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Pachymeningitis in a pediatric case of IgG4-related disease successfully treated with mycophenolate mofetil

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Immunoglobulin G4-related disease (IgG4-RD) is a systemic fibro-inflammatory disease that may affect any part of the body [1]. Pathologically it is characterized by storiform fibrosis, obliterative phlebitis, and dense lymphocytic infiltrate rich in IgG4+ plasma cells [1, 2]. In children, the most common manifestations are related to pancreatic, ocular, and lymph node involvement [3]. Central nervous system involvement is rare and may be in the form of pachymeningitis, tumor-like mass, and hypophysitis [3–5]. Herein, we present a pediatric case of IgG4-RD presented with ocular and cerebral involvement successfully treated with corticosteroids and mycophenolate mofetil.

A 17-year-old girl was admitted to the in-patient clinic for headache, double vision, and painful and limited right eye movements for 2 weeks. Ocular examination revealed right optic disc edema and cranial magnetic resonance imaging (MRI) showed diffuse dural thickening and contrast enhancement (Fig. 1). Acute phase reactants were elevated [erythrocyte sedimentation rate (ESR): 74 mm/h, C-reactive protein (CRP): 137 mg/L)]. Blood, urine, and cerebrospinal fluid (CSF) examinations were negative for infectious and autoimmune etiologies. She was treated with corticosteroids and diazomide for a month with the provisional diagnoses of optic neuritis and pachymeningitis. She had complete resolution of her ocular symptoms on the 3rd week and corticosteroids were discontinued on the 6th week. Four months later, the same complaints (headache, limited and painful right eye movement) recurred and the child was referred to pediatric rheumatology for the work-up. Physical examination was normal except for the restricted lateral movement of the right eye which was compatible with 6th nerve palsy. She had elevated acute phase reactants (ESR: 86

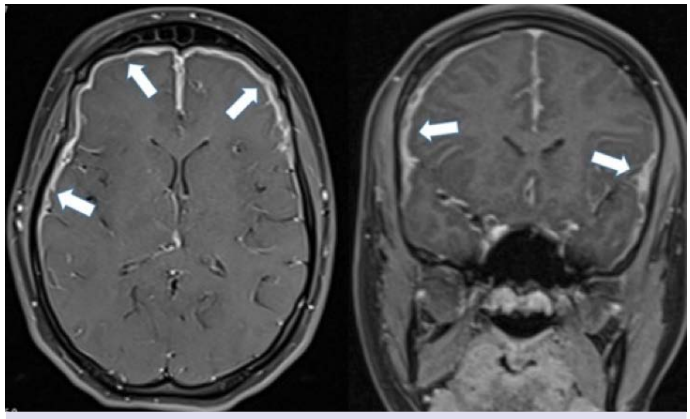

Figure 1. Axial (left) and coronal (right) post-contrast T1W images show diffuse dural thickening and contrast enhancement, which was more prominent at the level of bilateral frontal lobes, partially extending to the parietal and temporal lobes (arrows).

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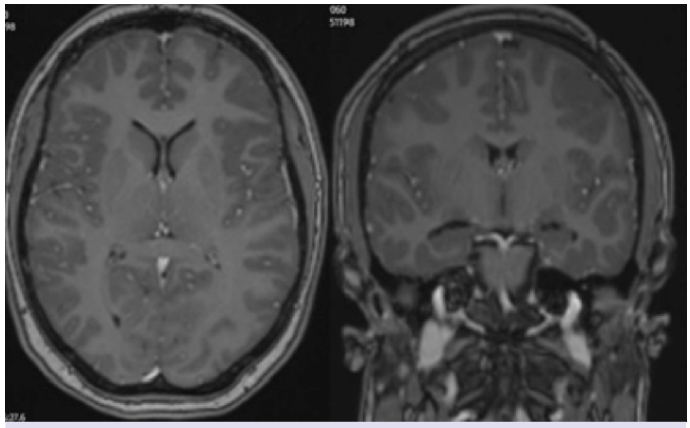

Figure 2. Axial (right) and coronal (left) post-contrast T1W images show resolution of all dural enhancement and thickening.

mm/h, CRP: 170 mg/L). Repeated extensive CSF and blood studies for an infectious or autoimmune etiology were negative. Thoraco-abdominal MR angiography was normal. Control cranial MRI showed the persistence of contrast enhancement and thickening of the meninx on the bilateral frontotemporal regions. IgG4-related disease was in the differential diagnosis list and both serum Ig G [2066 mg/dl (normal: 552–1631)] and Ig G4 [261 mg/dl (normal: 4.9–198.5)] were elevated. The child did not have any lymphadenopathy and pancreatic function tests were normal. Dural biopsy was performed and reported as inadequate material rich in fibrosis. The child was diagnosed as IgG4-RD and corticosteroid and mycophenolate mofetil treatments were started. Corticosteroids were discontinued on the 4th month. On the 6th month MRI features regressed totally (Fig. 2) and the control IgG4 levels were normalized (IgG4: 68 mg/dl). The child is being followed for 2 years under mycophenolate mofetil treatment with complete remission.

In conclusion, IgG4-RD in children may manifest as isolated pachymeningitis. Mycophenolate mofetil seems to be an effective corticosteroid sparing agent in IgG4-RD.

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