



## Case of Adult-Onset IgA Vasculitis

Catherine Teskin\*, Nikita Donti, Kunal Bhagat and Eric Russell

Department of Internal Medicine, Christiana Care Health System, USA

### Abstract

**Objective:** Review the presentation of IgA vasculitis, associated complications, and treatment.

**Methods:** We present laboratory findings and treatment related to this case, as well as, review of relevant literature.

**Results:** Seventy year-old male with a past medical history of gout recently restarted on allopurinol who presented with maculopapular lesions. Initial lab work was remarkable for acute kidney injury and urine analysis revealing proteinuria. Rheumatology was consulted and workup included elevated IgA, undetectable rheumatoid factor, cryoglobulinemia not present HIV/hepatitis panel negative, urine drug screen negative for cocaine, low C3, negative ENA panel, negative ANCA panel, and negative MPO/PR3 antibodies. A biopsy of a characteristic lesion of the skin revealed strong deposits of IgA leading to the diagnosis of IgA vasculitis.

**Conclusion:** This case demonstrates the importance of obtaining a biopsy, controversial benefit of steroids, and poorer prognosis with adults.

**Keywords:** IgA vasculitis; Henoch-Schonlein purpura; Palpable purpura; Renal dysfunction

### Introduction

IgA vasculitis, formerly called Henoch-Schonlein purpura, is an immune-mediated vasculitis associated with IgA deposition. The etiology remains unknown but a variety of triggers are recognized, including an abnormal inflammatory process deriving from immune reactions to various antigenic stimuli like bacterial, viral, or parasitic agents, in a genetically prone individual [1]. IgA vasculitis is more predominant in the pediatric population with male predominance, and higher incidence in Asian population. It is generally a self-limiting disease and characterized by a tetrad of clinical manifestations: palpable purpura without thrombocytopenia and coagulopathy, arthralgia, abdominal pain, and renal disease. Presentation may develop over days to weeks. Adults have increased risk for renal involvement and prompt diagnosis of IgA vasculitis may help prevent progression to advanced renal disease.

### Case Presentation

Seventy year-old male with a past medical history significant for severe tophaceous gout recently restarted on allopurinol, prior ICU admission for lactic acidosis of unknown etiology who initially presented for complaints of multiple, ecchymotic, hemorrhagic maculopapular lesions of his extremities that developed acutely over a 2 week period. Patient denies fevers, chills, cough, and history of cardiac or renal disease, as well as illicit drug use.

On presentation, the vital signs were normal. Lab work included unremarkable CBC, normal ESR, elevated CRP, mildly elevated CPK, significant acute kidney injury with creatinine 3.98 mmol/L with baseline of 0.9, and urine analysis remarkable for proteinuria with 300 mg/dl, significant red blood cells with >100 A, and 21-30 hyaline casts. Hospital course was complicated by an episode of significant coffee-ground emesis, lactic acidosis, and hypotension necessitating ICU transfer. Underwent endoscopy revealed esophagitis and severe portal hypertensive gastropathy. Physical exam was significant for diffuse hemorrhagic maculopapular lesions over all extremities with hemorrhagic plaques of distal fingers and tense vesicles of feet. Rheumatology was consulted and initial differential diagnosis was broad including various vasculitis such as cocaine induced, drug induced, Sweet's Syndrome, and systemic vasculitis like Wegener granulomatosis and MPO-ANCA-related vasculitis. Underwent rheumatologic workup including, elevated IgA, undetectable rheumatoid factor, cryoglobulinemia not present, HIV/hepatitis panel negative, urine drug screen negative for cocaine, C3 low at 66, C4 within normal limits, negative ENA panel, negative ANCA panel, and negative MPO and PR3 antibodies. A biopsy of a characteristic lesion of the skin revealed

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#### \*Correspondence:

Catherine Teskin, Department of Internal Medicine, Christiana Care Health System, Newark, Delaware, USA,

E-mail: catherine.teskin@gmail.com

Received Date: 03 Feb 2020

Accepted Date: 21 Feb 2020

Published Date: 25 Feb 2020

#### Citation:

Teskin C, Donti N, Bhagat K, Russell E. Case of Adult-Onset IgA Vasculitis. *Ann Clin Case Rep.* 2020; 5: 1805.

ISSN: 2474-1655

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strong deposits of IgA in vessel wall leading to the diagnosis of IgA vasculitis. Given the deposition was IgA rather than IgG or IgM without eosinophils seen, his otherwise unexplained gastropathy, negative work-up for infection and alternative vasculitides, IgA vasculitis was considered the most likely diagnosis. The lack of eosinophils makes drug induced vasculitis less likely. Patient later underwent a kidney biopsy that revealed focal segmental necrotizing and crescentic glomerulonephritis with deposits of IgA, which further supported a diagnosis of IgA vasculitis. He was stabilized from a bleeding and kidney function standpoint, and the patient was discharged on Prednisone with rheumatology follow-up.

## Discussion

The vast majority of IgA vasculitis is self-limited and can be managed in the ambulatory setting, unless there is inability to maintain oral intake, GI bleeding, changes in mental status, severe pain limiting ambulation, or renal insufficiency. Treatment is generally supportive with hydration, rest, and analgesics. The use of steroids is controversial with some studies that suggest glucocorticoids prevent renal involvement and gastrointestinal complications [2] but other studies reveal no benefit [3]. Renal biopsy is typically obtained to determine the degree of crescent formation and is the best indicator of prognosis [4]. Recurrence of IgA vasculitis is reported in about one-third of pediatric cases and relatively worse prognosis with adults. Adults are at increased risk of progression to significant

renal involvement and end-stage renal disease [5]. Treatment of IgA vasculitis nephritis is only considered in patients with marked proteinuria (>1 g/day) and/or impaired renal function during an acute episode. Immunomodulating drugs like cyclophosphamide and mycophenolate mofetil may also be considered in conjunction with IV steroids if no there is benefit with steroids alone [2].

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