Case Report

MICROSCOPIC POLYANGIITIS (MPO-ANCA) AND HENOCH-SCHOENLEIN PURPURA OVERLAP IN A CHILD

Sarah Hasham[1], Tetyana L. Vasylyeva[1]*

Author information: *Department of Pediatrics Texas Tech University HSC at Amarillo, 1400 S. Coulter str., Amarillo, TX, USA, 79106, e-mail: Tetyana.vasylyeva@ttuhsc.edu*

Abstract: MPO-ANCA and Henoch-Schoenlein purpura overlap are extremely rare in the pediatric population. We present a case with biopsy-proven IgA renal depositions and positive MPO in a 6-year-old child with the clinical presentation of vasculitis with cutaneous and renal involvement. The child was successfully treated with a combination of steroids and cyclophosphamide.

Key words: MPO-ANCA; Henoch-Schoenlein purpura; Pediatrics.

INTRODUCTION Microscopic polyangiitis (MPA) can be distinguished from Henoch-Schoenlein purpura (HSP) based on the presence of renal, pulmonary complications, MPO ANCA seropositivity, and pauci immune glomerulonephritis; these are characteristics of MPA not found in HSP [1].
We present a unique case of HSP overlapping with MPA in a young pediatric patient. A few cases in the literature have been described for adults with HSP and IgA nephropathy with ANCA positive tests but none for a child.

CASE PRESENTATION 6 yrs old girl was referred to a nephrology clinic on account of having hematuria, proteinuria, hypertension, and recurrent HSP during the last five months. She was treated with steroids for six months due to recurrent HSP but did not respond to the treatment. On physical examination, she had a prominent palpable purpuric rash for six months and Cushingoid features (Figure 1). She did not have renal involvement at the presentation time but developed non-nephrotic range proteinuria and microhematuria 4-5 months into the disease process.

The patient had recurrent abdominal pain, arthralgias and swelling of joints, bloody stools, and palpable purpura. Kidney biopsy showed focal proliferative glomerulonephritis and IgA deposition in glomeruli (Figures 2 and 3). ANCA test was done before a renal biopsy, and MPO was positive.

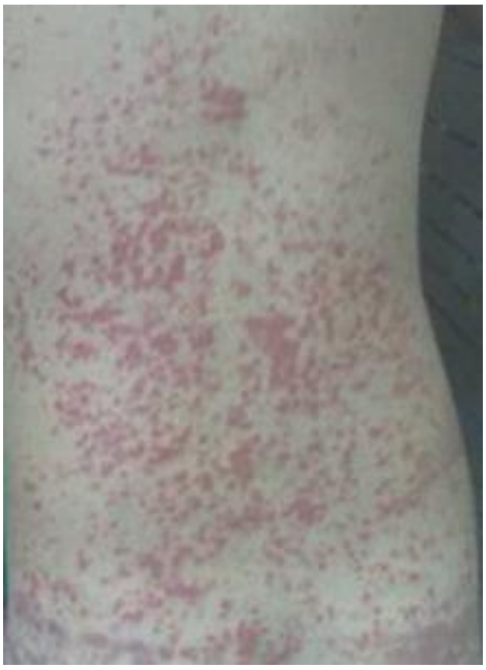

Figure 1. Typical HSP skin rash on the trunk of the patient.

She was treated with pulse-steroid therapy and Cyclophosphamide with clinical recovery.

*Corresponding author: *Tetyana L. Vasylyeva@ttuhsc.edu*
1400 S. Coulter str., Amarillo, TX, USA, 79106, e-mail: Tosin.Bewaji@ttuhsc.edu

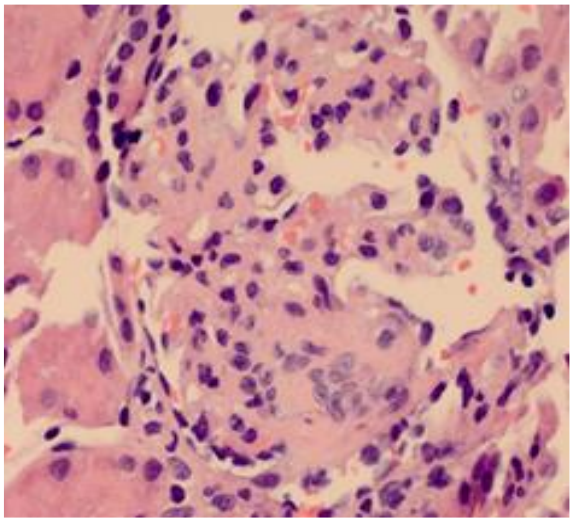

Figure 2. H&E: Mesangial expansion with increased cellularity

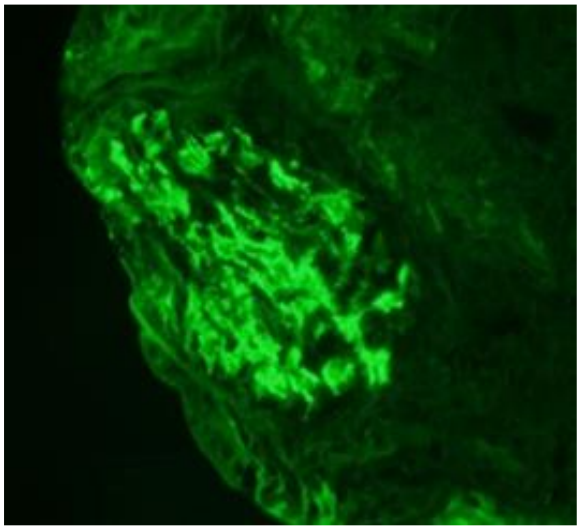

Figure 2. IF: IgA granular mesangial staining

DISCUSSION Microscopic polyangiitis is well known as ANCA systemic vasculitis, characterized by few or non-immune deposits in the affected small vessels without granuloma formation [1]. Clinical features vary, but nephritis and pulmonary bleeding and peripheral nerve neuropathy, abdominal pain, joint pain, and skin eruptions are common, whereas central nervous system manifestations are rare. It was stipulated that MPO-ANCA has complex and unique pathogenesis, with evidence for a loss of tolerance to neutrophil proteins, leading to ANCA-mediated neutrophil activation, recruitment, and injury effector T cells also involved [2]. The prognosis often remains guarded with frequent relapses and high cumulative morbidity [3].

HSP is a form of systemic vasculitis characterized by immunoglobulin A deposits in small vessels [4]. Clinical features include cutaneous purpura, arthritis, abdominal pain, gastrointestinal bleeding, and nephritis. HSP's etiology remains unknown, but IgA plays a critical role in the immunopathogenesis [4]. Renal insufficiency and pulmonary involvement are uncommon in pediatric patients, but the long-term prognosis of HSP is directly dependent on the severity of renal involvement.

Recent reports in adult literature have indicated an overlap of ANCA-associated systemic vasculitis and IgA nephropathy, although a link between these two entities has not been established [1,5].

In our case, a diagnosis of MPA and HSP overlapping syndrome was made based on clinical data and kidney biopsy and serological laboratory results. Clinical correlations suggest that immune complex deposits in the kidney may potentiate ANCA's effect in producing severe glomerulonephritis.

The same synergistic effect of IgA immune complexes and MPO ANCA may also result in rapidly progressive systemic vasculitis complications and persistent disease recurrence. Aggressive treatment with pulse steroids and cyclophosphamide for overlapping ANCA and HSP has been proposed in adult literature to prevent life-threatening multi-organ complications, which we chose for our pediatric patient led to substantial clinical improvement.

CONCLUSION Vasculitis is a rare condition with significant morbidity and mortality. Overlap MPO-ANCA with HSP, although was described in adult literature, extremely rare in pediatrics. Aggressive treatment with steroids and cyclophosphamide may lead to sustained remission of both medical conditions.

REFERENCES

1. Takeshi N, Jyuira M, Masafumi I, Kon Ron C. MPO-ANCA- and IgA positive systemic vasculitis: a possibly overlapping syndrome of microscopic polyangiitis and Henoch-Schoenlein purpura. J of cutaneous pathology 36(8): 871-7. doi: 10.1111/j.1600-0560.2008.01145.x

2. Kitching AR, Anders HJ, Basu N, Brouwer E, Gordon J, Jayne DR, Kullman J, Lyons PA, Merkel PA, Savage COS, Specks U, Kain R. ANCA-associated vasculitis. Nat Rev Dis Primers. 2020 Aug 27;6(1):71. doi: 10.1038/s41572-020-0204-y.
3. Jariwala MP, Laxer RM. Primary Vasculitis in Childhood: GPA and MPA in Childhood. Front Pediatr. 2018, 16;6:226. doi: 10.3389/fped.2018.00226.
4. Saulsbury, FT. Henoch-Schoenlein purpura in children: report of 100 patients and review of literature. Medicine (Baltimore). 1999;78(6):395-409. doi: 10.1097/00005792-199911000-00005
5. Kim, S, Dedeoglu, F. Update on pediatric vasculitis. Curr Opin Pediatr. 2005;17(6):695-702. doi: 10.1097/01.mop.0000187190.97166.83.