Igg4-related disease of the middle ear in a pediatric patient – the first reported case

Filip Hergešić[3], Lucija Grbac[2], Jakov Ajduk[1,2]

[1]Klinika za otorinolaringologiju i kirurgiju glave i vrata, KBC Sestre milosrdnice, Vinogradska 29, 10000 Zagreb, Hrvatska

[2]Medicinski fakultet Sveučilišta u Zagrebu, Šalata 3b, 10000 Zagreb, Hrvatska

[3]Klinika za otorinolaringologiju i kirurgiju glave i vrata, KB Sveti Duh, Sveti Duh 64, 10000 Zagreb, Hrvatska

filip.hergesic@gmail.com

Objective: To present the first reported case of IgG4-related disease (IgG4-RD) of the middle ear in a pediatric patient and to highlight diagnostic challenges and treatment options. Materials and methods: A 15-year-old girl presented with unilateral ear fullness and conductive hearing loss. Otoscopic, audiological, radiological, and histopathological examinations were performed, including immunohistochemical staining for IgG4. Results: The initial diagnosis was nonspecific chronic otitis media. After two surgeries, soft tissue from the tympanic cavity and mastoid was removed. Histology revealed dense plasmacytoid infiltration with fibrosis, and immunohistochemistry showed more than 50 IgG4-positive plasma cells per high-power field and an IgG4+/IgG ratio above 40%. Serum IgG4 levels were normal. MRI of the head, neck, thorax, and abdomen excluded additional lesions. Following surgery and a six-month course of oral corticosteroids, symptoms partially improved and no recurrence was observed during a one-year follow-up. Conclusion: IgG4-RD of the middle ear is extremely rare and has not previously been described in children. As it may mimic chronic otitis media, this diagnosis should be considered in atypical presentations. Early recognition enables targeted surgical and immunosuppressive treatment, preventing potential complications.

Key words: Immunoglobulin G4-related disease, middle ear, pediatrics