Hypertrophic Cranial Pachymeningitis with Inflammatory Valvular Disease

To the Editor: We read with interest the article by Tojo et al (1) indicating an autopsy case of Wegener's granulomatosis associated with pachymeningitis. We describe a rare case of hypertrophic cranial pachymeningitis (HCP) presenting with valvular disease of the heart and a complete atrioventricular (A-V) block.

A 47-year-old man with a 4-year history of HCP was admitted to our hospital because of exertional dyspnea. Four years earlier, the patient had been diagnosed with HCP by a head magnetic resonance imaging (MRI) and had received two courses of steroid therapy (methylprednisolone 500 mg/day for 3 days) with resolution of the cranial nerve paralysis. The next year, he began to complain of dyspnea on exertion. On admission, he was febrile (37.2°C). His lungs were clear, but pansystolic murmurs were heard over the apex. He had neither arthritis nor tonsillitis. A neurological examination showed oculomotor palsy in the right eye and a left cranial nerve XII palsy. The erythrocyte sedimentation rate was 65 mm/h, and the C-reactive protein titer was 7.7 mg/dl. His serum immunoglobulin G and M were 2,203 mg/dl and 352 mg/dl, respectively. Rheumatoid factor, antinuclear antibody, antistreptolysin O and serum angiotensin converting enzyme were all normal. A sagittal T1-weighted MRI showed an isointense lesion involving the upper cervical cord. An electrocardiogram showed a complete A-V block which had not been detected two years earlier. A subsequent echocardiographic examination revealed diffuse thickening of both mitral and aortic valves, with the absence of calcification and verrucae. Cardioangiography demonstrated both aortic and mitral valve regurgitation. An aortic valve replacement, a mitral valvuloplasty, and the implantation of a permanent pacemaker were performed. The removed aortic valve was soft and thick, and microscopic examination revealed that the aortic valve had been infiltrated with lymphocytes and plasma cells, but without granulation or vasculitis; this is consistent with acute and chronic inflammation. However, bacteriological and fungal cultures were negative. After the operation, his clinical course was uneventful and he was subsequently discharged.

HCP is a rare disease characterized by thickening of the dura mater, which leads to cranial nerve palsy, confusion and/or cerebellar ataxia (2). The

pathogenesis of HCP is unclear, but in some studies it has been suggested to be related to autoimmune and connective disorders (3, 4). The histological findings of the affected dura in patients with HCP generally reveal acute and chronic inflammatory changes (2, 3), which was similar to the resected aortic valve in our patient. This may suggest a relationship between the HCP and the heart disease in our case, although no evidence indicating an underlying autoimmune process was noted. There has only been one other reported case of HCP associated with a complete A-V block (5). In both cases, the underlying etiologic mechanisms remain obscure. However, to our knowledge, this is the first case report of HCP associated with valvular disease.

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