MIR in atypical idiopathic inflammatory demyelinating disease treated with methylprednisolone and cyclophosphamide

U. Kallweit, D. Pöhlau, D. Pauleit¹, M. Harzheim

Department of Neurology, Kamillus-Klinik, Asbach, 1Radiologische Gemeinschaftspraxis, Bad Honnef, Germany

The differential diagnosis of multi-focal enhancing lesions in cranial magnetic resonance imaging (MRI) is often difficult and includes a wide spectrum of differential diagnoses including atypical idiopathic inflammatory demyelinating lesions.^[1,2] We report an 18-year-old woman who developed severe multi-focal neurological symptoms within few weeks presenting with somnolence, dysarthria, asymmetric tetraparesis and moderate ataxia. The cerebrospinal fluid (CSF) analysis revealed a mild pleocytosis (17/3 mononuclear cells), a dysfunction of the blood brain barrier and oligoclonal bands without corresponding findings in the serum. The MRI on the day of admission showed large, partly confluent lesions with perifocal edema, T1-hypo-intensity as a correlate of severe tissue damage as well as concentric gadolinium enhancing lesions



Figure 1: (a) Day 1, prior to treatment: large, subcortical and juxtacortical, partly confluent lesions with perifocal edema and a slight mass effect in the supratentorial white matter in T2 weighted images, hypointensity and also concentric gadolinium enhancing lesions in T1 weighted images. (b) Day 7, after treatment with cumulative doses of 10000 mg methylprednisolone and 1000 mg cyclophosphamide: decrease of mass effect and edema, of lesion volume and especially of gadolinium enhancement. (c) Day 53, after treatment with cumulative doses of 10000 m methylprednisolone and 2000 mg cyclophosphamide: massive progression in lesion load and especially new juxtacortical lesions in T2 weighted images as well as more than 10 gadlinium enhancing lesions on T1 weighted images. (d) Month 17, after treatment with cumulative doses of 20000 m methylprednisolone and 3000 mg cyclophosphamide: new gadolinium enhancing lesion on T1 weighted images in the brainstem (arrow). (1.0 T, FOV 230/1.2, TR (T1) 678, TR (T2) 5056), TE (T1) 15, TE (T2) 100)

Michael Harzheim

Department of Neurology, Kamillus-Klinik, Hospitalstr. 6, D-53567 Asbach, Germany. E-mail: michael.harzheim@kamillus-klinik.de

[Figure 1a]. Immediately, high-dose methylprednisolone (2000 mg/day on seven following days) and high-dose cvclophosphamide (1000 mg = 600 mg/m^2) were administered intravenously. A second cyclophosphamide pulse of identical dosage was performed exactly one week later. The MRI on day 7 showed a decrease of mass effect and edema, of lesion volume and especially of gadolinium enhancement [Figure 1b]. Thirteen days after the initiation of the therapy neurological examination revealed mild psychomotor deficit and a latent left-sided hemiparesis. Although the clinical course seemed to be stabile six weeks later, once more 5×2 g methylprednisolone and 1 g cyclophosphamide was administered because brain MRI revealed new partly confluent and gadolinium enhancing lesions [Figure 1c]. After 17 months a nuclear facial palsv occurred and brain MRI showed a gadolinium enhancing lesion in the brainstem [Figure 1d]. Due to clinical, laboratory and MRI findings, multiple sclerosis (MS) with dramatic multi-focal onset mimicking a Marburg type of the disease was diagnosed.^[3,4] The

further clinical course was relapsing-remitting. Frequent clinical and MRI controls are essential for diagnosis and effects of treatment in malignant forms of idiopathic inflammatory demyelinating diseases.

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Accepted on 01-02-2008