Clinical Case Reports and Reviews

Case Report ISSN: 2059-0393

Adult outset of IgA vasculitis with pulmonary and renal involvement

Sharif Muhammad, Khalid Moddathir Mohmmed*, Manal Shahid, Muhammad Haris shah and Reem Abbas

Department of Medicine, Cavan &Monaghan Hospital (RCSI Group), Ireland

Abstract

IgA vasculitis (formerly Henoch schonlein purpura) is an IgA mediated small vessel vasculitis, commonly presents in children and extremely rare in adults. We present a case of IgA vasculitis in a seventy year old man, who presented with painful pruritic rash on the lower extremities along with polyarthralgia.

Acute kidney injury evident by mildly impaired creatinine, hematuria and non-nephrotic range proteinuria were reported at presentation. Although extremely rare pulmonary involvement was evident in our case by hemoptysis and radiographic findings. Final diagnosis of IgA vasculitis and evidence of IgA deposition was confirmed with skin and kidney biopsies.

In view of kidney and lung involvements patient received short course of oral prednisone along with symptomatic management. Long term follow up showed slow but steady recovery with resolution of skin rash, joint pain, pulmonary symptoms and renal abnormalities

Although extremely rare, IgA vasculitis may need to be considered as one of the differential diagnosis in adult patient presenting with vasculitic skin rash.

Introduction

IgA vasculitis is a small vessel vasculitis associated with immunoglobulin A (IgA) complex deposition [1,2]. The immune complexes are composed of IgA1 and IgA2 but only IgA1 is found in the inflammatory infiltrate of the disease.

IgA vasculitis is commonly presenting as small vessel vasculitis in pediatric age group, while the occurrence in adults has been rarely reported. The diagnosis can be easily missed in adult patients. A high degree of suspicion and performing a biopsy of involved tissue along with immuno-fluorescence studies in suspected cases are mandatory to establish the diagnosis. Presence of Leukocytoclastic vasculitis with IgA deposition in biopsied tissue is pathognomonic finding for IgA vasculitis. Clinical manifestations can be seen in the skin, joints, gastrointestinal tract, and kidneys. Very unusual presentations it may involve pulmonary system [3].

Case presentation

A seventy-year-old man presented with nonspecific polyarthralgia and widespread lower body painful pruritic vesicular eruptions which did not blanch (Figure 1). He was admitted for further workup and management. Initial urine dipstick revealed hematuria and non-nephrotic range proteinuria. During the time in the hospital he developed mild acute kidney injury and the rash also extended more to involve the buttocks and genitals. Furthermore, he had 2 episodes of hemoptysis and a new right sided lung opacity shown up in the chest x-ray (Figure 2). In view of kidneys and lung involvement patient was treated with steroid along with analgesics and topical emollients. Subsequently he had skin biopsy which consistent with a small vessel leucocytoclastic vasculitis and IgA deposition. A Kidney biopsy shows proliferation of the mesangium, with IgA deposits on immunofluorescence and electron microscopy confirmed diagnosis of IgA vasculitis (Figure 3). Long term follow up showed slow but steady recovery with resolution of skin rash, pulmonary and renal abnormalities.

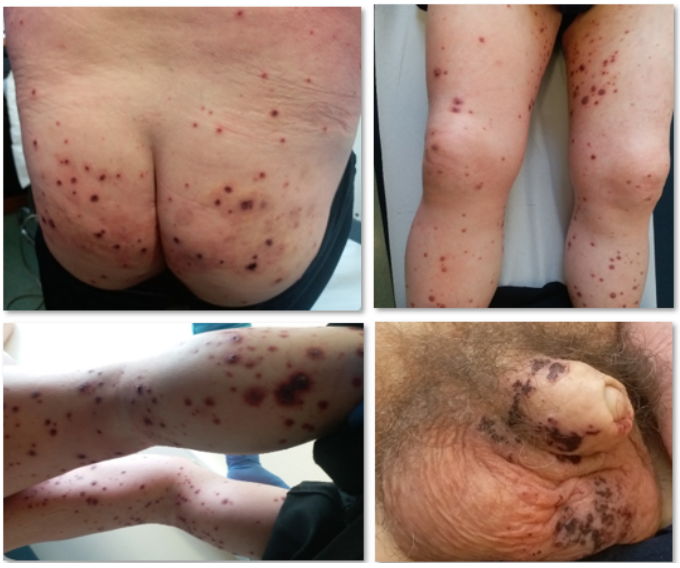

Figure 1. Typical rash of henoch schonlein purpura involving lower extremities, buttocks and genitalia

Correspondence to: Khalid Moddathir Mohmmed, Department of Medicine, Cavan &Monaghan Hospital (RCSI Group), Ireland, Tel: 00353894814563; E-mail: khalidmoddathir@gmail.com

Key words: *henoch schonlein purpura, IgA vasculitis, pulmonary, kidney, biopsy*

Received: August 19, 2019; **Accepted:** August 26, 2019; **Published:** August 29, 2019

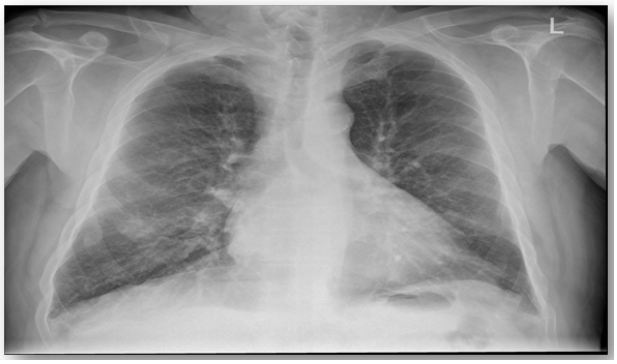

Figure 2. Chest x-ray showing right middle lobe and part of right lower lobe infiltration

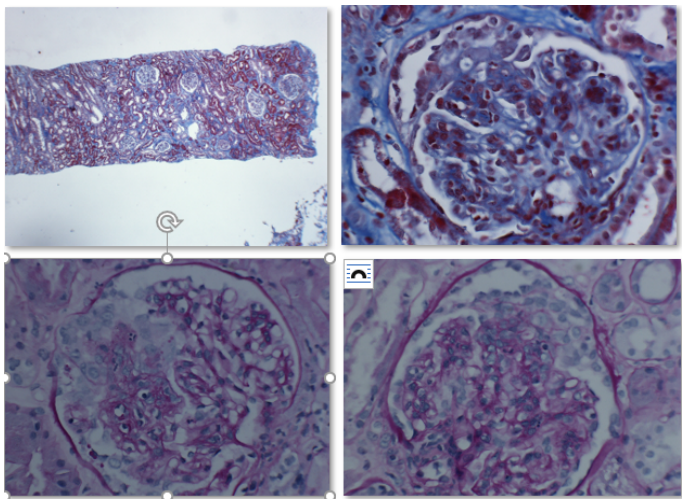

Figure 3. Kidney biopsy specimens showed deposition of IgA nephropathy, mesangial hypercellularity in a minority of glomeruli, endocapillary hypercellularity and cellular crescents, moderate arteriosclerosis and focally moderate –severe arteriosclerosis in keeping with IgA vasculitis

Discussion

Ninety percent cases of IgA vasculitis present in children with the few cases reported in adults. It is a multisystem disease, characterized by cutaneous rash, nonspecific polyarthralgias, abdominal pain and renal involvement. Rarely may involve scrotum, central nervous system, eyes and lungs.

Adults being more likely to have chronic kidney disease in the long term compared to children [4].

Extremely rare pulmonary involvement can manifest as diffuse alveolar hemorrhage and occasionally as usual interstitial pneumonia or interstitial fibrosis.

Tissue biopsy is the gold standard investigation to confirm the diagnosis and to rule out other conditions. Hypersentivity vasculitis, primary vasculitis (e.g ANCA positive vasculitidies), vasculitis secondary to connective tissue disease or infections such as Hepatitis need to be considered in the differential diagnosis.

Findings of IgA vasculitis on superficial skin biopsy are leucocytoclastic vasculitis with IgA deposition. However, renal biopsy which is usually done in case of renal involvement reveals mesangial proliferation with IgA deposition.

Management of IgA vasculitis is usually symptomatic as the disease tends to have a self-limiting course particularly in the absence of renal disease [5]. Spontaneous resolution is common within 8 weeks but can have longer course and relapses are possible.

Renal involvement is the major cause of mortality and morbidity and usually determines the prognosis therefore long-term follow-up is necessary to avoid serious and sometimes fatal complications [6].

Conclusion

Although extremely rare and easily missed, IgA vasculitis may need to be considered as one of the differential diagnosis in adult patients presenting with vasculitic skin rash.

Adults with IgA vasculitis are more likely to have more severe disease compared to children particularly if organs such as the kidneys and lungs are affected. Consequently, prompt treatment and regular follow up is often needed to prevent serious complications.

Declarations

Acknowledgments

It is a great pleasure for us to acknowledge the assistance and contributions of many individuals in making this dissertation a success. We also would like to thank all the staff members of Cavan general hospital whose contribution one way or the other aided in the management of the patient.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Ethics approval and consent to participate

Not applicable.

Competing interests

The authors declare that they have no competing interests regarding the publication of this paper.

Funding

We receive no funding support.

Availability of data and materials

Datasets used and/or analyzed for this study have been included in this published article.

Authors' contributions

Study concept and design: KMM and MS. Acquisition of data: SM, KM, MS, MHS, RA Analysis and interpretation of data: KMM, MS, and MS. Drafting of the manuscript: KMM, and MS. Critical revision of the manuscript for important intellectual content: All authors. All authors read and approved the final manuscript.

References

1. López Meiller MJ, Cavallasca JA, Maliandi Mdel R, Nasswetter GG (2008) Henoch-Schönlein Purpura in adults. *Clinics* 63: 273-276. [Crossref]

2. Saulsbury FT (1999) Henoch-Schonlein purpura in children. Report of 100 patients and review of the literature. *Medicine (Baltimore)* 78: 395-409. [Crossref]

3. Usui K (2007) Diffuse pulmonary hemorrhage as a fatal complication of Schonlein-Henoch purpura**.** *J Dermatol* 34: 705-708. [Crossref]

4. Reamy BV, Williams PM, Lindsay TJ (2009) Henoch-Schonlein purpura. *Am Fam Physician* 80: 697-704.

5. Fogazzi G, Pasquali S, Moriggi M (1989) Long-term outcome of Schönlein-Henoch nephritis in the adult. *Clin Nephrol* 31: 60-66. [Crossref]

6. Blanco R, Martinez-Taboada VM, Rodriguez-Valverde V (!997) Henoch-Schönlein purpura in adulthood and childhood: two different expressions of the same syndrome. *Arthritis Rheum* 40: 859-864. [Crossref]