Bullous Rash Leading to a Diagnosis of IgA Vasculitis with Renal Involvement Responsive to Therapy

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Bullous Rash Leading to a Diagnosis of IgA Vasculitis with Renal Involvement Responsive to Therapy

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BACKGROUND

• Formerly known as Henoch-Schönlein purpura (HSP)
• Immune complex small vessel vasculitis with IgA1-dominant immune deposits
• Most common systemic vasculitis in children, more severe disease course in adults
• Clinical Manifestations:
  - Palpable purpura
  - Arthralgias/arthritis
  - Enteritis
  - Glomerulonephritis

REFERENCE


CASE PRESENTATION

A 60-year-old male presented to the hospital with an initial complaint of a severe headache. During the admission, he developed a bullous rash on his right foot concerning for vasculitis. Lab work was significant for a leukocytosis, acute kidney injury, and elevated sedimentation rate. There also was a concern for an infectious etiology, so the patient was started on acyclovir, vancomycin, and ceftriaxone until cultures returned negative. Due to spread of the rash to the bilateral lower extremities and upper extremities, he was started on intravenous methylprednisolone.

REFERENCE


RESULTS

• Negative Rheumatologic Serologies: ANA profile, ANCAs, complements, RF/CCP, cryoglobulins, and SPEP
• Punch Biopsy of the Lower Extremity: small-medium vessel vasculitis
• Deeper Tissue Biopsy of Lower Extremity: mild IgA staining and strong perivascular C3 staining, but was felt to be insufficient evidence for IgA vasculitis
• CTA Head: no evidence of CNS vasculitis
• Kidney Biopsy: IgA nephropathy with presence of one crescent and three glomeruli with necrosis

REFERENCE


DISCUSSION

• IgA nephropathy in the setting of IgA vasculitis is generally accepted as a predictor of poor prognosis for the adult population
• Promising prognostic factors for predicting progression to ESRD include proteinuria level, macroscopic hematuria, hypertension, and proteinuria level on follow up
• Successful response to therapy with treatments such as corticosteroids, cyclophosphamide, and colchicine has been documented
• Our case outlines a biopsy proven IgA nephropathy in an adult responsive to mycophenolate mofetil and corticosteroid combined immunosuppressant therapy
• Further studies detailing successful treatment and prognostic factors are needed for the critical early detection of IgA nephropathy in adults

REFERENCES
