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## Case Report

# IgA vasculitis as a presentation of human immunodeficiency virus infection<sup>☆</sup>

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## ABSTRACT

IgA vasculitis is a small-vessel vasculitis mediated by immune complexes. In clinical terms, it is characterised by palpable purpura in the lower limbs, joint involvement in the form of arthralgia or arthritis, and gastrointestinal and renal involvement (this will mark a poorer prognosis in adults). Infectious processes, mainly in the upper respiratory tract, are frequently found to be triggers. On the other hand, human immunodeficiency virus (HIV) causes immune dysfunction, which triggers hypergammaglobulinemia and can trigger autoimmune disorders. At times, this can affect the vascular endothelium, giving rise to vasculitic manifestations, although there are few reports in the literature of its role in the presentation of HIV.

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## Vasculitis IgA como forma de presentación de la infección por el virus de inmunodeficiencia humana

## RESUMEN

La vasculitis IgA es una vasculitis de pequeño vaso mediada por inmunocomplejos. Clínicamente se caracteriza por la púrpura palpable en miembros inferiores, la afectación articular en forma de artralgias o artritis y la afectación gastrointestinal y renal (esta última marcará el mal pronóstico en adultos). Es frecuente encontrar procesos infecciosos como desencadenantes, principalmente de vías respiratorias altas. Por otro lado, el VIH causa una disfunción inmunitaria que desencadena una hipergammaglobulinemia y puede desencadenar alteraciones autoinmunes. En ocasiones este efecto se realiza sobre el endotelio vascular dando lugar a cuadros vasculíticos, aunque como forma de inicio los casos descritos en la literatura son escasos.

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### Palabras clave:

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## Introduction

IgA vasculitis (IgAV), commonly called Schönlein-Henoch purpura, is a small-vessel leukocytoclastic vasculitis not associated with anticytoplasmic antibodies of the neutrophil antibodies, and is char-

acterised by the presence of immunocomplex deposits which predominantly contain IgA-1.<sup>1,2</sup> Although it may present at any age, this vasculitis usually presents in children (between the ages of 2 and 10<sup>3</sup>), and is characterised by the standard triad of palpable purpura, abdominal pain and arthralgias. Prognosis is generally favourable.<sup>1</sup> The IgAV are usually preceded by upper respiratory tract infections and exposure to drugs,<sup>4</sup> with cases described as being triggered by viruses, such as Epstein-Barr, cytomegalovirus or human B19<sup>3,5</sup> parvovirus. Cases of IgAV which are secondary to HIV in adults are exceptional, and we therefore believe it is of interest to present a new case.

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## Clinical case history

A woman, aged 28, with no toxic substance habits or any remarkable medical history presented at the hospital with fever, asthenia, odyndophagia, erythematous lesions in lower members, abdominal pain, arthralgias and swollen ankles, of one month onset. She initially received treatment with azitromycin, paracetamol and dexametopofen, with no alleviation.

Physical examination revealed pain on palpation in the periumbilical region, palpable purpuric lesions distal to the knees and a slight perimalleolar oedema, with no signs of inflammation. Blood count, biochemical tests, urine test, ANA, ANCA, rheumatoid factor and complement levels were normal or negative. A moderate raising of IgA immunoglobulins were observed (5.56 g/dl, normal values between 0.71 and 3.91). A biopsy of skin lesions was conducted which showed evidence of a leukocytoclastic vasculitis with IgA deposits in direct immunofluorescence test.

A diagnosis of adult IgAV was made and treatment with a dose of oral prednisone of 30 mg/day in descending order was prescribed, with improvement of abdominal clinical symptoms and of skin lesions. After 4 weeks the patient presented again with a recurrence of skin pupura, which coincided with the withdrawal of the steroids. At the time the patient presented with 4000 leukocytes/mm<sup>3</sup> (900 lymphocytes/mm<sup>3</sup>). HIV serology was positive, and confirmed with the Western blot technique, with negative serologies of lues, HCV and HBV. She presented with 183 lymphocytes CD4<sup>+</sup>/mm<sup>3</sup> and a HIV RNA of 93,348 copies/ml. Antiretroviral treatment was started with efavirenz, tenofovir and lamivudine, with suppression of viral replication being achieved and the disappearance of skin and joint symptoms. The patient has remained asymptomatic during the 4 year follow-up.

## Discussion

IgAV diagnosis is clinical and should be suspected in patients who present with the prototypical triad of non thrombocytopenic palpable purpura, abdominal pain and arthritis.<sup>6</sup> Skin biopsy is useful for demonstrating leukocytoclastic vasculitis with IgA deposits (the sample must be sent fresh in order to undertake direct immunofluorescence).

In 2012 consensus was reached regarding the term Chapel-Hill vasculitis, recognised as a class of vasculitis associated with

a probable aetiology, and as an example those associated with hepatotropic<sup>7</sup> viruses. We recommend adding the aetiology when classifying, so that in our case this would be an HIV-associated IgAV.<sup>1</sup>

Although rare, IgAV that has been linked with HIV infection is already a known fact. As in our case, and exceptionally, the study of the vasculitis may be the form of the HIV diagnosis.<sup>5-7</sup> However, the most normal occurrence is that the vasculitis is diagnosed in patients with chronic infection resulting from known HIV and precipitated by opportunistic infections or exposure to drugs.<sup>5</sup>

## Conclusions

With the presentation of this case we wish to draw attention to adult IgAV with poor evolution and suggest HIV screening, regardless of whether the clinical history contains at risk activities since we believe that a causal relationship may be established between the vasculitis and the viral infection. It appears that only antiviral treatment will be able to control the disease and this will prevent the unnecessary use of steroids or immunosuppressants.

## Conflict of interests

The authors have no conflict of interests to declare.

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