A Rare Case of IGA Vasculitis Presenting as Severe Bullous Rash

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INTRODUCTION

Henoch-Schönlein purpura (HSP) also known as IgA vasculitis is the most common vasculitis in children and presents with palpable purpura, abdominal pain, and renal involvement. It is most common in males and children less than 10 years old. It often presents after recent upper respiratory illness. The classic rash associated with HSP is palpable purpura or petechiae, however other rashes have been described and can make the final diagnosis challenging.

CASE PRESENTATION

Our patient is a 9-year-old male who presented with lower extremity rash characterized by small erythematous non-blanching lesions on the ankles and lower extremities. The rash progressed up the legs along with scattered lesions on the abdomen and elbows 14 days prior to admission. Lesions progressed to larger purpura with vesicles 6 days later (Figure 1) and he was seen in the ED and prescribed oral steroids. 4 days later he developed hematuria and oral lesions and was referred to rheumatology. Upon presentation to the rheumatology clinic, he was sent to the ED given the extent vasculitis involvement. On hospital admission the patient had multiple scattered annular bullous and non-bullous lesions involving the ankles, legs, and buttocks (Figure 2). Lab work showed positive p-ANCA, negative myeloperoxidase antibody, and positive anti-streptolysin O. Skin biopsy was positive for IgA with vascular destruction and epidermal necrosis most consistent with late-stage IgA vasculitis. He was started on high-dose IV methylprednisolone for three days followed by a prolonged steroid taper, oral methotrexate, and analgesics.

CASE PRESENTATION

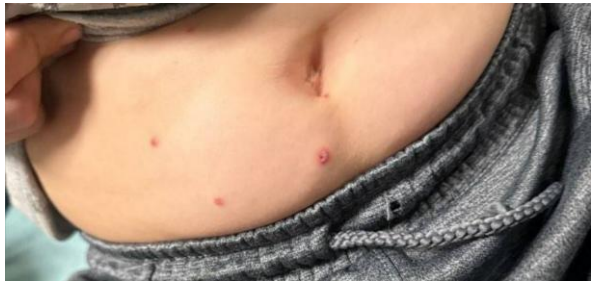

Figure 1: Abdominal skin lesions on day 7 of rash

Figure 2: Bullous lesions on the lower extremities on day of admission (day 14 of rash).

DISCUSSION

- IgA vasculitis can have multiple skin manifestations beyond the typically associated purpura.
- Clinicians should have a high suspicion for IgA vasculitis with atypical rash if the distribution and clinical scenario favors the diagnosis.
- When clinical uncertainty is present a skin biopsy can help confirm the diagnosis.

CONCLUSION

- IgA vasculitis is the most common childhood vasculitis
- While purpuric lesions are commonly associated, other skin manifestations are also possible
- If uncertain, skin biopsy may aid in the diagnosis of IgA vasculitis
- High dose steroids and immune modulators may be used in severe cases of IgA Vasculitis

REFERENCES

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