

## IMMUNOGLOBULIN G4 (IGG4) RELATED DISEASE AND GRANULOMATOSIS WITH POLYANGIITIS (GPA) IN CHILDHOOD: A CASE REPORT OF NEW OVERLAP SYNDROME

*Milena Manojlović<sup>1</sup>, Dragana Lazarević<sup>1,2</sup>, Marija Ratković Janković<sup>1,2</sup>, Sandra Stanković<sup>1,2</sup>, Emilija Golubović<sup>1,2</sup>, Jelena Vojinović<sup>1,2</sup>*

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis (WG) is one of major variants of anti-neutrophil cytoplasm antibody (ANCA)-associated vasculitis (AAV) and one of the most common vasculitis in children affecting small- to medium-sized blood vessels. Immunoglobulin G4 (IgG4)-related disease is characterised by inflammatory pseudotumours with elevated serum IgG4 concentrations. Orbital pseudotumour in children can be initial clinical presentation of both IgG4 related disease and GPA. Herein, we describe a young girl with a new overlap syndrome, initially presented with orbital pseudotumour. Furthermore, diagnostic challenges and applied therapy are described.

*Acta Medica Medianae 2023;62(3): 103-108.*

**Key words:** *granulomatosis with polyangiitis (GPA), immunoglobulin G4 (IgG4)-related disease, orbital pseudotumour*