Case Report

CT and MR Imaging in Neuro-Bęhcet Disease

Willeit, E. Schmutzhard, F. Aichner, U. Mayr, F. Weber, and F. Gerstenbrand

Abstract: Computed tomography and magnetic resonance (MR) studies were done in one case of Bęhcet disease and showed a focal lesion in the upper brain stem. Early institution of therapy was followed by a complete clinical remission and disappearance of the CT and MR changes. Computed tomography and MR are important modalities for the early detection of Bęhcet disease when reversibility of the pathological changes is still possible. Index Terms: Bęhcet disease—Brain, diseases—Computed tomography—Magnetic resonance imaging.

Bęhcet disease, a rare condition in central Europe, is more common in Turkey, where it was originally described, as well as in the Arab countries, Japan, and China (1–3). The classic symptom triad comprises oral and genital aphthous ulcers, and relapsing iritis (4). Knapp first described its neurological complications (5), which affect up to one-third of Bęhcet patients (6–8). Focal necrotic lesions, mainly localized in the brain stem and basal ganglia, represent the most common neuropathological findings (9,10). In the early stages, perivascular lymphocytic infiltrations and, less frequently, thrombosis of blood vessels predominate (11,12). Only a few authors have reported CT findings in neuro-Bęhcet disease (13–16) and, to our knowledge, the magnetic resonance (MR) findings have not yet been described. The aim of this study is to demonstrate the possibility of establishing by CT and MR the diagnosis of the CNS manifestations of Bęhcet disease at an early stage, when the reversibility of the cerebral lesions with the appriopriate treatment may still be feasible.

CASE REPORT

A 35-year-old woman had a 7 year history of oral and genital aphthous ulcers, relapsing iridocyclitis, and migratory polyarthralgias. In 1978 a skin biopsy showed typical histological signs of vasculitis (3). In 1984 she was admitted because of dizziness, headache, unsteadiness, and partial loss of vision in the right eye for a period of 4

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1a,b

1c,d

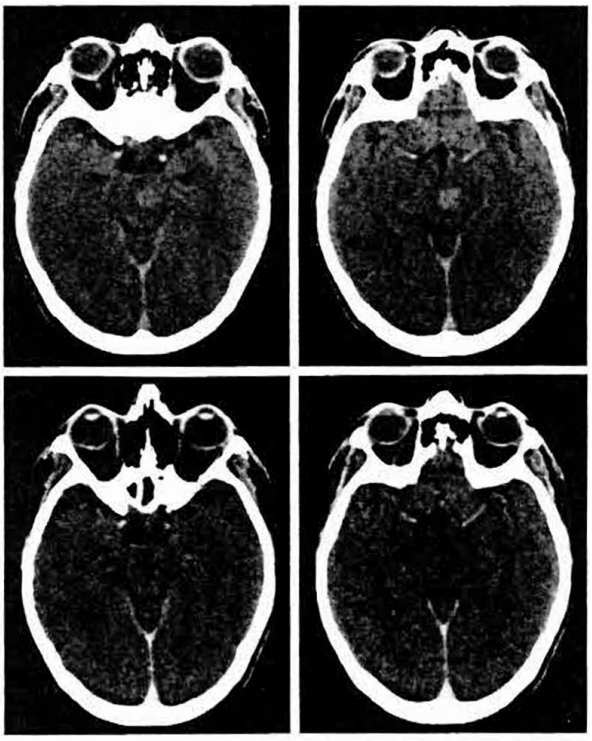

FIG. 1. a and b: Contiguous postcontrast CT scans show slightly enhancing focal lesion in upper brain stem. c and d: Follow-up study 3 weeks later reveals normal findings.

weeks. Neurological examination showed impairment of upward conjugate gaze, nystagmus on downward gaze, and a mild horizontal nystagmus on gazing to the right side. The pupils were anisocoric, the right pupil being irregular and miotic. In addition, a slight right hemiparesis was present. The deep tendon reflexes were brisk, and Babinski sign was present bilaterally. The cerebellar functions were normal. Cerebrospinal fluid (CSF) analysis showed a slightly increased cell count (26/mm^3) and a normal protein content. IgG, IgA, and IgM and CSF electrophoresis were in the normal range. The C-reactive protein in the CSF was 28.6 mg/L ml (normal: <1.8 mg/100 ml). The electroencephalogram was abnormal with diffuse theta activity. Ophthalmological examination revealed iridocyclitis of the right eye and signs of retinal vasculitis. HLA-B5 was positive and the antinuclear factor and rheumatoid factor (Latex) were negative. Complement-fixation reactions to a wide range of neurotropic viruses were negative. Besides a mild leukocytosis and increased erthrocyte sedimentation rate (24 mm in the 1st h) all routine laboratory parameters were within normal limits. The electrocardiogram was normal.

Precontrast CT was considered normal, whereas after infusion of contrast medium an enhancing mass measuring 12 mm in diameter located in the left upper brain stem was detected (Fig. 1). Magnetic resonance was performed on a 0.15 T superconducting magnet with sagittal and axial images obtained with spin echo (SE) and inversion recovery (IR) sequences. The T1 weighted images [SE: repetition time (TR), 500 ms; echo time (TE), 40 ms] were normal (Figs. 2a and b)]. The T2 weighted images (SE: TR, 2,320 ms; TE, 80 ms) showed a high signal in the left upper brain stem corresponding to the CT findings as well as to the clinical focal symptoms (Figs. 2c and d). Using an IR sequence (TR, 1,900 ms; T1, 500 ms) a low signal was seen in the corresponding area (Fig. 2e).

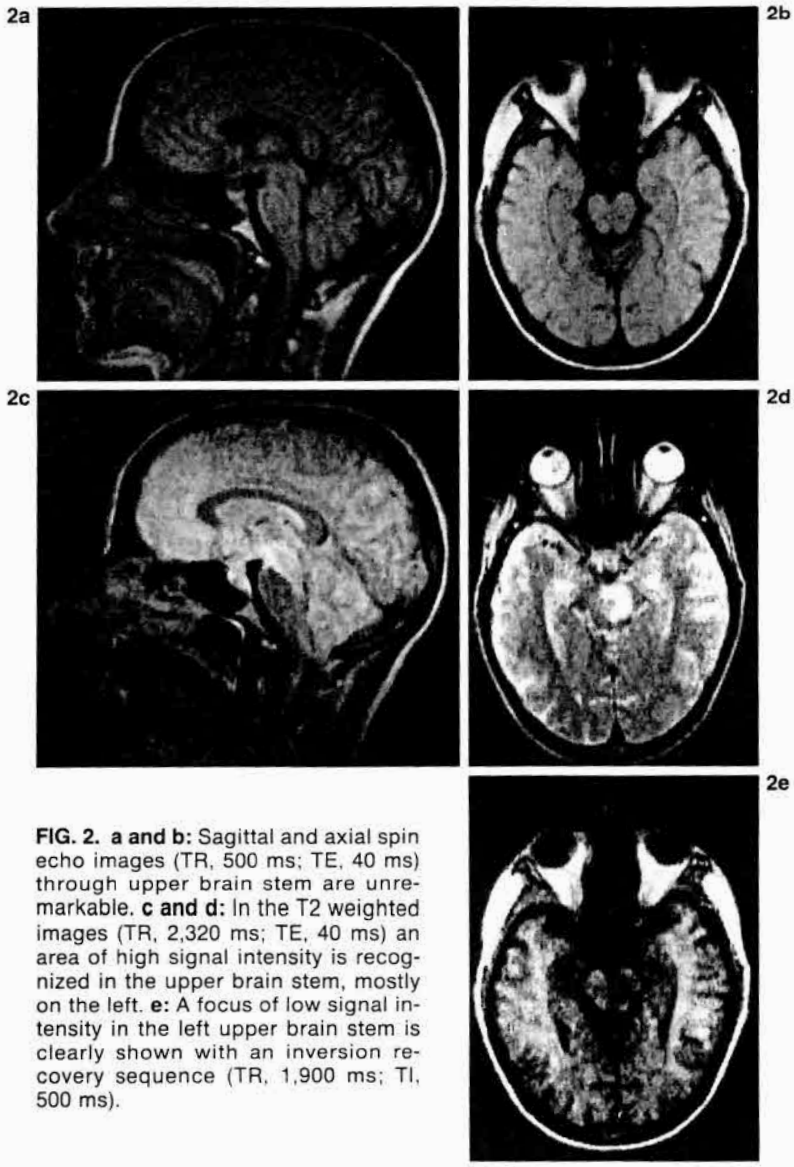

FIG. 2. a and b: Sagittal and axial spin echo images (TR, 500 ms; TE, 40 ms) through upper brain stem are unremarkable. **c and d:** In the T2 weighted images (TR, 2,320 ms; TE, 40 ms) an area of high signal intensity is recognized in the upper brain stem, mostly on the left. **e:** A focus of low signal intensity in the left upper brain stem is clearly shown with an inversion recovery sequence (TR, 1,900 ms; TI, 500 ms).

Therapy with intravenous prednisolone (60 mg daily) and azathioprine (50 mg three times a day) was initiated and was followed by a dramatic clinical improvement within 3 weeks. The focal symptoms related to the upper brain stem resolved completely. The CSF returned to normal. Repeat CT (Figs. 1c and 1d) and MR scans (not shown) revealed that the focal change in the upper brain stem had disappeared.

DISCUSSION

The etiology of Behcet disease, suspected to be viral (4,9) or based on an autoimmune disorder (17,18), is unknown. The diagnosis is based on the presence of at least two of the three features of the classic triad—recurrent mouth and genital aphthous ulcers and ocular inflammation—or one of these features combined with changes of the CSF (19–22). On CT hypodense lesions (13,15,16) have been described; we are not aware of any MR report in this syndrome. In our case the focal lesion of the left upper brain stem could be demonstrated on both CT and MR. In CT the lesion was visible only after contrast enhancement. This might be due to a focal abnormality of the blood–brain barrier related to neurovasculitis encountered in Behcet disease (11,12,17,18). Magnetic resonance revealed corresponding changes in the upper brain stem. As the neurological deficit and the CT and MR findings in our case proved to be reversible, we think that necrosis and scarring had not yet occurred at the early stage of the disease. Thus, our study demonstrates the ability of CT and MR to show focal cerebral abnormalities, early in the course of neuro-Behcet disease.

The prognosis of Behcet disease is variable and unpredictable and spontaneous remissions can occur. No reliably effective therapeutic program has yet evolved. Wolf et al. (6) found a mortality of 41% in 65 cases; others did not confirm this adverse prognosis (1,2,23). The best response to corticosteroids or other immunosuppressive agents is obtained with an early institution of therapy (12). Hence, cerebral CT and MR might be helpful for the early diagnosis, as in some cases the involvement of CNS may precede the classic symptom triad (14,15,23,24) and is not always associated with pathological CSF changes (1,25).

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