Reumatol Clin. 2010;**6(4)**:231–232

Reumatología Clínica

www.reumatologiaclinica.org

Images in clinical rheumatology

Schönlein-Henoch purpura: a case observation

Inmaculada Macías Fernández

Sección de Reumatología, Hospital Universitario, Puerta del Mar, Cádiz, Spain

ARTICLE INFO

Article history:
Received December 29, 2008
Accepted March 4, 2009

Case report

A 58 year old male with chronic obstructive lung disease came to the clinic due to confluent purpuric lesions which tended to ulcerate, which began during the days prior on the lower limbs; they increased in number and extended to the buttocks, trunk, upper limbs and face an were accompanied by joint pain of the hands and feet. No other gastrointestinal problems, fever or clinical data suggesting connective tissue disease existed. Upon examination, the patient presented pitting edema of the lower limbs and a universally distributed palpable purpura which did not affect mucosal tissue, with ulcers due to the loss of epidermis on the lower limbs (Figure 1, Figure 2).

Laboratory analysis showed: normal urea and creatinine; an erythrocyte sedimentation rate of 95 mm/h; C reactive protein of 5.8 mg/dl; microhematuria; 24 h urine proteins 2,859 mg en orina de 24 h; normal complement and elevated IgA. Autoantibodies were negative. Serology for HBV was compatible with a chronic infection and the viral load was negative. Rheumatoid factor was positive at a low titer and was interpreted as secondary to HBV infection. Skin biopsy was compatible with purpuric leukocytoclastic vasculitis, with an absence of immunofluorescnet deposits. Renal biopsy showed non-hyaline glomeruli and no half moons, with a mild increase in the mesangial matrix and some areas of cellularity, in addition to granular IgA and C3 deposits, detected through immunofluorescence, in the mesangium.

Suspecting Schönlein-Henoch purpura (HSP), oral prednisone was started (1 mg/kg/day for 2 weeks, reduced at a rate of 2.5 mg every 14 days, reaching 5 mg per day); losartan, atorvastatine and weekly alendronate plus cholecalcipherol. After one year, the patient has not developed new symptoms and is maintained with low dose steroids, and is being treated with low dose steroid, has good renal function, proteinuria under 500mg/dl and the HBV viral load is still negative.

Discussion

HSP is vasculitis manifested as non thrombocytopenic purpura, arthritis/joint pain, abdominal pain and, in a variable proportion, nephropathy.[1] It is histologically characterized by leukocytoclastic vasculitis with IgA deposits in small caliber vessels of the affected organs (skin, kidneys and intestine[1]). Nephritis, which appears in 20%-100% of cases, is the main prognostic factor,[2] although there are others such as: the greater the age, the worse the prognosis and the greater the risk for nephritis, persistence of nephritic syndrome and extension of the IgA deposits to the capillary walls. In 90% there is only microhematuria. Renal affection is dynamic and may evolve to chronic renal failure in 2%-5% even when there are no extrarenal manifestations.[3]

Skin lesions are treated only with rest, although sometimes steroids are employed when they are extensive or chronic. Arthritis

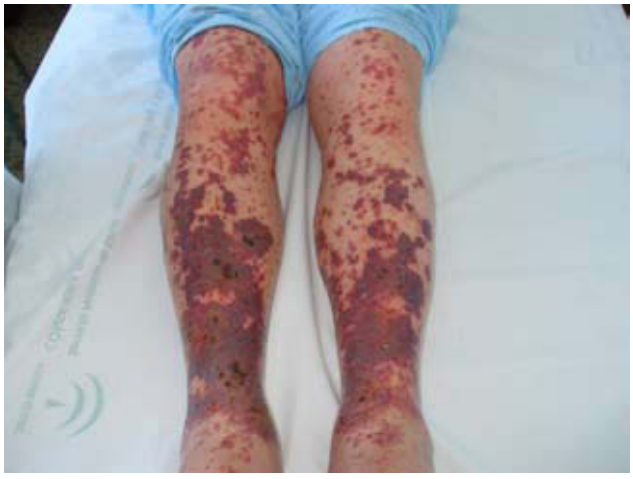

Figure 1. Red purpuric lesions which are confluent, petechial and tend to ulcer.

E-mail address: imaciasfdez@hotmail.com

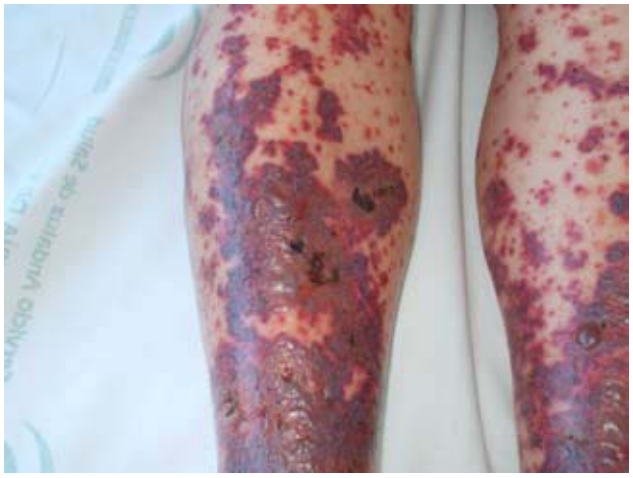

Figure 2. Extense and confluent areas with some ulcers due to epidermal loss.

responds to non-steroidal anti-inflammatory drugs or low dose steroids. A protective effect of steroids over the development of nephropathy has been suggested,[4] and its use seem reasonable in digestive tract affection and persistent skin lesiones (>2 months), in order to avoid acute complications (abdominal pain and surgery).[5] As immunosuppressants, immunoglobulins, cyclophosphamide, azathioprine and mycophenolate mofetil have been employed, but until there are controlled prospective studies, treatment of renal and/or intestinal HSP will continue to be controversial.

Disclosures

The authors have no disclosures to make.

References

1. Lie JT. Members and Consultants of the American College of Rheumatology Subcommittee on classification of vasculitis. Illustrated histopathologic classification criteria for selected vasculitis syndromes. Arthritis Rheum. 1990;33:1074-87.
2. Blanco R, Martínez Taboada VM, Rodríguez Valverde V, García-Fuentes M, Gonzalez-Gay MA. Henoch-Schönlein purpura in adulthood and in childhood: two differents expressions of the same syndrome. Arthritis Rheum. 1997;40:859-64.
3. Goldstein AR, White RHR, Akuse R, Chantler C. Long-term follow-up of childhood Henoch-Schönlein nephritis. Lancet. 1992;339:280-2.
4. Kawasaki Y, Suzuki J, Nozawa R, Suzuki S, Suzuki H. Efficacy of methylprednisilone and urokinase pulse therapy for severe Henoch-Schönlein nephritis. Pediatrics. 2003;111:785-9.
5. Weiss PF, Feinstein JA, Luan X, Burnham JM, Feudtner C. Effects of corticosteroid on Henoch-Schönlein Purpura: a systematic review. Pediatrics. 2007;120:1079-87.