

A case with Immunoglobulin G4 related Hypertrophic Pachymeningitis mimicking brain tumor

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63 male with right frontal scalp mass visited to neurosurgery department. Brain magnetic resonance imaging (MRI) revealed 1.5 cm enhancing mass with adjacent subgaleal tissue thickening/enhancement and bony destruction in the right frontal area. Dural thickening/enhancement and leptomenigeal enhancement along the right cerebral convexity was shown. Differential diagnosis recommended for malignant bone tumor or bone metastasis with combined pachymeningeal and leptomenigeal metastasis. Operation was done for diagnose and remove mass. Biopsy showed hypertrophic pachymeningitis with marked lymphoplasmacytic, neutrophilic and histiocytic infiltration, dense fibrosis, destruction of skull. There were increased Immunoglobulin G4 (IgG4)-positive cells (up to 68/HPF). Consultation to rheumatology was done and patient was transferred to rheumatology department for further workup and management. He had dry eye and both ear hearing difficulty for several years. However, there was no further immunoglobulin infiltration in other organ systems. Plasma IgG was 1094.4 mg/dL (680-1620) and IgG4 testing 9.6 mg/dL (3.9-86.4). Antinuclear antibody, antineutrophil cytoplasmic antibody, rheumatoid factor, anti-cyclic citrullinated peptide, angiotensin converting enzyme level were all normal. Erythrocyte sedimentation rate (ESR) and C- reactive protein (CRP) were 13 mm/hr (0-10) and 1.5 mg/dL (0-0.5) respectively before the operation, and they decreased to 0.68 mm/hr, and 0.06 mg/dL at the 4 days after operation (POD 4). Although naproxen and antibiotics had been done after operation, they increased 21 mm/hr and 3.99 mg/dL at consultation day. ESR and CRP decreased dramatically just one day after starting prednisolone (0.5 mg/kg). They became to be normalized at the 5 days after steroid therapy. Now we are tapering oral prednisolone. Followed-up brain MRI findings was no mass redeveloped except mild dural thickening/enhancement along the right convexity. Here we report a case with IgG4 related- hypertrophic pachymeningitis mimicking brain tumor, which was successfully treated with oral steroid.

