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

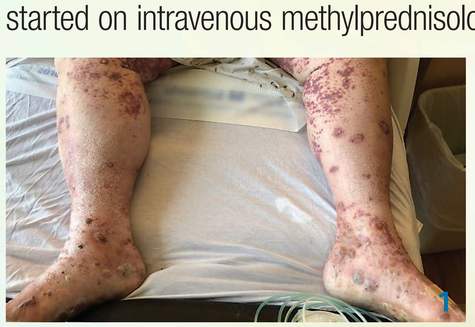
- Formerly known as Henoch-Schönlein purpura (HSP)
- Immune complex small vessel vasculitis with IgA1-dominant immune deposits[1]
- Most common systemic vasculitis in children, more severe disease course in adults[1,2]
- Clinical Manifestations:
  - Palpable purpura
  - Arthralgias/arthritis
  - Enteritis
  - Glomerulonephritis[1]

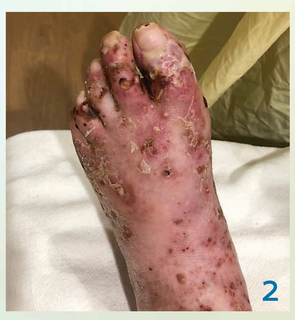
Degree of Renal Failure	Incidence [3]
End Stage Renal Disease (ESRD)	11%
Severe Renal Failure (eGFR <30)	13%
Moderate Renal Failure (eGFR <50)	14%

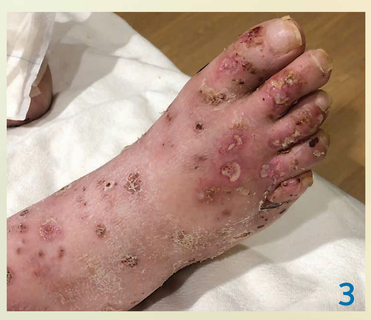
- Factors associated with progression to ESRD include a baseline proteinuria level >1 gm/day, macroscopic hematuria, hypertension, and proteinuria level >1 gm/day on follow-up[1,2]
- No optimal treatment defined with limited research in the adult population

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.







1. 1.Vasculitis rash present on the patient's bilateral lower extremities with prominent bullae on the feet.

2–3. Ruptured bullae with erosions and crusting on the patient's bilateral feet two weeks after the initial rash developed.

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

DISCUSSION

- IgA nephropathy in the setting of IgA vasculitis is generally accepted as a predictor of poor prognosis for the adult population[1,2]
- Promising prognostic factors for predicting progression to ESRD include proteinuria level, macroscopic hematuria, hypertension, and proteinuria level on follow up[3]
- Successful response to therapy with treatments such as corticosteroids, cyclophosphamide, and colchicine has been documented[1]
- **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

[1]Audemard-Verger A, Terrier B, Dechartres A, et al. Characteristics and Management of IgA Vasculitis (Henoch-Schönlein) in Adults: Data From 260 Patients Included in a French Multicenter Retrospective Survey. *Arthritis & Rheumatology*. 2017;69(9): 1862-1870. doi:10.1002/art.40178.

[2]Audemard-Verger A, Pillebout E, Guillevin L, Thervet E, Terrier B. IgA vasculitis (Henoch–Shönlein purpura) in adults: Diagnostic and therapeutic aspects. *Autoimmunity Reviews*. 2015;14(7):579-585. doi:10.1016/j.autrev.2015.02.003.

[3]Pillebout E. Henoch-Schonlein Purpura in Adults: Outcome and Prognostic Factors. *Journal of the American Society of Nephrology*. 2002;13(5):1271-1278. doi:10.1097/01.asn.0000013883.99976.22.

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