
MR of Cerebral Whipple Disease

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Summary: A case of cerebral Whipple disease is reported. MR findings of the brain are discussed in relation to neuropathologic lesions reported previously. Repeated MR investigations may serve as a valuable tool to evaluate long-term efficacy of treatment in cerebral Whipple disease.

Index terms: Brain, inflammation; Brain, magnetic resonance

We present a case of Whipple disease, a systemic infection frequently affecting the small bowel, lymph nodes, joints, muscle, and other tissues, in which there was neurologic involvement.

Case Report

A 50-year-old man was admitted with a 3-year history of weight loss and recurring episodes of fever. Histologic examination of an inguinal lymph node showed characteristic periodic acid-Schiff-positive macrophages, compatible with Whipple disease. Three weeks later, blurred vision developed. On neurologic examination, dyskinetic eye movements, generalized myoclonic jerks, and an amnesic syndrome were found. Cerebrospinal fluid examination showed a mild lymphomonocytic pleocytosis. Magnetic resonance (MR) of the brain showed minimal morphologic changes with small areas of hyperintensity on T2-weighted images in the left mediobasal temporal lobe (Fig 1A). The clinical course initially worsening, generalized seizures followed by respiratory failure occurred despite the recommended treatment for Whipple disease. Four weeks later, MR of the brain showed confluent hyperintense lesions in the mediobasal parts of both temporal lobes, the hypothalamus, and the basal ganglia on both sides with a moderate mass effect and a circumscribed contrast enhancement (Fig 1B-D). Treatment was changed to ceftriaxone followed by a course of chloramphenicol. In the following weeks, myoclonic jerks and dyskinetic eye movements slowly improved. Six months later, MR investigation showed nearly complete resolution of the previously observed lesions, but moderate cerebral atrophy was present.

Discussion

Neurologic involvement is a serious complication of Whipple disease, with a frequency of approximately 10% (1). In our patient myoclonus, generalized seizures, an amnesic syndrome, and dyskinetic eye movements developed, all of which appear to occur frequently in cerebral Whipple disease (1-7). MR investigations in our patient as well as in previous reports (2-5) showed good correlation regarding preferential sites of the well-known neuropathologic lesions which consist of granulomatous polioencephalitis or panencephalitis, involving the basal parts of the telencephalon, the hypothalamic nuclei, thalamus, the periaqueductal gray, and the tectum pontis (1, 6). Signs of nodular-granular ependymitis, another typical neuropathologic finding, could not be demonstrated on our MR scans. Repeated MR investigations showed different extent and degree of signal alteration at various stages of the disease, indicating different stages in the inflammatory process and different degrees of edema. Contrast enhancement of the mamillary bodies may be attributable to a focal disruption of the blood-brain barrier; the combination of that enhancement with bilateral signal-intensity changes on T1-images might indicate a progression of the inflammation. Despite the recommended treatment for cerebral Whipple disease (8), both clinical symptoms and MR lesions initially worsened. Therefore, treatment was changed to ceftriaxone and chloramphenicol, leading to moderate clinical improvement confirmed by radiologic resolution of the lesions. MR investigation in our patient 6 months after onset of symptoms showed moderate cerebral atrophy. MR findings in our patient therefore document both delayed treatment

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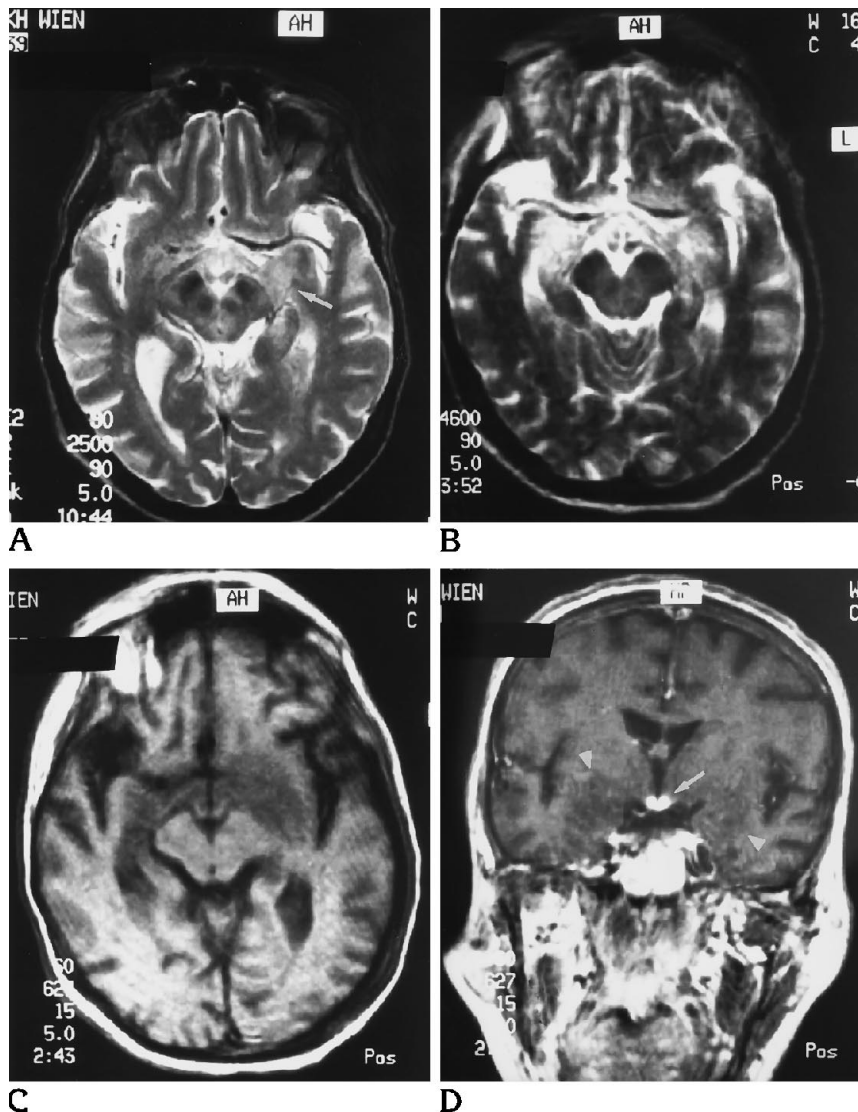


Fig 1. A, Axial T2-weighted MR image shows increased signal intensity in the temporal lobe (amygdala, hippocampal gyrus), mamillary bodies, and optic tract on the left (arrow).

B, Axial T2-weighted MR image shows bilaterally increased signal intensity in the medial temporal lobes, the amygdala, the hippocampal gyrus, the mamillary bodies, optic tracts, and hypothalamus.

C, Axial T1-weighted MR image shows bilaterally decreased signal intensity in the same areas as in A, with moderate mass effect.

D, Coronal T1-weighted MR image after intravenous gadolinium-DTPA shows intensive enhancement of mamillary bodies (arrow) with decreased signal intensity in the parahippocampal gyrus, hypothalamus, olfactory area, and optic tracts (arrowheads).

response and irreversible brain damage in cerebral Whipple disease.

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