

# MULTIPLE SCLEROSIS OR MULTIPHASIC DISSEMINATED ENCEPHALOMYELITIS?

## A NEW QUESTION ABOUT AN OLD PROBLEM

### CASE REPORT

PÉRCILES A. MARANHÃO-FILHO\*

---

**ABSTRACT** - A 42 year-old woman developed paraplegia that resolved in six months, followed by sudden right hemiparesis and dysphasia two years later. The clinical work-up, including CT and MR scans, visual evoked potentials, CSF examination and cerebral biopsy suggested the possibility of either multiple sclerosis or multiphasic disseminated encephalomyelitis. The differential diagnosis between both conditions is discussed.

**KEY WORDS:** multiphasic disseminated encephalomyelitis, multiple sclerosis, magnetic resonance imaging.

**Esclerose múltipla ou encefalomielite disseminada multifásica? uma nova questão sobre um antigo problema: relato de caso**

**RESUMO** - Relato de caso de uma mulher com 42 anos de idade, que apresentou paraplegia, com resolução em seis meses seguida por hemiparesia direita e disfasia de início súbito. Sua história, os aspectos da tomografia computadorizada e da ressonância magnética do crânio, os resultados do potencial evocado visual, do exame do líquido cérebrospinal, da biópsia cerebral e da ressonância magnética de controle, sugeriram as possibilidades diagnósticas de esclerose múltipla ou encefalomielite disseminada multifásica. O diagnóstico diferencial entre ambas é discutido.

**PALAVRAS-CHAVE:** encefalomielite disseminada multifásica, esclerose múltipla, ressonância magnética.

---

Multiple sclerosis (MS) is a chronic inflammatory disease of the central nervous system (CNS) with variable clinical manifestations. Pathological and immunological features indicate an autoimmune attack directed towards the myelin<sup>17</sup>. While MS is nosologically well delineated, diseases in which large and confluent territories of the cerebral white matter become demyelinated correspond to a less understood group<sup>5</sup>. Few conditions have caused more confusion at bedside and in the neurologic literature than disseminated vasculomyelinopathy<sup>13</sup>, which includes the disseminated encephalomyelitis (DEM).

The present report discuss on a case of a 42-year-old woman who had paraplegia followed by right hemiparesia two years later. Both episodes recovered completely with steroids. The investigation that included CT and MR scans, visual evoked potentials, CSF examination and a cerebral biopsy favored the diagnoses of MS or multiphasic disseminated encephalomyelitis (MDEM).

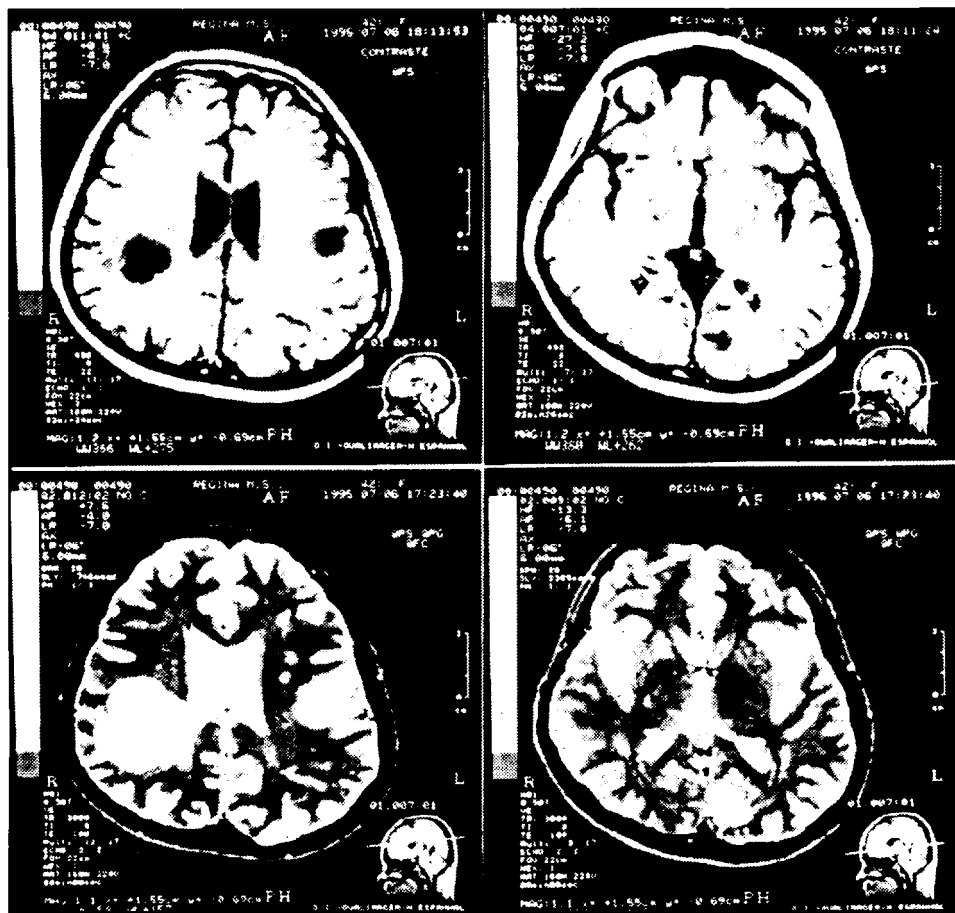
---

\*Neurologist, Department of Neurosurgery, National Institute of Cancer (INCa).Rio de Janeiro, Brazil.  
Aceite: 18-março-1996.

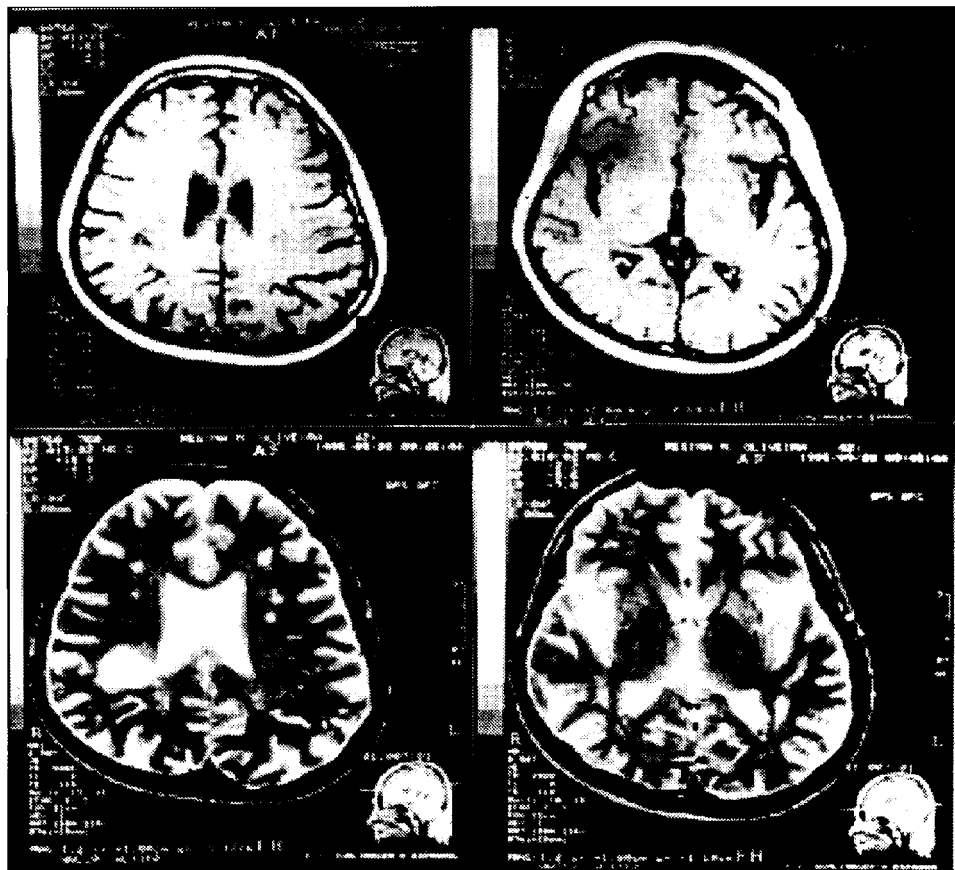
## CASE REPORT

A 42 year-old right handed white woman was seen as an outpatient in July 1995 because of right hemiparesis and dysphasia for 15 days. Two years earlier she suddenly presented paraplegia together with hypoesthesia up to D4 level, bladder and retal dysfunction. The diagnosis of transverse myelitis was given elsewhere and she was treated with oral corticosteroids. She became able to walk with assistance within two months. Six months later the patient became fully recovered except for urinary retention. For the last six months she noticed intermittent diplopia and one week earlier she had developed paroxysm of vertigo and "Jabs-like" headache. A right hemiparesis with hyperreflexia and Babinski sign were present, as well as motor dysphasia and marked pallor of both optic discs. Systemic examination was normal. She reported hemorrhoids and an uterine myoma surgery fifteen years earlier. Her medical history was otherwise unremarkable. There was no history of previous infection or recent immunization against rabies, measles or influenza.

The routine laboratory screening was normal. A CT brain scan and a RMI (Fig 1) showed ring-like enhancing lesions with oedema in the right and left fronto-parietal lobes, and in the left occipital lobe. Visual evoked potentials and the CSF were normal, without oligoclonal bands or IgG increase. A CSF anti-HIV test was negative.



**Fig 1.** MRI scan sequence. T1-weighted image after gadolinium injection showing: (a) two ring-like enhancing lesions in the right and left fronto-parietal subcortical white matter; (b) another small lesion with the same aspect in the left occipital lobe. T2-weighted scan showing: (c) the same largest lesions bilaterally, without mass effect and multifocal small lesions with high signal intensity in the subcortical white matter mostly in the left side; (d) high signal lesion in the left occipital lobe.



*Fig 2. RMI scan sequence two-and-a-half months after the first MRI scan. T1-weighted scan after gadolinium administration showing: (a) the two lesions previously seen practically disappeared as well as the occipital one (b) T2-weighted image showing: (c) the largest lesion with high signal on the left side disappeared and the other in right side shrank. There were multifocal periventricular white matter lesions bilaterally. (d); the high signal lesion of the left occipital lobe disappeared.*

A needle biopsy of the right fronto-parietal lobe lesion was carried out showing inflammatory primary demyelination without evidence of malignancy.

She was given oral dexametazone 4 mg qid. One-and-a-half month later speech and strength were normal. A control MR taken 2.5 months after the first MR, showed great improvement (Fig 2). There has been a considerable reduction of the right side lesion while the two largest lesions on the left side almost disappeared.

## DISCUSSION

Enhancing lesions with mass effect have been associated with MS<sup>1,6,7,10</sup>. As far as the MR is concerned, the present patient fulfills the criteria for the "strongly suggestive of MS" category<sup>11</sup> with three lesions hyperintense in T2 longer than 3.0 mm, one of which being periventricular. She also fulfills the criteria for clinically definite MS (CDMS)<sup>14</sup> as there were two attacks, clinical and paraclinical evidence supporting two separate lesions. Besides, the two attacks involved different parts of the CNS, occurred more than a month apart and lasted more than 24 hours each. It is generally recognized that the diagnosis of CDMS has an accuracy of 90 to 95%<sup>11</sup>.

More recently, Poser<sup>12</sup> stressed that the diagnosis of MS based on clinical grounds, CSF examination, and biopsy of the lesion without considering important aspects of RM, such as size and topography of the lesion(s), may be wrong. Interestingly, Poser considers that large demyelinating lesions with clinical, radiological and histological features simulating brain tumors considered as "tumoral form of MS"<sup>1-3,6,7,15</sup>, are actually forms (or subforms) of DEM.

Acute transverse myelopathy may occur in both MS and DEM. It is generally accepted that acute transverse myelopathy without associated neurologic signs or symptoms is a manifestation of DEM in 75% to 80% of cases<sup>13</sup>.

Monophasic acute DEM, the so-called ADEM, is not difficult to differentiate from MS since the diagnosis of MS should never be based on a single episode. Poser<sup>12</sup> proposed that DEM may be divided into two types. In the first type (recurrent DEM-RDEM), an initial episode of ADEM is followed by one or more episodes that reproduce all or some of the symptoms of the original attack. This presentation form is rare in MS. In the second type, there are two or more separate clinically different acute episodes (multiphasic-MDEM).

Any MS clinical feature may be present in encephalomyelitis and vice-versa<sup>9</sup>. Therefore, it is impossible to differentiate MDEM from MS on clinical grounds. This differentiation is nevertheless important since the prognosis of MDEM is comparatively better<sup>4</sup>. Both conditions respond to high-potency steroids as well. RMI may distinguish the two subtypes of DEM from MS. Unlike MS, the lesions in DEM are extensive and may involve the cortex. Alternatively, they consist of large globular areas usually located at the posterior angle of the ventricular atrium, with increased signal intensity but no mass effect. The cerebellum, thalamus or the basal ganglia are occasionally involved. The corpus callosum is usually not affected. In control RMI examinations new lesions may be seen in MDEM but they never occur in RDEM. The large lobar high intensity lesions, which may be seen in DEM, are never seen in MS<sup>4,12</sup>.

CSF usually does not help in differentiating DEM from MS, except by the fact that in DEM oligoclonal bands are rare and may disappear. Some authors consider biopsy as an important diagnostic tool<sup>3,6</sup> although some others<sup>4</sup> believe this may lead to erroneous diagnosis, since demyelination in MS and DEM are identical and shows perivascular oedema and inflammation associated with demyelination. Thus, the site of the lesion becomes the most important differentiating feature<sup>12</sup>.

An acute encephalitic, myelitic, or encephalomyelitic process may occasionally occur without any previous illness or vaccination. Miller et al.<sup>8</sup>, using only clinical data, reported nine examples of MS with onset or exacerbation following vaccination or inoculation against smallpox, rabies, typhoid fever, tuberculosis and tetanus. Interestingly, in four of them there were exacerbations of established MS. Kepes<sup>5</sup> studied 31 biopsied patients, suggesting that most large, focal demyelinating lesions of the brain represent an intermediate entity between classical MS and ADEM. Thirteen of their patients were more than 57 years old at disease onset. In three others additional lesions suggested a MS-like evolution, and only one received a flu vaccine 10 days before the onset of her clinical symptoms.

Whether influenza virus vaccine produces a form of ADEM or precipitates attacks of MS is an unsolved question. Until now, it is still far from whether ADEM and MS represent distinct disease entities or simply two different clinical expressions of the same reactive process<sup>16</sup>.

**Acknowledgement** - The author wish to thank Dr. Maurice Vicent and Prof. Mitchel A. Robuck Jr. for manuscript preparation assistance.

## REFERENCES

1. Giang DW, Poduri KR, Eskin TA, Ketonen LM, Friedman PA, Wang DD, Herndon RM. Multiple sclerosis masquerading as a mass lesion. *Neuroradiology* 1992;34:150-154.
2. Gütling E, Landis T. CT ring sign imitating tumor, disclosed as multiple sclerosis by MRI: a case report. *J Neurol Neurosurg Psychiatry* 1989;52:903-906.

3. Hunter SB, Ballinger WE Jr, Rubin JJ. Multiple sclerosis mimicking primary brain tumor. *Arch Pathol Lab Med* 1987;111:464-468.
4. Khan S, Yaqub BA, Poser CM, Al Deeb SM, Bohlega S. Multiphasic disseminated encephalomyelitis presenting as alternating hemiplegia. *J Neurol Neurosurg Psychiatry* 1995;58:467-470.
5. Kepes JJ. Large focal tumor-like demyelinating lesions of the brain: intermediate entity between multiple sclerosis and acute disseminated encephalomyelitis? a study of 31 patients. *Ann Neurol* 1993;33:18-27.
6. Lecky BRF, Jeyagopal N, Smith ETS, Foy PM. Cerebral CT lesions in multiple sclerosis mimicking multiple metastases (Letter). *J Neurol Neurosurg Psychiatry* 1991;54:92.
7. Maranhão-Filho PA, Moraes-Filho L, Camara LSA, Salema CC. Fulminant form of multiple sclerosis simulating brain tumor: a case with parkinsonian features and pathologic study. *Arq Neuropsiquiatr* 1995;53:503-508.
8. Miller H, Cendrowski W, Schapira K. Multiple sclerosis and vaccination. *Br Med J* 1967;2:210-213.
9. Miller H, Schapira K. Aetiological aspects of multiple sclerosis. *Br Med J* 1959;1:811-815.
10. Nesbit GM, Forbes GS, Scheithauer BW, Okazaki H, Rodriguez M. Multiple sclerosis: histopathologic and MR and/or CT correlation in 37 cases at biopsy and three cases at autopsy. *Radiology* 1991;180:467-474.
11. Paty DW, McFarlin DE, McDonald WI. Magnetic resonance imaging and laboratory aids in diagnosis of multiple sclerosis (Editorial) *Ann Neurol* 1991;29:3-5.
12. Poser CM. The epidemiology of multiple sclerosis: a general overview. *Ann Neurol* 1994;36(S2):S180-S193.
13. Poser CM. The diseases of the myelin sheath. In Baker AB (ed). *Clinical neurology*. Philadelphia: Harper & Row, 1981:1-188.
14. Poser CM, Paty DW, Scheinberg L, McDonald I, Davis FA, Ebers GC, Johnson KP, Sibley WA, Silberberg DH, Wallace WT. New diagnostic criteria for multiple sclerosis: guidelines for research protocols. *Ann Neurol* 1983;13:227-231.
15. Tilbery CP, Guidugli-Neto J. Esclerose múltipla simulando tumor cerebral: apresentação de caso com comprovação histopatológica. *Arq Neuropsiquiatr* 1995;53:302-306.
16. Yahr MD, Lobo-Antunes J. Relapsing encephalomyelitis following the use of influenza vaccine. *Arch Neurol* 1972;27:182-183.
17. Zhang J. Multiple sclerosis perspectives on autoimmune pathology and prospects for therapy. In Apple SH (ed). *Current neurology* 15. Chicago: Year Book, 1995:115-155.