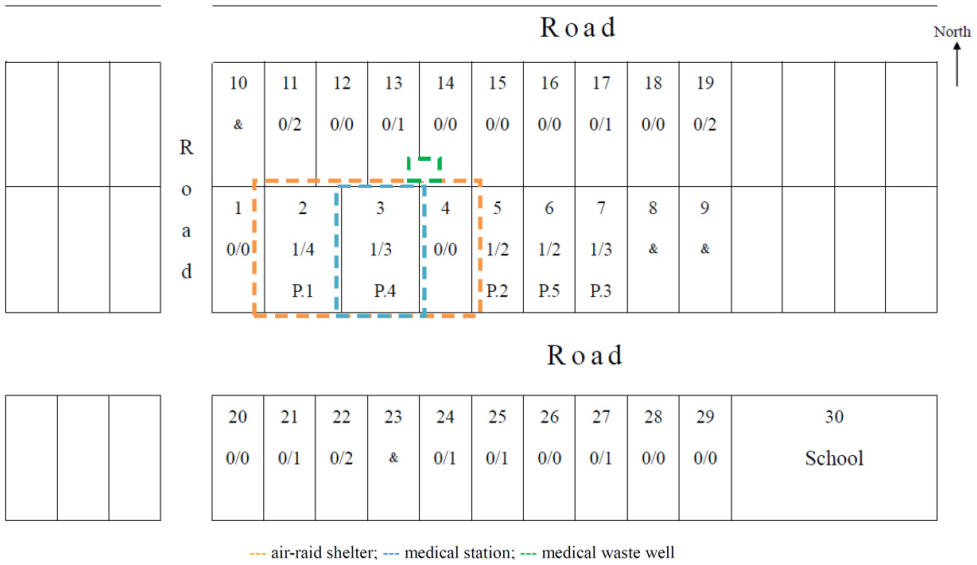

Figure 2. Simplified Schematic Diagram of Geographical Position of the five patients. n/N shows patients (n) of children under 15 years old now (N);& means empty house.

2. CASE REPORTS

As shown in Table 1, all five cases, live in one street and study in the same primary school. There are also 21 children who were younger than 15 years old lived paralleled with the street. Patients were all previously healthy without any allergic history to medication or food and didn't have any meditation before HSP occurrence. Their families were free of history of HSP. All patients or their guardians provided orally informed consent and agreed to participate in the study.

Table 1. Clinical findings of cases with Henoch-Schönleinpurpura

Case no.	House no.	Sex	Age at HSP onset	time of disease	Preceding infection	Symptoms	Diagnosis	Treatment	Outcome
1	2	M	8 years	February 2012	+	leg pain , diffused leg swelling, purpuric rash	HSP	glucocorticoid	recover
2	4	F	8 years	October 2012	-	purpuric rash	HSP	glucocorticoid	relapsed in December 2013
3	7	F	6 years	January 2014	-	purpuric rash, abdominal pain	HSP	dexamethasone	relapsed in November 2015
4	3	M	10 years	July 2014	-	leg pain, purpuric rash	Synovitis, HSP	loratadine, cefixime, dipyridamole and tablet thymosin	relapsed in December, 2015 and January, 2016
5	6	M	8 years	November 2014	NK	purpuric rash, leg pain , diffused articular swelling	HSP	Traditional Chinese medicine	Henoch–Schoölein purpura nephritis

+, presence; –, absence; NK, not know.

2.1 Case 1

The first case, an 8-year-old boy, lived in the house 2 in the street with his parents and three siblings. He was admitted to the outpatient department of Xi'an Children's Hospital in February 2012. About ten days prior to admission, he had experienced runny nose and had not received any treatment. Two days prior to admission, he had developed pain and swelling in his lower limbs. The next day, a sudden onset of purpuric rash, which was described as red and purple in color without pain or prurit, appeared on both of his ankles. He was clinically diagnosed with HSP, accepted empirical therapy with glucocorticoid for 2 months and recovered gradually without any relapse. His siblings, born in 2012, 2013 and 2014 separately, were free of this disease.

2.2 Case 2

The second case, an 8-year-old girl, lived in the house 4 from western side in the street with her parents and one sister. She was admitted to the outpatient department of Xi'an Children's Hospital in October 2012, with a two-day history of purpuric rash on her lower limbs. The rash was diffuse, symmetric, pruritic with no pain. She had no joint, abdominal or renal manifestations. The family members denied any upper respiratory tract symptoms before admission. Conventional treatment was initiated and lasted for one week in the local hospital. She recovered rapidly and relapsed in December 2013 because of a cold. But since she moved away from the street in 2014, she hasn't relapsed yet. Her sister born in 2008 was free of this disease.

2.3 Case 3

The third case, a 6-year-old girl, lived in the house 7 with her parents and two siblings. She was hospitalized in the Xi'an Children's Hospital in January 2014. Five days prior to admission, a diffuse painful rash appeared on her lower extremities. Two day prior to admission, a sudden onset of abdominal pain with vomiting occurred. There were no preceding upper respiratory tract symptoms or infections but a history of urticaria which is easy to relapse after catching a cold. The patient was clinically diagnosed with HSP and administrated with 10 mg dexamethasone/day. She recovered rapidly but relapsed with rash and abdominal pain in November 2015. Her siblings born in 2003 and 2012 were free of this disease.

2.4 Case 4

The forth case, a 10-year-old boy, lived in the house 3 with his parents and two siblings. He was admitted to the local hospital in July 2014, with a two-day history of leg pain. A tentative diagnosis of synovitis was made in local clinic, and pertinent treatments were performed for 10 days; however, his condition did not improve and sooner he developed a typical painful rash on her ankle. Finally HSP was clinically diagnosed and medications were applied, including loratadine, cefixime, dipyridamole and tablet thymosin. The patient recovered rapidly but relapsed in December, 2015 and January, 2016 because of colds. His siblings born in 2010 and 2013 were free of this disease.

2.5 Case 5

The fifth case, an 8-year-old boy, lived in the house 6 in the street with his parents and one sister. He was admitted to the local hospital in November 2014, with a one-day history of purpuric rash, leg pain and diffused articular swelling. He received Traditional Chinese Medicine treatments. Since then HSP relapsed several times because of colds and finally developed into Henoch–Schoöleinpurpura nephritis in October, 2015. His sister born in 2014 was free of this disease.

2.6 Medical record

We found medical record of patient 3 from Xi'an Children Hospital. She was admitted on January 3rd with a diffuse painful rash (see Figure 3) and abdominal pain. Physical examination results showed her temperature was 36.5°C, pulse 84/min, respiratory rate 22 breaths/min, manual blood pressure 100/65 mmHg, weight 21.0 kg. A diffuse, symmetric, purpuric rash with angioneurotic edema was present on the extremities. The rash swelled slightly over the surface of the skin and did not fade when pushed. Her abnormal pain was paroxysmal around the navel and could abate by its own.

Laboratory investigations results were as follows: hemoglobin 112g/L, white blood cell count 3.2×10^9/L, platelet count 294×10^9/L, neutrophils ratio 45.1%, lymphocyte ratio 42.2% and urine microalbumin 78.10 mg/L. Occult blood and glucose in the urine was weakly positive. Microscopic examination of the urinary sediment revealed 1-2 erythrocytes/hpf. Thrombin indicators were almost within normal ranges. Serum immunoglobulin at admission such as rheumatoid factor (RF), antinuclear antibody (ANA), antineutrophil cytoplasmic antibodies (ANCAs), and mycobacterium tuberculosis antibody were negative. She was diagnosed as having HSP and upper respiratory infection, and was administrated with 100 mg dexamethasone, 4 ml cimetidine, 0.2 g ribavirin per day for 8 days. She recovered gradually and was discharged in the ninth day.

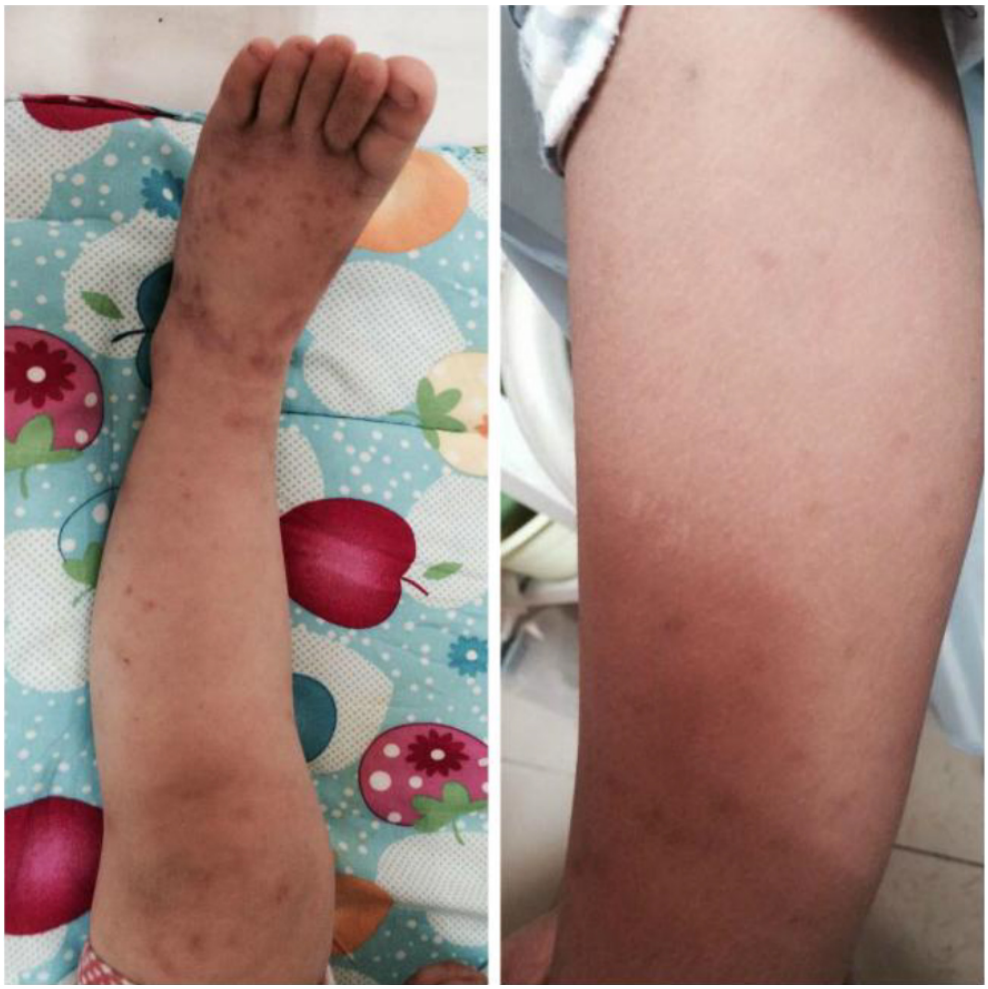

Figure 3. Clinical Presentation of HSP in Zheng village. Panels show images of purpuric rash on lower extremities of Patient 3

3. DISCUSSION

HSP is considered a benign and self-limiting disorder characterized by the presence of a palpable purpuric, joint manifestation, abdominal pain and nephritis, which typically occurs in childhood. The most common trigger events as previously reported were infection[6] which can be collectively indicated from the prominent autumn-winter peak in incidence rates and the frequent occurrence after an upper respiratory tract infection. And familial cluster cases indicated that genetic factors may also be responsible for this disease.[7] But in our report, only two patients had slight upper respiratory tract inflammation prior to the onset of first symptoms and all patients were from different families. As a result, it seems that normally reported reasons did not have a role in the etiology of HSP.

The special occurrence of HSP in our five patients may shed a different light on the etiology of this disease, just as the research in the past.[8, 9] We deduced this shelter cave and medical waste well may account for the outbreak of the disorder in this street, as all these hazardous substances can endanger human health, especially in children with higher susceptibility. However, since the results of potable water's and soil's routine quality monitoring in patients' houses showed that they were within quality requirements, an explicit etiologic role could not be determined and the pathogenesis could not be proved yet. It implied that circumscribed uncommon environmental factors in the district, either infectious or allergic, triggered the outbreak of the disorder. Patient 2 who moved to another distinct, recovered from HSP and didn't relapse, which further confirmed our hypothesis, sincerelapses often happen on children who had less commonly infections, had a longer duration of the first episode of palpable purpura and had abdominal pain and joint manifestations,[10] which was not suitable in this report.

Consequently, although no certain etiologic factor was finally determined in our report, we deduced that the outbreak of HSP had been triggered with an analogical environment background in the street related to air-raid shelter cave and the medical waste well. To our knowledge, this is the first and largest outbreak of HSP confined to a street in the world.

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CONFLICTS OF INTEREST DISCLOSURE

The authors declare that they have no competing interests.

REFERENCES

[1] Chen O, Zhu XB, Ren P, et al. Henoch Schonlein Purpura in children: clinical analysis of 120 cases. African Health Sciences. 2013 Mar; 13(1): 94-9. PMid:23658574. https://dx.doi.org/10.4314/ahs.v13i1.26

[2] Huang L, Liu A, Dai YW, et al. Clinical analysis of childhood anaphylactoid purpura:report of 760 cases. Chinese Journal of Dermatology. 2015 Jan; 48(1): 11-4.

[3] Piram M, Mahr A. Epidemiology of immunoglobulin A vasculitis (Henoch-Schonlein): current state of knowledge. Current Opinion in Rheumatology. 2013 Mar; 25(2): 171-8. PMid:23318735. https://doi.org/10.1097/BOR.0b013e32835d8e2a

[4] Li KL, Liu DW, Wu WD, et al. Study on the Incidence of Hospitalized Henoch-Schonlein Purpura Cases in 6 Cities in China, 2007-2009. Chinese Journal of Vaccines and Immunization. 2011 Feb; 17(2): 128-32.

[5] Gardner-Medwin JM. Incidence of Henoch-Schonlein purpura, Kawasaki disease, and rare vasculitides in children of different ethnic origins. Lancet. 2002 Oct; 360(9341): 1197-202. http://dx.doi.org/10.1016/S0140-6736(02)11279-7

[6] Albaramki J. Henoch-schonlein Purpura in children a fifteen-year experience at a tertlary hospital. The Lebanese Medical Journal. 2016 Jan; 64(1): 13-7.

[7] Motoyama O, Iitaka K. Familial cases of Henoch-Schonlein purpura in eight families. Pediatrics international: official journal of the Japan Pediatric Society. 2005 Dec; 47(6): 612-5. http://dx.doi.org/10.1111/j.1442-200x.2005.02138.x

[8] Levy-Khademi F, Korman SH, Amitai Y. Henoch-Schonlein purpura: simultaneous occurrence in two siblings. Pediatric Dermatology. 2000 Mar; 17(2): 139-40. PMid:10792806. https://doi.org/10.1046/j.1525-1470.2000.01733.x

[9] Cakir N, Pamuk ON, Donmez S. Henoch-Schonlein purpura in two brothers imprisoned in the same jail: presentation two months apart. Clinical and Experimental Rheumatology. 2004 Mar; 22(2): 235-7. PMid:15083895.

[10] Calvo-Río V, Hernández JL, Ortiz-Sanjuán F, et al. Relapses in patients with Henoch-Schönlein purpura: Analysis of 417 patients from a single center. Medicine (Baltimore). 2016 Jul; 95(28): e4217. http://dx.doi.org/10.1097/MD.0000000000004217