

MULTIPLE SCLEROSIS WITH EARLY CHILDHOOD ONSET

A CASE REPORT

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SUMMARY — A 2 year old boy was admitted owing to a subacute episode of ataxic gait and hearing deficit. Computerized tomography (CT) was normal and cerebrospinal fluid (CSF) analysis revealed gamma globulins level of 15.4% (normal 7 to 14%). There was spontaneous remission after 7 months. At 5 years of age the boy incurred a second episode with predominantly right apendicular ataxia and tonic gaze deviation to the right side. CT showed a low-density lesion in the white matter adjacent to the right frontal horn. Visual and auditory evoked potentials were abnormal. CSF revealed a mild increase in gamma globulins level of 14.5% with an abnormal T lymphocyte subsets study. The combination of visual, cerebellar, brain stem and paraventricular lesions with clear remissions and exacerbations, supported by CT, CSF and evoked potentials findings suggests the diagnosis of multiple sclerosis even at this early age.

Esclerose múltipla na infância: relato de um caso.

RESUMO — Os autores relatam o caso de um menino com provável esclerose múltipla, de início aos 2 anos de idade, com um segundo surto aos 5 anos. O acometimento foi predominantemente de tronco cerebral, cerebello, nervo óptico e paraventricular. Tomografia computadorizada, exame do líquido cefalorraqueano com eletroforese de proteínas e estudo de subpopulações linfocitárias T, bem como a avaliação dos potenciais evocados visuais, auditivos e sômato-sensitivos sugeriram o diagnóstico.

The prevalence of multiple sclerosis (MS) is 50:100000, depending on geographical area and ethnic origin¹⁴. It rarely occurs before 10 years of age, comprising some 0.2 to 0.4% of the total patients¹⁴. The present paper reports a case starting early in childhood and emphasizes that such a diagnosis should be considered even at such an early age.

CASE REPORT

LBL (RG 2267469J) a 2 years 3 months old boy was admitted to the hospital due to a subacute episode of ataxic gait and hearing deficit for two months. There was no previous remarkable disease. He has healthy parents with no consanguinity or neurological diseases in the family. General physical examination was normal. Neurological examination — Axial cerebellar signs, with ataxic gait, falling frequently; hypoacusia. Laboratory examinations — Computed tomographic (TC) scan was normal. Cerebrospinal fluid (CSF): 0 cells/mm³, a total proteins 22 mg%, glucose 52 mg%; VDRL, Wassermann and Weinberg

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were nonreactive. CSF protein electrophoresis showed a slight gamma globulins increase of 15.4% (normal 7 to 14%); no oligoclonal banding was present. The boy was discharged with the diagnostic hypothesis of cerebellar syndrome and hypoacusia of undetermined etiology. No specific treatment was given; his cerebellar signs normalized over a two month period while a moderate improvement in hypoacusia occurred over a 7 month period.

At the age of 5 years 1 month he was rehospitalized with the onset of acute gaze deviation to the right, starting two weeks previously. Neurological examination — Apendicular ataxia greater on the right side; tendency to a tonic gaze deviation to the right, with unimpaired fixation and tracking capacities; ocular fundi and optic discs were normal. Laboratory examination — CT showed a low-density lesion in the white matter adjacent to the right frontal horn with no mass effect and no contrast enhancement after intravenous contrast medium. Audiometry confirmed bilateral hypoacusia. CSF: 9 cells/mm³ (60% lymphocytes, 30% reticulomonocytes, 5% neutrophils, 2% plasmocytes, 3% macrophages), total proteins 18 mg%, glucose 58 mg%; VDRL, Wassermann and Weinberg were nonreactive. CSF gamma globulins were slightly increased (14.5%) and no oligoclonal banding was present. The CSF T lymphocyte subsets study showed a decrease in the T-active subset and an increase in the T-sensitized subset which suggests local lymphocyte signaling in the CSF system (10,11). Electroencephalogram was normal. Somatosensory evoked potential (SEP) was normal; pattern-reversal visual evoked potential (VEP) revealed right optic nerve impairment. The brain stem auditory evoked potential was suggestive of a bilateral impairment of the auditory pathway at the pontomesencephalic level. The child was treated with prednisone 1 mg/kg/day with remission of the oculomotor deficit over a two month period.

COMMENTS

At the first occurrence, clinical diagnosis of MS is generally impossible to establish as there are no specific signs, symptoms or laboratory tests¹⁴. Ghezzi and Manara⁷ reported 58 childhood cases of MS; Gall et al.⁶ described 40 children and adolescents with MS, reviewed from 1920 to 1952; Bye et al.⁴ reported 5 cases in childhood, most of these patients^{4,6,7} with first episode studied retrospectively. The case discussed here underwent clinical and laboratorial evaluation in both episodes. Initial bouts in childhood^{6,11,17} are usually accompanied by visual or brainstem impairment followed, in order of frequency, by hemiparesis, paraparesis, ataxia and diffuse encephalopathy. Our patient had predominantly ataxia and brainstem signs while visual impairment was not clinically detectable but revealed by VEP. The course of the disease in children^{2,8,9} is mainly of the relapsing-remitting type as in adults. An initially progressive course occurs in only 22%⁵ while complete recovery after the first attack is found in 68%⁵. In the patient discussed here, the two episodes with a remissive course supports the diagnosis of MS. Onset before the age of 10 years occurred in 7 of Gall et al's.⁶ 40 cases with none before 7 years old. In the large series reported by Ghezzi and Manara⁷, MS was not encountered prior to 6 years of age; in the 125 cases of MS prior to 16 years of age analysed by Duquette et al.⁵, 5 years was the earliest age at onset. Besides the case discussed here, the onset of MS at 2 years of age has been described by Bejar et al.¹, Brandt and Ziegler², and Hauser and Bresnan⁸. Brandt et al.² mentioned another case with onset at the same age, with clinical evaluation and necropsy by Nobel, in 1912.

The diagnostic difficulties are well exemplified by our patient who at the first bout was suspected of having a posterior fossa tumour ruled out by CT and the clinical course. After the first hospitalization, the etiology of his cerebellar signs and hypoacusia remained unclear. During the second episode, a relapsing-remitting course was established, MS was suspected and supported by laboratory findings. CSF gamma globulins levels are elevated in 50 to 65% of MS patients and in the case discussed here, a mild increase was detected. Lymphocyte subsets with a decrease in T-active cells and increase in T-sensitized cells were present in our case which is the most frequent pattern in the chronic, repetitive, inflammatory model of which MS is a typical example, as pointed out by Livramento et al.¹⁰. VEP demonstrated unilateral but otherwise undetected optic nerve impairment in the patient here described which occurs in 14% of children with MS¹⁶. VEP and SEP are established as superior to clinical routine evaluation in demonstrating lesions in these pathways, in MS³.

The clinical course, with two distinct episodes and remissions, the two lesions, detected by history and clinical examination, as well as other white-matter and optic

nerve lesions evidenced by CT and VEP supported by CSF pattern suggest the diagnosis⁹ of MS despite the early age of onset. These characteristics fulfill all the necessary criteria of Schumacher et al.¹⁶ with the exception of the precipitate age of onset. For this same reason, the case here reported does not fit into the 'definite MS' category using the guidelines of Rose et al.¹⁵ but would be better considered as 'probable MS'.

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