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CLINICAL CASE

Bullous Henoch-Schönlein purpura. Case report

Púrpura de Schönlein-Henoch Buloso. Caso clínico

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Abstract

Henoch- Schönlein purpura (HSP) or IgA Vasculitis is the most common childhood vasculitis. The classic tetrad of signs and symptoms include palpable purpura, arthralgia, abdominal pain and renal disease. The occurrence of hemorrhagic bullae in children with HSP is rarely encountered. **Objective:** To report an unusual cutaneous manifestation of HSP in children. **Case report**: A 14-year-old girl complained about a 2-week painful bullous rash in both lower extremities and multiple arthralgias. There was no history of abdominal pain or urinary symptoms. In both lower extremities, there were numerous palpable purpura and hemmorrhagic bullae. In light of clinical findings, laboratory tests and skin biopsy are requested. The histopathology described intraepidermal blisters, acanthosis, spongiosis and perivascular dermal infiltrate. Direct immunofluorescence (IFD) (+) for IgA. The diagnosis of bullous HSP was made and treatment with endovenous corticosteroids was initiated. Three days after overlapping to oral corticosteroids, new ecchymotic lesions appeared in both legs. Due to the persistence of cutaneous involvement and negative control tests, azathioprine was associated obtaining a good response. **Conclusion:** Although bullous lesions in HSP does not add morbidity, it is often an alarming phenomenon with multiple differential diagnoses. The anti-inflamatory effect of corticoids is likely to be beneficial in the treatment of patients with severe cutaneous involvement through inhibition of proinflammatory transcription factors and decreasing the production of the metalloproteinases.

Keywords:
Schönlein Henoch,
bullous purpura,
vasculitis,
blisters

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Introduction

Henoch- Schönlein purpura (HSP) or IgA Vasculitis is the most common vasculitis in the pediatric age. It affects small vessels due to the storage of IgA in them[1]. It is presented as a tetrad, palpable purpura, arthralgia, abdominal pain and acute renal compromise[2], however, some of them might not appear. The diagnosis is based on the classification made by the European League Against Rheumatism and the Paediatric Rheumatology European Society in 2006. They proposed a diagnosis based on the presence of palpable purpura with one of the following symptoms: diffuse abdominal pain, skin biopsy with IgA deposits, arthritis or arthralgia, and/or renal compromise (hematuria and/or proteinuria)[3].

Characteristic cutaneous manifestations consist of palpable purpura (80 to 100% of the cases) and edema, which can be followed by urticarial exanthema or maculopapular exanthema. Palpable purpura presents a symmetric distribution and it is localized mainly in lower limbs and gluteus, but it can also affect the face, trunk, and upper limbs[1,5]. Despite being common in adults, cutaneous blisters are very uncommon in children, with an incidence rate lower than 2%[6].

The objective was to report an uncommon presentation form of HSP in children and to emphasize its relevance in the differential diagnosis of bullous dermatosis in the pediatric age.

Clinical case

Female patient, 14 years old, without relevant personal and family morbid history, consulted as outpatient due to a 2-week history of painful bullous lesions on both lower limbs associated with arthralgia, without compromise of the general health status, abdominal pain, urinary symptoms or previous discomforts. The treatment started with flucloxacillin, but there was no improvement. Thus, it was decided to hospitalize the patient on the 7th day.

The patient was evaluated by dermatology unit, which confirmed multiple blisters, some of them were confluent, forming wide bulla, erythematous plaques, some of them necrotic, and scabs in both lower limbs (figure 1). The arms had erythematous papules and small vesicles. The trunk, head, and mucosa did not have lesions. Due to a suspicion of autoimmune bullous disease, many laboratory tests were requested, complete blood count, biochemical, hepatic, and lipid profile, antistreptolysin O titer, complete urinalysis, thyroid tests, creatinine, complement C3 and C4, and antinuclear antibodies, in addition to a polymerase chain reaction to detect herpes 1 and 2, however, all of them had normal results. A biopsy of the skin was performed, which indicated intracorneal vesicle, epidermal acanthosis, and spongiosis associated with dermal infiltrate predominantly perivascular (figure 2). The direct immunofluorescence was positive for fine granular IgA deposits in vessels of the superficial plexus (figure 3), which confirmed bullous HSP.

The treatment started with intravenous hydrocortisone for three days, then, due to the good clinical course and the absence of systemic compromise, the patient was discharged with oral prednisone (1 mg/kg/day) associated with antihistamines. Due to the persistent cutaneous compromise after two months of treatment and negative follow up studies, it was decided to administer oral azathioprine and to start the tapering corticoids, which produced a good clinical response.

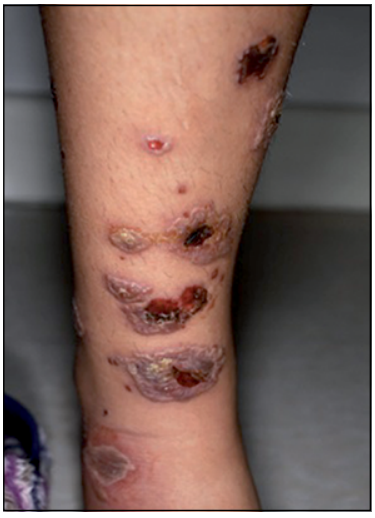

Figure 1. Necrotic blisters at the time of hospitalization.

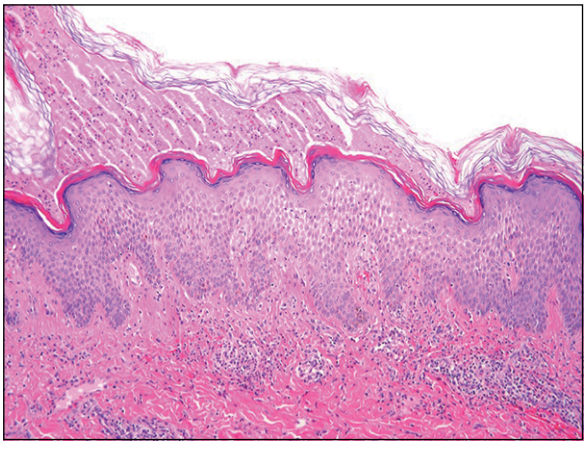

Figure 2. Skin biopsy, highlighting intracorneal vesicle with proteinaceous material, epidermis with acanthosis and spongiosis, and perivascular dermal infiltrate. Hematoxylin eosin 40x.

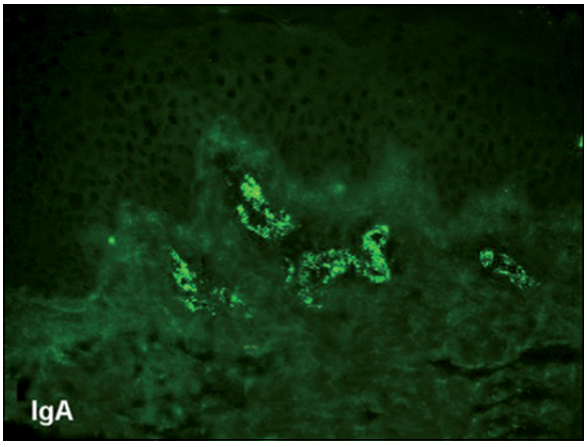

Figure 3. Positive direct immunofluorescence for IgA, with fine granular pattern in the superficial plexus vessels.

Discussion

The first reference to bullous HSP in the literature was made by Garland, 1985[7]. There is no evidence of an association of the HSP with a worse diagnosis, however, it is important to define the differential diagnosis with other bullous diseases in the childhood, such as erythema multiforme, bullous impetigo, dermatitis herpetiformis, staphylococcal scalded skin syndrome and linear IgA bullous dermatosis of children. Sometimes a cutaneous biopsy is necessary to determine the definitive diagnosis[8].

The etiology of HSP is unknown. Multiple bacterial and viral infectious agents have been associated with histories of respiratory tract affections. Other known triggers include drugs, food and bug bites. These elements would be the triggers of an immune response which leads to the formation of IgA-immune complexes, that which would be deposited in the blood vessel walls of susceptible people[9], triggering a local inflammatory response which leads to a leukocytoclastic vasculitis with small blood vessels necrosis[10].

Regarding the pathogenesis of blister formation, it seems like they are formed due to a metalloproteinase-9increase, which is an enzyme secreted by polymorphonuclear leukocytes in the dermic side of the dermo-epidermal junction. These enzymes break down components of the basement membrane, which leads to the formation of subepidermal blisters[11].

Although there is no consensus on the management of bullous lesions in the HSP, the anti-inflammatory effects of corticoids might be beneficial for the treatment of patients with acute HSP. It has been described that they might inhibit the proinflammatory transcription factor and reduces the concentration of nuclear factor kappa-B, matrix metalloproteinase -2 and -9[12,13]. It has also been demonstrated that topical corticoids are effective in the treatment of mild-moderate bullous HSP in adults, and in refractory cases, the use of immunosuppressors could be an alternative (such as azathioprine), although it is mainly indicated in cases of HSP with an acute renal compromise[14].

The HSP without renal compromise is usually auto-limited. The duration of the symptoms is variable, however, they disappear within the first eight weeks. In spite of this, recurrences are common, and they happen in 30-40% of the patients within the first year, but with lower intensity and duration[10].

Conclusion

The appearance of bullous lesions is an uncommon form of cutaneous presentation of HSP. Despite its worrying clinical symptoms, it does not imply a worse prognosis, thus it can be generally treated in the same conservative form as common HSP.

Ethical responsibilities

Human Beings and animals protection: Disclosure the authors state that the procedures were followed according to the Declaration of Helsinki and the World Medical Association regarding human experimentation developed for the medical community.

Data confidentiality: The authors state that they have followed the protocols of their Center and Local regulations on the publication of patient data.

Rights to privacy and informed consent: The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the correspondence author.

Financial Disclosure

Authors state that no economic support has been associated with the present study.

Conflicts of Interest

Authors declare no conflict of interest regarding the present study.

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