Case Report

Henoch–Schonlein purpura presenting sequentially as nodular rash, erythema nodosum, and palpable purpura

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ABSTRACT

We describe a 26-year-old woman who presented with a nodular rash on the elbows following an insect bite. Two days later, she developed erythema nodosum. Both these lesions were treated symptomatically. One week later, she had purpura, abdominal pain, hematuria, and arthralgias, following which steroids were administered. Her investigations revealed only microscopic hematuria that disappeared with therapy. This pattern of sequential appearance of rash and a nodular morphology are both unique features not previously reported.

Key words: Adult, erythema nodosum, Henoch–Schonlein purpura, nodular rash

INTRODUCTION

Henoch–Schonlein purpura (HSp) is an immune complex mediated small vessel vasculitis involving IgA deposition in the capillaries of the kidney, skin, and gastrointestinal tract.[1,2] Medications, vaccinations, infections, tumors, and insect bites have been mentioned as causes of HSp.[3] Purpura is the presenting symptom in 50% cases and is mandatory for diagnosis.[1] We report a rare case of HSp following an insect bite in a 26-year-old woman, who, in succession, had a nodular rash on the elbows, erythema nodosum, and palpable purpura.

CASE REPORT

A 26-year-old farm laborer presented to our outpatient department (OPD) with a nodular rash that had developed on both elbows [Figure 1a and b] following an insect bite while she sleeping. She had pruritus over the affected areas and her lower limbs as well, but without any rash [Figure 1c]. She was treated with antihistamines in the emergency department. Two days later, she presented to the OPD with complaints of tender and erythematous nodules on the legs [Figure 1d]. Her investigations revealed the following: Hemoglobin 10.9 g/dl, total leukocyte count 10.6 × 10^9/l with 76% neutrophilia, erythrocyte sedimentation rate 40 mm/h, urea 9.94 mmol/l, creatinine 44 μmol/l, platelets 284 × 10^9/l, and 40-50 RBCs/hpf on urinalysis without proteinuria. She was unwilling to be admitted and was, therefore, sent home and was prescribed acetaminophen and chlorpheniramine. She came back 1 week later with a purpuric rash on her lower limbs [Figure 1e], diffuse abdominal pain, and arthralgias of the knees, ankles, and elbows. Examination showed non-palpable purpura, pedal edema, tender peri-umbilical region, and an otherwise normal systemic examination. Her urea and creatinine were 12.07 mmol/l and 52.8 μmol/l, respectively. Microscopic hematuria persisted while the spot urinary protein was 23 mg/dl. Her electrocardiogram and liver function tests were normal, while ultrasonogram showed minimal free fluid in the pelvis. Her immunological profile was as follows: Rheumatoid factor negative, antistreptolysin O (ASO) titer <200 IU, antinuclear antibody 0.6 U (<1.0 U), complement C3 0.94 g/l, and complement C4 0.41 g/l. Testing for cytoplasmic-antineutrophil cytoplasmic antibodies (c-ANCA) could not be done since the patient refused to undergo any further investigations. She did not want a skin biopsy. She was initiated on methylprednisolone 8 mg daily, ranitidine, and hydroxyzine, and over the next 3 days, her signs gradually

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subsided. Her urine became bland prior to discharge, 7 days after admission.

DISCUSSION

Vasculitic disorders are classified according to whether the small, medium, or large arteries are involved.[4] HSp, urticarial vasculitis, cryoglobulinemic vasculitis, and cutaneous small vessel vasculitis are referred to as small vessel vasculitides.[5] Johann Lukas Schonlein and his pupil Edouard Heinrich Henoch contributed to the recognition of this entity.[3] Incidence of HSp is about 14/100,000, with the highest incidence in the autumn.[6] This is the commonest vasculitis in the pediatric age group.[2] The adult: Child ratio is 1:2-3 with a slight male preponderance.[3] Adults may or may not have more severe kidney disease depending upon the series studied.[3]

The capillaries, post-capillary venules, and non-muscular arterioles are inflamed.[5] Thirty percent of patients have a preceding upper respiratory tract infection.[1] Many viruses and bacteria have been implicated, but in the majority of cases, none have been found.[1] Infections

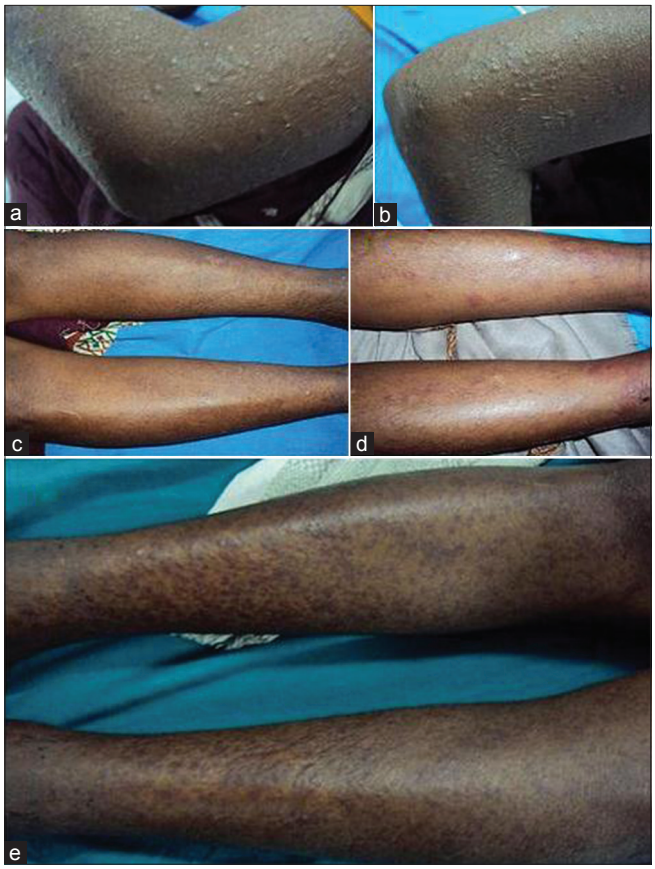

Figure 1: (a, b) Nodular rash around the left and right elbows developed few hours after the insect bite; (c) absence of any rash in the lower limbs during the first visit; (d) erythema nodosum bilaterally 48 h after the first visit; (e) purpuric rash in both lower limbs 1 week after the second visit

including streptococci, parvoviruses, Epstein–Barr virus, and vaccinations related to measles and enteric fever have been reported as etiologies.[6] The first case of HSp following insect bite was reported in 1954.[7] Abnormalities in HSp include dysregulation of interleukin (IL)-1β homoeostasis and homozygous and heterozygous mutations of Mediterranean fever (*MEFV*) gene that is involved in caspase-1 activation.[2] *MEFV* gene mutations are associated with severe nephritis.[2] Disease susceptibility arises from particular human leukocyte antigen (HLA) types which influence IgA function and/or clearance or those influencing immunomodulation.[2]

Features in descending order of frequency are purpura, arthritis, abdominal pain, nephritis, and gastrointestinal bleeding.[4] Also, 40-50% patients have renal involvement in the form of either hematuria or proteinuria.[1] Adults may have severe glomerulonephritis.[4] Nodules as a manifestation of vasculitis are more common with medium vessel vasculitides, whereas purpura, papules, vesicles, and urticaria are commoner with small vessel vasculitides.[5] HSp presents mostly with palpable purpura and papules.[5] Our patient had a nodular rash at onset, a feature which had not been previously described. Subcutaneous nodules can be seen in HSp, but are more common in other conditions such as rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), and rheumatic fever. [8] Such vasculitic nodules have been described only in children with HSp.[8] These subcutaneous nodules, as in RA, develop at pressure sites like the elbow, as was seen in our patient.[8] Erythema nodosum has been reported in only one instance of HSp and that too in a child.[9] This occurred 2 weeks prior to purpura and improved with clarithromycin that was administered for *Mycoplasma pneumoniae* infection.[9] Sequential development of three morphological patterns (erythema nodosum, erythema marginatum, and purpura) of rash was described in this child. This is only the second account of a similar case with three different patterns of rash occurring in succession and the first in which a nodular rash is a precursor of HSp. Tissue biopsies are useful when the presentation is atypical.[3] But in our case, the patient refused biopsies on two occasions. Vesicles and hemorrhagic bullae are occasionally seen.[3] Other manifestations involve the heart, liver, lung, nervous system, pancreas, and adrenals.[2] Systemic lupus, disseminated intravascular coagulation, and septicemia should then be considered in the differential diagnosis.[3] Based on the HSp EULAR/PRINTO/PRES Ankara 2008 classification, our patient had 3/4 criteria of the optional criteria.[2] Prognosis with hematuria without proteinuria is mostly benign.[10] The disease generally lasts for less than a month, with half of the patients having at least one recurrence, usually milder.[3] Our patient's urinary sediments became bland within 1 week of illness. About

45-80% of adults can have renal involvement, and the risk of progression to chronic kidney disease is 30%.[11]

In conclusion, we have described here a unique sequential pattern of rashes in HSp occurring after an insect bite that improved with steroids. Clinicians must be aware that HSp may present with several patterns of rashes in succession. Also, nodular rash and erythema nodosum could precede typical purpuric lesions in HSp.

REFERENCES

1. Marzano AV, Vezzoli P, Berti E. Skin involvement in cutaneous and systemic vasculitis. Autoimmun Rev 2013;12:467-76.
2. Smith G. Management of Henoch Schonlein purpura. Paediatr Child Health 2008;18:358-63.
3. Uppal SS, Hussain MA, Al-Raqum HA, Nampoory MR, Al-Saeid K, Al-Assousi A, *et al.* Henoch-Schonlein's purpura in adults versus children/adolescents: A comparative study. Clin Exp Rheumatol 2006;24:S26-30.
4. Khasnis A, Langford CA. Update on vasculitis. J Allergy Clin Immunol 2009;123:1226-36.
5. Xu LY, Esparza EM, Anadkat MJ, Crone KG, Brasington RD. Cutaneous manifestations of vasculitis. Sem Arthritis Rheum 2009;38:348-60.
6. Kraft DM, McKee D, Scott C. Henoch-Schonlein purpura: A review. Am Fam Physician 1998;58:405-8,11.
7. Sharan G, Anand RK, Sinha KP. Schonlein-Henoch syndrome after insect bite. Br Med J 1966;1:656.
8. Robson WL, Leung AK. Subcutaneous nodules in Henoch-Schonlein purpura. Pediatr Nephrol 2000;14:493-4.
9. Shimizu M, Hamaguchi Y, Matsushita T, Sakakibara Y, Yachie A. Sequentially appearing erythema nodosum, erythema multiforme and Henoch-Schonlein purpura in a patient with Mycoplasma pneumoniae infection: A case report. J Med Cas Rep 2012;6:398.
10. Sano H, Izumida M, Shimizu H, Ogawa Y. Risk factors of renal involvement and significant proteinuria in Henoch-Schonlein purpura. Eur J Pediatr 2002;161:196-201.
11. Pillebout E, Thervet E, Hill G, Alberti C, Vanhille P, Nochy D. Henoch-Schonlein Purpura in adults: Outcome and prognostic factors. J Am Soc Nephrol 2002;13:1271-8.

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