

## CAS CLINIQUE/CASE REPORT

### BULLOUS PRESENTATION OF HENOCHE-SCHÖNLEIN PURPURA IN AN ADULT

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**ABSTRACT :** Henoch-Schönlein purpura is a small vessel vasculitis occurring mainly in childhood and rarely in adulthood. Typical cutaneous eruption may begin as macular or urticarial erythematous lesions progressing to a palpable purpura. In adults, the disease has a propensity to be more severe and chronic and affects mainly the ankles and feet. Bullae, vesicles and ulcers are occasionally seen in this group. The prognosis depends on renal involvement, commonly seen in adults.

We report a new case of Henoch-Schönlein purpura in a 36-year-old man presenting with a bullous eruption followed by the appearance of abdominal pain and hematuria.

#### INTRODUCTION

Henoch-Schönlein purpura (HSP) is a small-vessel vasculitis that usually affects children and adolescents [1]. In adults, abdominal pain and fever have a lower frequency than in children. However, arthralgias and renal involvement are frequent and the latter can worsen the prognosis [2]. Cutaneous lesions begin with macular or urticarial erythematous eruption that typically evolve into palpable purpura. Bullae, vesicles and ulcers are occasionally seen in adults [3]. The diagnosis of HSP can rely on the clinical symptoms, physical examination and histology findings. It should be evoked in front of a classical triad of rash, gastrointestinal symptoms or hematuria, and arthritis [4]. We report a new case of HSP in a 36-year-old man, presenting with a bullous eruption and palpable purpura followed by the appearance of abdominal pain, joint swelling and hematuria.

#### CASE REPORT

A 36-year-old man without previous medical history, presented with palpable purpuric lesions and bullous

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**RÉSUMÉ :** Le purpura rhumatoïde ou syndrome de Henoch-Schönlein (SHS) est une vasculite des petits vaisseaux atteignant surtout l'enfant et rarement l'adulte. L'éruption cutanée débute par des lésions érythémateuses maculaires ou urticariennes évoluant vers un purpura palpable. Chez l'adulte, la maladie est plus sévère et tend à être chronique privilégiant les membres inférieurs, notamment les chevilles et les pieds. La forme bulleuse, les vésicules et les ulcérations sont occasionnellement observées. Le pronostic dépend de l'atteinte rénale qui paraît être plus fréquente dans la population adulte. Nous rapportons un nouveau cas de SHS chez un adulte de 36 ans sous forme d'éruption bulleuse suivie de l'apparition de douleur abdominale et d'hématurie.

eruptions evolving toward necrosis over the lower extremities (Fig. 1). He was previously seen by a physician and received empirical treatment with an antibiotic without any improvement. Ten days later, he presented a severe abdominal pain, unresponsive to treatment by non steroidal anti-inflammatory drugs (NSAIDs) with bilateral swelling of the ankles. There was no fever, no neurological or pulmonary symptoms nor additional abnormalities on physical examination. The patient had no history of recent infection, insect bites or drug intake before the onset of cutaneous lesions. The only altered laboratory tests consisted of a leukocytosis (white blood cell count of 16000 per mm<sup>3</sup>) with a left shift, thrombocytosis (490000 per mm<sup>3</sup>), microscopic hematuria and albuminuria (1+). Findings from the following tests were negative or within normal range : ESR, liver and renal function tests, ASO, PT, PTT, antinuclear antibodies, rheumatoid factor, anti-double-stranded DNA antibodies, c- and p-ANCA, antiphospholipid antibodies, cryoglobulinemia, C3, C4, serum IgA level and serologies for hepatitis B and C. Stool guaiac examination was negative. Chest X-ray was normal. Skin biopsy from a necrotic lesion showed vascular ectasia, extravasation of red blood cells, focal fibrinoid necrosis of certain capillaries in papillary dermis associated with a neutrophilic infiltrate and leukocytoclasia. Direct immunofluorescence analysis of the skin was negative. The diagnosis of HSP was made based on the clinical findings (palpable purpura, abdominal angina, joints swelling and hematuria) in correlation with the histology and laboratory

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FIGURE 1. Bullous purpura with a necrotic center over the right leg.

tests findings. Treatment with prednisone 60 mg per day led to resolution of abdominal pain and improvement of cutaneous lesions. A progressive withdrawal of corticosteroid was achieved within two months with complete healing of skin lesions.

#### DISCUSSION

HSP is an inflammatory disease of unknown etiology characterized by immune complexes deposits of IgA in veinules, capillaries and arterioles [4]. The disease is rare in adults and is more commonly found in men. In children, the antigen that induces HSP may be a viral pathogen while in adults the difference in severity and the more frequent occurrence in summer suggest a different pathogen. The signs of complement activation and deposition in the lesions indicate an immune-complex process [5]. The cutaneous eruption consists of erythematous papules typically followed by a palpable purpura, abdominal pain, arthritis or nephritis [4]. The rash tends to occur mainly over the feet and ankles and fades over one to four weeks but can be recurrent over months to years [3]. In severe cases, hemorrhagic, pur-

puric or necrotic lesions may occur [4]. Bullous eruption in HSP, as in our case, appears often in adults while it is very rare in children [6]. The cutaneous lesions, including bullous eruption, can result from a dysregulated humoral immunity [7]. Microscopic hematuria, melena, cutaneous ulcerations and sometimes the association with some drugs, predominate in adult population [8-10]. Abdominal pain of colicky type may occur and simulate an acute abdomen [11]. Pulmonary, cardiac, genital and neurological symptoms have also been described [5]. Nevertheless, the most serious sequela of HSP is renal involvement, occurring during the first three months of the disease and mostly revealed by hematuria. A monthly follow-up of urinalysis with measure of creatinine and uremia is required when hematuria is present [4]. The incidence of renal involvement in adults varies from 45 to 85% with a progression to renal failure in approximately 30% of cases [5, 12]. In adults, the disease tends to be more severe : 40% have persistent hematuria and 10% sustain chronic renal failure [5]. A recent infection, fever, extensive purpura over the trunk and biological markers of inflammation are predictive factors for renal involvement. However,

the presence of bullous lesions, cutaneous necrosis and the severity of histological findings (thrombosis, necrosis, depth of lesions) are not predictive of renal involvement [13].

Differential diagnoses of HSP include hypersensitivity vasculitis, vasculitis secondary to drugs or malignancies, cryoglobulinemia, polyarteritis nodosa, systemic lupus erythematosus, connective tissue diseases or infections [11]. The bullous eruption in our patient could be a source of diagnostic confusion. Hypersensitivity vasculitis shows a relatively chronic clinical course with presence of necrotizing vasculitis involving the whole dermis. In our case, the acute presentation, the absence of drug intake and infection prior to the onset of skin lesions and the leucocytoclastic vasculitis involving mainly the upper dermis are in favor of HSP, despite the negative direct immunofluorescence [14]. No focus of infection or malignancy nor laboratory tests compatible with cryoglobulinemia, systemic lupus erythematosus or connective tissue diseases were found. The presence of vasculitis involving capillaries rather than medium-sized vessels makes the diagnosis of polyarteritis nodosa less likely.

There is no specific laboratory test for HSP, a transitory elevation of serum IgA is observed in 50% of cases. Leukocytosis, eosinophilia, increased ESR and thrombocytosis may exist. Urinalysis may reveal the presence of hematuria [1, 4]. The direct immunofluorescence test from fresh lesions (done within 48 hours) shows deposits of IgA and C3 in the capillary walls. In older lesions, these immunoreactants may lack [15-16]. Moreover, Van Hale et al. found an 87% positivity of direct immunofluorescence in skin lesions of HSP, making the finding of negative test possible in 13% of cases [17]. The negative result in our patient may be explained by the delay before taking the skin biopsy (after 48 hours of onset of cutaneous lesions).

Kidney biopsy is nonspecific for HSP and is not necessary in the majority of cases. However, the biopsy may be done when the diagnosis is uncertain but is mainly indicated when renal involvement is persistent and/or worsening (appearance of renal failure, proteinuria superior to 2 g per 24 hours and arterial hypertension) [1, 5]. The normal renal function tests with the resolution of hematuria and albuminuria were not in favor of the decision of taking a kidney biopsy in our case.

There is no specific treatment for HSP. Bed rest and adequate hydration are indicated. NSAIDs may alleviate arthralgias while systemic corticosteroids are indicated to treat severe complaints or to prevent the progression of renal disease [2]. Dapsone has beneficial effects on cutaneous, gastrointestinal and articular manifestations in adults, especially those with chronic HSP [5]. Immunosuppressive treatment with cyclophosphamide, cyclosporine or azathioprine is sometimes necessary in life-threatening disease or when renal function is compromised [2, 5].

## CONCLUSION

HSP is the most frequent vasculitis in children but may occur in adults. It usually presents with palpable purpura which may progress into hemorrhagic, necrotic or less often into bullous eruption as in our case. The cutaneous lesions may be associated with abdominal, joints, renal, pulmonary or neurologic involvement [3, 5, 10]. The prognosis is excellent in the absence of persistent renal or other systemic involvement [11]. In our case, the skin lesions healed completely within two months of treatment with corticosteroids and regular follow-up of urinalysis showed a sustained complete resolution of hematuria and albuminuria over a period of three years.

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### فرقية فقاعية رثوية عند كهل

موجز : الفرقة الرثوية او متلازمة شونلين - هينوك التهاب العروق الصغيرة تصيب غالبا الاطفال ونادرا الكهول. يبدأ الطفح الجلدي بافات حمامية لطاخية او حاكة وتتطور الى فرقية مجسوسة. الافة عند الكهل شديدة وتتحول الى الازمان. التوضع الغالب يكون على الطرفين السفليين وخاصة الكاحل والقدم. الشكل الفقاعي والحويصلات والتقرحات تلاحظ اخيانا. يتوقف الانذار على الاصابة الكلوية وهي غالبا ما تصيب الكهول. نأتي على سيرة حالة جديدة للفرقية الرثوية عند كهل عمره ٣٦ عاما بشكل طفح فقاعي تتبعه الام بطنية وبيلة دموية.