Vasculitis IgA que afecta a cuatro sistemas

IgA Vasculitis involving four systems

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ABSTRACT

Henoch-Schonlein Purpura (HSP) is a systemic vasculitis now called IgA vasculitis, that infrequently occurs in adults. While rheumatological and gastrointestinal complications are common, lung involvement is a rare complication. We report the case of a 70 year-old female that presented to the hospital with purpuric lesions on her legs and ileitis. During her hospital stay she developed respiratory distress, and the computed tomography (CT scan) showed evidence of Diffuse Alveolar Hemorrhage (DAH). She also had renal dysfunction, and the renal biopsy confirmed the presence of leukocytoclastic vasculitis on histology, consistent with HSP. The patient was initiated on corticosteroid therapy and cyclophosphamide, in addition to supportive management, leading to the resolution of her symptoms.

This case is unique, as it presents a rare complication of HSP, with pulmonary disease, causing DAH. Furthermore, adult onset HSP is also an uncommon occurrence.

It is very important to recognize DAH early in HSP, as it holds a high mortality rate.

Keywords: Henoch–Schönlein purpura; Adult; Vasculitis; Corticosteroid therapy; Diffuse Alveolar Hemorrhage.

Palabras clave: Púrpura de Henoch–Schönlein; Adulto; Vasculitis; Terapia con corticosteroides; Hemorragia alveolar difusa.

INTRODUCTION

Henoch-Schönlein purpura (HSP) is an acute immunoglobulin A (IgA)–mediated disorder characterized by a generalized vasculitis involving the small vessels of the skin, the gastrointestinal tract, the kidneys, the joints, and, rarely, the lungs and the central nervous system. HSP has been renamed IgA vasculitis (IgAV), but the eponymic name remains in wide use.[1] HSP is one of the most common vasculitis in childhood, with an incidence of 10–20 cases per 100,000 children. It most often occurs between the ages of 3 and 15 years old. The prevalence of adult HSP is unknown, and its annual incidence is 1 in 1 million. This disease also seems to occur more frequently in the male gender. While the exact cause of HSP is unknown, various infections and chemical triggers have been associated with it. Studies show genetic, immunological and environmental factors that may play a role in the pathogenesis of the disease.[2,3]

A hallmark of Henoch–Schönlein purpura is a nonthrombocytopenic purpura, most commonly on the lower limbs. Histopathologically, Henoch–Schönlein purpura is characterized by leukocytoclastic vasculitis associated with deposition of immunoglobulin A immune complexes in vessel walls.[3]

HSP usually involves the kidneys, joints, gastrointestinal system and skin. Severe lung involvement, such as pulmonary hemorrhage, is very rare.[3,4]

The long term prognosis is heavily dependent on the presence and severity of nephritis. Studies with prolonged follow-up show that up to one third of adult patients reach end stage renal failure.[2-5]

We present a rare case of HSP with alveolar hemorrhage.

CLINICAL CASE

We present the case of a 70 years old caucasian female, without significant past medical history. About 13 days before hospitalization, a generalized cutaneous erythema erupted. She was seen at her local Health Center and prescribed bilastine. About 3 days later, she went to the Emergency Department (ED) with nausea and vomiting. Acute gastroenteritis was assumed, and she was treated with metoclopramide and pantoprazole at discharge. Another 3 days went by, and the patient's condition persisted, so, an analytical evaluation was carried out. The blood test showed no significant alterations, but she did present haematoproteinuria in the urinary sediment (US). Acute cystitis was then assumed, she was prescribed ciprofloxacin. Again, she maintained nausea and vomiting, and concomitantly initiated abdominal pain in the upper quadrants. The purpuric lesions began to scatter (Image 1-2), so she resorted again to the ED. The exames carried out showed that she maintained US with hematoproteinuria; analytically she now presented leukocytosis and C-reactive protein (CRP) of 30 mg/dl. Abdominal and pelvic CT-scans suggested ileitis. She was then admitted as an inpatient for further investigation on suspicion of vasculitis.

From the follow-up exams carried, stood-out a proteinuria of 3.2 g/24h. During her hospitalization she presented clinical deterioration, with partial respiratory failure. Chest CT-scan was performed, and revealed evidence of diffuse alveolar hemorrhage (Image 3).

She was then transferred to the Nephrology Department to perform a renal biopsy. The results were compatible with crescentic IgA vasculitis (Image 4-5). The diagnosis of IgA vasculitis was then assumed. Corticosteroid therapy was initiated, first with pulse IV methylprednisolone (500 mg to 1 g daily for three days), followed by oral prednisone (60 mg daily). By multiorgan involvement and severity of clinical case we

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Cómo citar este artículo: Diogo J, Coelho C, Lencastre Monteiro R, Ghiletchi Â
IgA Vasculitis involving four systems. Galicia Clin 2022; 83-4: 48-50
Recibido: 19/08/2021 ; Aceptado: 11/10/2021 // https://doi.org/10.22546/67/2633

Image 1

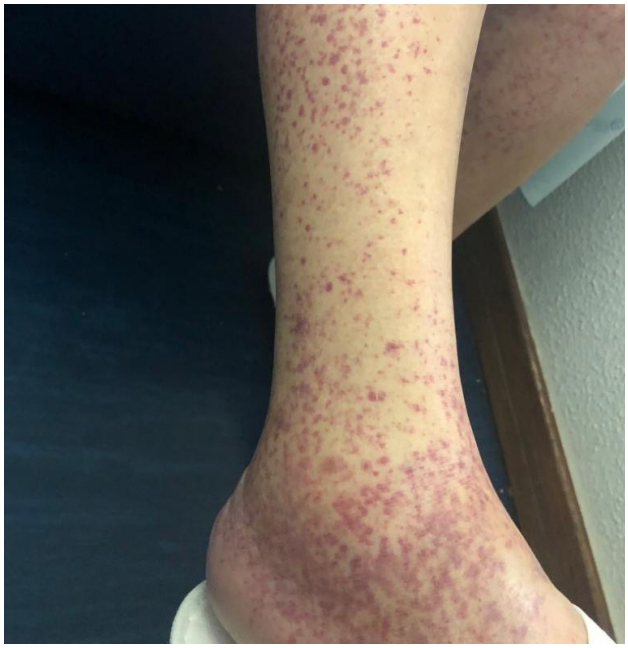


Image 2

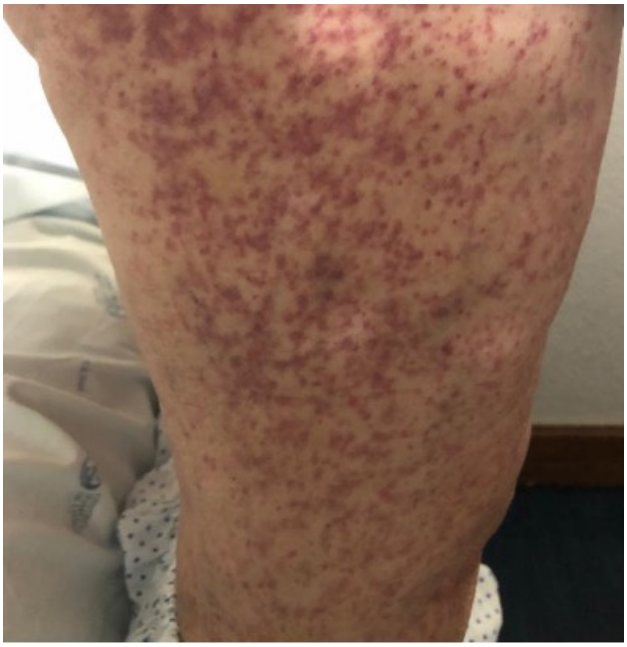
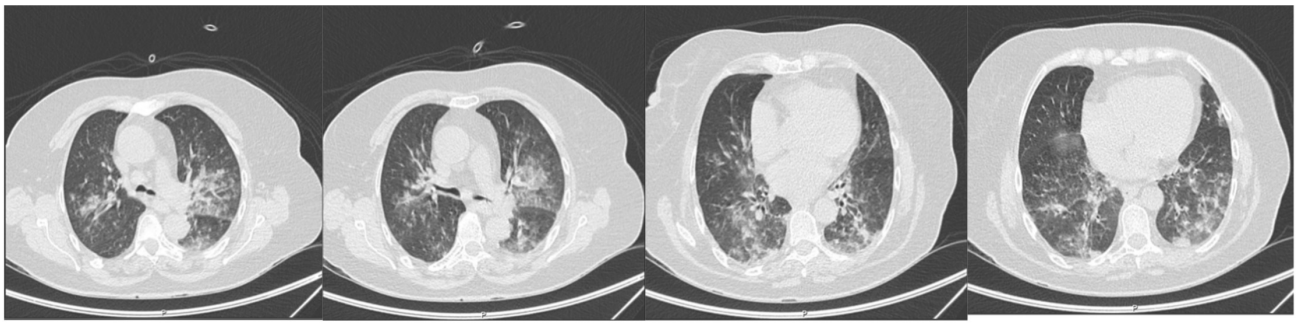


Image 3



also decided to treat with cyclophosphamide in addition to course of glucocorticoids. A clear improvement of her clinical status, with total regression of the purpuric lesion and normalization of kidney function was observed (Table 1). She also performed a bronchoscopy, thoracic, abdominal and pelvic CT scan and endoscopic exams, all excluding evidence of an occult cancer.

Three months later, on appointment she performed a pelvic and abdominal CT scan, also with regression of ileitis.

DISCUSSION

IgA vasculitis is a systemic vasculitis that infrequently occurs in adults. While rheumatological and gastrointestinal complications are common, lung involvement is a rare complication. The occurrence of DAH in HSP is rare, the reported prevalence ranging from 0.8% to 5%. The onset of DAH occurs variably after the diagnosis of HSP, from 2 days to 18 years. Hemoptysis (75%), a decrease in hemoglobin (74%), and chest infiltrates (94%) are the most common clinical findings. DAH is frequently severe, and 50% of the patients require mechanical ventilation.[6]

The clinical case presented unequivocally demonstrates the severity that HSP can have in adult age, unlike in the pediatric population, where this pathology is usually self-limited and has a favorable long-term prognosis. Usually, in adults, joint symptoms are more prevalent. Renal involvement is also more frequent and more severe in adults, strongly compromising the course of the disease. Concomitant pulmonary involvement is rare in ANCA-negative vasculitis but possible.

The difficulty in diagnosing this systemic vasculitis in adults is related to the fact that the symptoms of this clinical condition do not manifest themselves simultaneously, and often complaints such as arthralgia or abdominal pain precede the cutaneous manifestations. In this specific case the CT scan shows an ileitis. This is more frequent than lung involvement and represented the symptom that with the purpuric lesions makes the suspicion. That makes the decision of hospitalization.

When purpura is the only manifestation of HSP, the disease is usually self-limiting, and only conservative treatment is recommended.[3] On the other hand, when there is evidence of organ dysfunction, particularly common in adults, corticosteroid therapy is recommended. In cases of severe or relapsing disease, treatment with immunosup-

Image 4

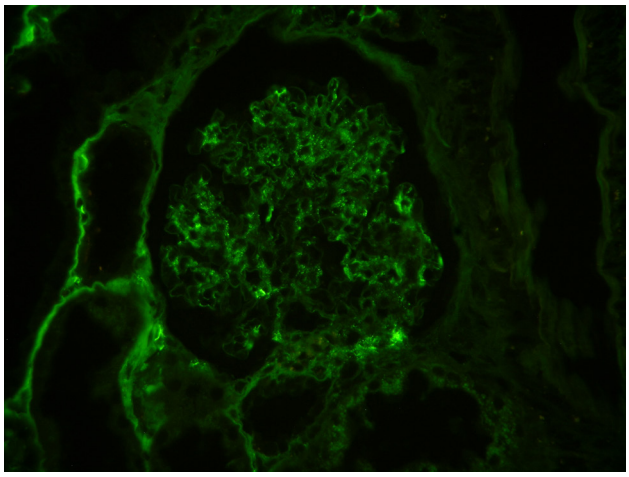


Image 5

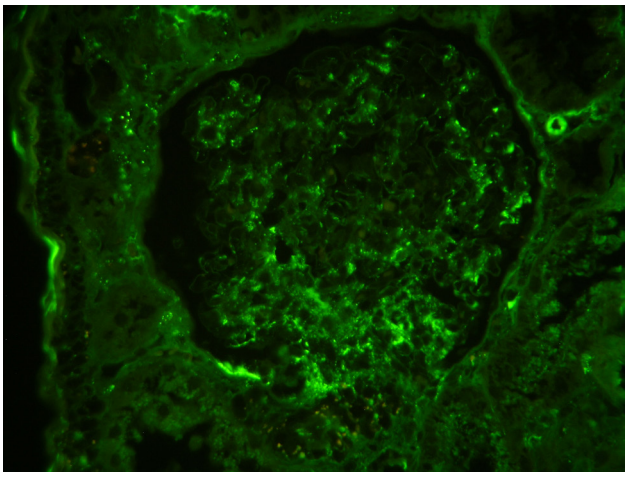


Table 1. Evolution of renal function during hospitalization

	Day 1	Day 4	Day 6	Day 7	Day 8	Day 9	Day 10	Day 11	Day 15	Day 30	After 3 months	After 1 year	After 2 years
Creatinine (mg/dl)	0,99	0,94	1,40	1,61	2,36	2,57	2,71	2,53	1,74	1,86	1,65	1,59	1,11

pressive agents, like cyclophosphamide, rituximab or MMF is recommended. Some authors prefer to give cyclophosphamide, based upon its effectiveness in the treatment of other forms of rapidly progressive crescentic glomerulonephritis.[1]

Vasculitis is associated with cancer, with an incidence of ~2–5%, the majority of cases related to haematological malignancies. The onset of vasculitis may appear before, during, or after the cancer diagnosis. HSP is more commonly associated with solid tumours than with haematological malignancies.[2-4] The gastrointestinal tract, respiratory organs and urinary tract are the most affected organs. These patients are mostly male, approximately 60 years of age, and screening for cancer in this subgroup could be indicated in the case of unexplained development of HSP.[3-5]

CONCLUSION

IgA vasculitis is a multisystemic heterogeneous disease. Histopathological examination of the skin and renal biopsy confirms the presence of leukocytoclastic vasculitis with IgA deposits. In renal involvement the presence of hematuria is possible, so it is important to analyze the urinary sediment.

Renal involvement, with concomitant pulmonary involvement, is rare in ANCA-negative vasculitis, but possible.

Adults have worse prognosis, and the development of hematuria may infer the appearance of serious complications, such as nephritic and nephrotic syndrome.

Malignancy is a possibility in the adult with PHS and therefore must be excluded. These patients should have regular follow-up for an extended period, given the possibility of long-term complications.

CONFLICTS OF INTEREST
The authors declare that there is no conflict of interest in this work.

FUNDING SOURCES
The authors declare that there were no external sources of study for the performance of this article.

RIGHT TO PRIVACY AND INFORMED CONSENT
The authors declare that no data that allows identification of the patient appears in this article.

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