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SCIENTIFIC LETTER

IgG4-Related Coronary Aneurysm in a Child

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To the Editor: A 13-y-old boy presented with fever and erythematous lips for 2 wk. Erythrocyte sedimentation rate (ESR) was 65 mm/h and C-reactive protein (CRP) 31 mg/L. American Heart Association (AHA) diagnostic criteria could not be met for classic or incomplete Kawasaki disease. Echocardiography was normal. Treated as pneumonia based on radiographic findings, fever subsided and he was discharged.

On follow-up after 6 wk, he was asymptomatic. ESR increased to 80 mm/h, CRP 84 mg/L and high-resolution computed tomography (HRCT) revealed pericardial effusion, multiple mediastinal lymphadenopathy, and persistent lung consolidation. F-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) had metabolic activity in thymus, pericardium, right iliac bone, and mediastinal lymph nodes.

Repeat echocardiography after 4 wk showed coronary artery aneurysm; left coronary artery diameter was 4.5 mm (z score 5 to 10) and left anterior descending (LAD) artery diameter was 6 mm (z score 5 to 10), proximal aortitis and impaired left ventricular (LV) function. Serum immunoglobulin G (IgG), immunoglobulin A (IgA), and immunoglobulin G4 (IgG4) levels were elevated, IgG4 - 292 mg/dL (4.90–198.5 mg/dL). Mediastinoscopy-guided lymph node biopsy was done; on immunohistochemistry, IgG4-positive plasma cell count per high power field was 30–35. The ratio of IgG4+/ IgG+ plasma cells was 35%–40%. He was diagnosed with IgG4-related disease based on clinical and pathological findings.

He was treated with Deflazacort equivalent to 1 mg/kg/d of prednisolone for 7 mo. ESR and CRP became normal, IgG4 decreased to 143 mg/dL and cardiac magnetic resonance imaging (MRI) showed normal sized coronaries. The boy had no relapse at 1-y follow-up after cessation of treatment.

IgG4-related disease is diagnosed by the presence of serum IgG4 level > 134 mg/dL, tissue infiltration of >10 IgG4+ cells per high power field, and an IgG4+/IgG+ cell ratio > 40% [1]. The absolute number of IgG4+ plasma cells per high power field do vary in organs [2]. IgG4-related coronary involvement has been reported in adults [3]. Cyclophosphamide, mycophenolate mofetil, and rituximab are the steroid sparing agents [4].

IgG4-related coronary involvement has not been reported in pediatric literature and we also wish to emphasize the remarkable treatment response.

Declarations

Conflict of Interest None.

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