PAEDIATRIC DERMATOLOGY

MICROSCOPIC POLYANGIITIS AND GRANULOMATOSIS WITH POLYANGIITIS (WEGENER'S GRANULOMATOSIS) IN CHILDREN - CUTANEOUS MANIFESTATIONS

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Introduction: Antineutrophil cytoplasmic antibodies (ANCA)-associated vasculitides are rare diseases in children.

Objective: To investigate clinical, histological and serological parameters of children with microscopic polyangiitis (MPA) and granulomatosis with polyangiitis (GPA) – Wegener's granulomatosis.

Results: We summarized the data of 11 children (10F, 1M, 12.0+/-2.6 years, range: 5-15 years), with MPA and 4 patients (3F, 1M, 10+/-3 years, range: 5-16 years) with GPA, who were diagnosed and treated between 1997-2016. The 8/11 MPA patients had cutaneous manifestations at presentation: 7/11 had palpable purpura, while necrotizing vasculitis of the skin had 3/11 patients. All 4 children with GPA had cutaneous manifestations at presentation: two had necrotizing vasculitis, one had palpable purpura, and one had right upper-eyelid edema and infiltration and proptosis caused by extraocular pseudotumor, initially histologically misdiagnosed as orbital immunoglobulin G4 (IgG4)-related disease. All MPA patients had perinuclear (p) ANCA specific to myeloperoxidase (MPO), while 3/4 GPA patients had cytoplasmic (c) ANCA specific to proteinase 3 and 1/4 GPA patients had pANCA specific to MPO. Patients were followed for 3 - 12 years. All 15 patients were treated with standard immunosuppressive therapy (cyclophosphamide and methylprednisolone pulses) with gradual tapering of prednisone and introduction of mycophenolate mofetil, azathioprine or methotrexate. None of our patients died. The 3/11 MPA patients who had acute renal failure (ARF) progressed to the end-stage renal disease. ARF, necrotizing vasculitis and central nervous system involvement at presentation were parameters of poor outcome in MPA. In GPA patients, upper-airway and orbital inflammation, unlike cutaneous vasculitis and glomerulonephritis, were resistant to immunosuppressive therapy.

Conclusions: Our report emphasizes that children presenting with cutaneous vasculitis, chronic eyelid swelling, sinusitis, hoarseness and/or hematuria/proteinuria should be tested for ANCA. Timely diagnosis and adequate immunosuppressive treatment enable a favorable prognosis of MPA and GPA in children.