Eur Arch Psychiatry Clin Neurosci (1992) 242:69–71

European Archives of Psychiatry and Clinical Neuroscience

Marchiafava-Bignami Disease. A Case Studied by Structural and Functional Brain Imaging

T. Humbert[1], P. De Guilhermier[2], C. Maktouf[3], G. Grasset[3], F. M. Lopez[2], and P. Chabrand[1]

[1]Service de Psychiatrie B1, Hôpital Gaston Doumergue, C.H.U. 30000 Nimes, France
[2]Département d'Imagerie Médicale, Hôpital Gaston Doumergue, C.H.U. 30000 Nimes, France
[3]Service Central de Médecine Nucléaire et de Biophysique Médicale, Hôpital Gaston Doumergue, C.H.U. 30000 Nimes, France

Received August 25, 1992

Summary. Marchiafava-Bignami disease (MBD) is a rare complication of chronic alcoholism. The clinical features, X-ray, CT and MRI findings are well documented. However, functional brain imaging has not been used in cases of MBD. We used single-photon emission computed tomography (SPECT) to monitor the regional cerebral blood flow in a patient suffering from a acute form of MBD, from which he subsequently recovered. Several abnormalities were found. A more frequent use of functional brain imaging in MBD could improve our knowledge of pathogenesis and prognosis for MBD.

Key words: Marchiafava-Bignami disease – CT – MRI – SPECT – Corpus callosum

Introduction

Marchiafava-Bignami disease (MBD) is a rare complication of chronic alcoholism. Its aetiology is completely unknown [12, 13, 15, 17, 19]. The clinical features and neuroradiological (CT and MRI) data are well documented [4, 5, 23]. However, these imaging techniques cannot account for the totality of the symptomatology [3].

There is good evidence that blood flow imaging can be of clinical value. It is a valuable diagnostic tool in the evaluation (functional changes), characterisation, and management of a variety of neuropsychiatric disorders [1, 11, 24]. We therefore performed single-photon emission tomography (SPECT) using ^{99m}Tc-HMPAO in the investigation of a patient suffering from an acute and regressive form of MBD.

Case Report

A 43-year-old man, was admitted to the Psychiatry Department B for psychic symptoms and cachexia on 15 April 1991. He was right-handed and a chronic alcoholic. His general condition improved greatly during the first few days. However, his neuropsychiatric condition remained severe. He suffered from confusion, astasia-abasia, mutism and limb hypertonia, but had no convulsive episodes. The stretch reflexes were rapid and symmetrical, the plantar reflexes were neutral. There were no pyramidal signs. The pupils were equal and reactive and there was no oculomotor paralysis.

On admission, his serum potassium level was 2.5 mmol/l, GOT 48 U/l, GPT 134 U/l and γGT 179 U/l; prothrombin rate 80%. All other laboratory studies, including vitamin B12, folic acid and HIV ELISA, gave normal or negative results.

The EEG revealed a symmetrical, reactive sub-alpha rhythm of 7 Hz, with a good visual arrest reaction. These clinical features and the lack of signs indicating other types of alcoholic encephalopathies suggested that the patient was suffering from MBD. This diagnosis was confirmed by a cerebral scan 19 April. It showed a hypodensity extending over the whole of the corpus callosum. The ventricular system was of normal size with discrete diffuse cortico-subcortical atrophy. After feeding and treatment with vitamin B1, PP and small doses of benzodiazepines, the patient's behaviour improved. MRI was therefore performed on 5 June. It clearly demonstrated central zones of liquefaction necrosis stretching over the entire corpus callosum (Fig. 1), but without other lesions. Electrophysiological studies were all pathological (particularly BAEPs and VEPs). Doppler-sonographic measurements of blood flow through the internal and external carotic arteries, the anterior and medial cerebral arteries on both sides and the basilar artery were normal.

The patient was examined on 13 June (see Fig. 2) and 4 July. The 4 July examination was carried out 8 days after the patient had been placed on amineptine (200 mg/day). Walking, although ataxic, was possible. This was not the case on 13 June. However, there was much less progress in the phasic disorders. The two SPECT examinations showed biparietal hypofixation and distinct

Correspondence to: T. Humbert

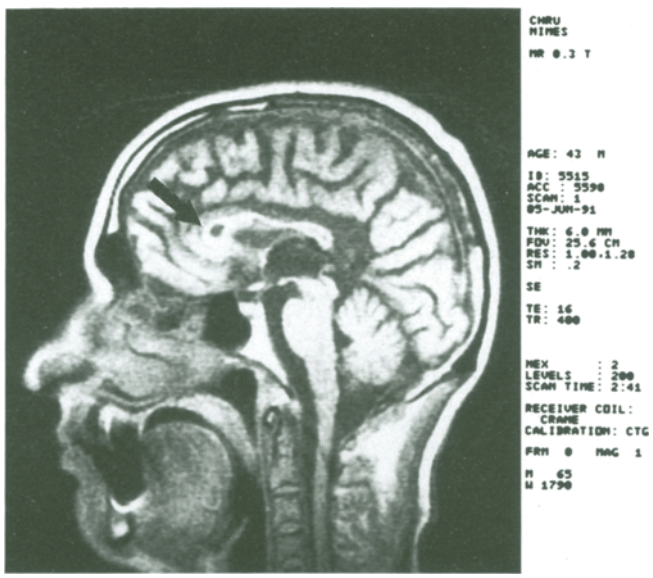

Fig. 1. MRI performed 6 weeks after the first clinical signs, without contrast medium. T1-weighted sagittal SE images with TE = 16 ms and TR = 400 ms showing poorly defined areas of central necrosis in the corpus callosum (homogeneous hyposignal in T1 similar to that of the CSF and homogeneous hypersignal in T2). MRI specifications: Bo = 0.3 Tesla, Matrix 256, SE = TE: 16 ms, TR: 400 ms

hypofixation of grey matter in the region of the left lobulus paracentralis. This latter hypofixation was less extensive during the second examination.

The neurological examination carried out 3 months after the patient's admission was still disturbed by Broca's aphasia. However, a left unilateral ideomotor apraxia was clearly observed as well as agraphia.

Discussion

The clinical and neuroradiological findings are consistent with the diagnosis of MBD. The interhemispheric disconnection syndrome signs have rarely been described [2, 16]. There are a few reports of a favourable outcome for acute, often necrotic forms of MBD. All the established MRI appearances have shown *partial* lesions of the corpus callosum limited *to this latter* area [3, 8, 22]. In our patient, the lesions (shown by CT and MRI) were limited to the corpus callosum, but this body was *entirely* necrosed; Therefore, the prognosis was *relatively* favourable.

While the diagnosis and treatment of a case of MBD is not novel, the use of functional brain imaging is. To date there are no reports of cerebral blood flow (CBF) studies in MBD, although positron emission tomography (PET) and SPECT have been widely used to examine many neuropsychiatric disorders [1].

The two most commonly used rCBF tracers are ^{123}I-labeled n-isopropyliodo-amphetamine (IMP) and ^{99m}Tc-HMPAO. Both of these agents provide data that are generally in agreement with the metabolic data obtained by PET.

We first examined the SPECT findings for their reliability and specificity. Studies on normal controls and

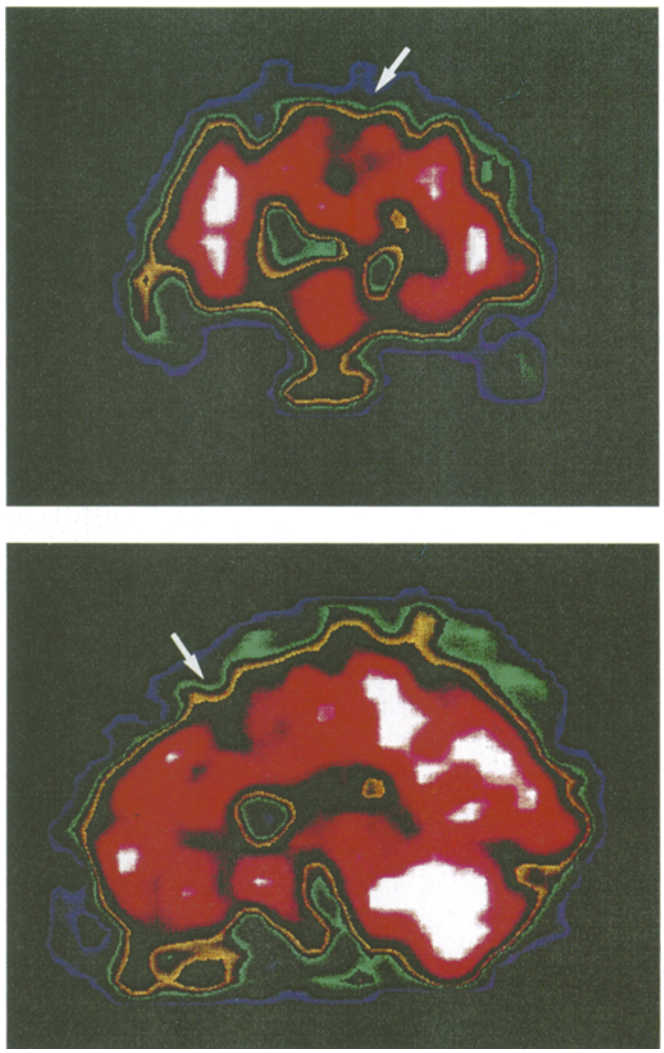

Fig. 2a and b. SPECT examination on 13 June 1991 showing focal hypometabolism in the region of the left lobulus paracentralis: frontal section (**a**) and parasagittal section (**b**): at the lobulus paracentralis level. In the supine position, the patient was injected intravenously with 20 m Ci of freshly prepared ^{m99}Tc HMPAO and imaged 30 min later with a GENESYS ADAC gamma camera interfaced with a PEGASIS ADAC computer. Acquisition parameters: 64 projections. 64 × 64 matrix. 25 s per projection. Slice thickness: one pixel or 5 mm

patient populations indicate that auditory, visual and cognitive stimulation, as well as anxiety, produce patterns of rCBF different from those of the resting state [20, 24, 26, 27]. We therefore confirmed the first focal abnormalities with another SPECT scan performed 3 weeks later. Images were also reconstructed in three planes (transaxial, sagittal and coronal) with 39 slices in each plane. The images were interpreted by two nuclear medicine physicians who had no clinical data. Decreased signal areas were indentified only if they were seen on 3 or more contiguous slices.

Some abnormalities are difficult to interpret:

– The left functional abnormalities (lobulus paracentralis) could be produced by an acute distant lesion, in this case in the corpus callosum and resulting from MBD.

MRI performed 2 months later (August) confirmed that there was no structural modification of this left fronto-parietal region (cortical and subcortical white matter). In the present case, this interpretation, which suggests diaschisis [10, 14, 18, 21], could also explain such neuropsychological disorders as aphasia. Nevertheless, this hypothesis, based on a single case report, must remain speculative.

– Another explanation seems much more plausible, because of the neuropathological changes that occur in chronic alcoholism, and because of the recent functional brain imaging studies performed on alcoholic patients. Alcohol has diffuse effects, on the corpus callosum and the central hemispheric white matter, and also on layers 3 and 5 of the cerebral cortex, and such microscopic lesions have been seen in MBD [6, 7, 9, 25]. Moreover, the combination of MBD and Wernicke's encephalopathy, for instance, occurs in about 15–20% of cases [3].

Several authors have found an alteration in the cerebral metabolism of alcoholic patients, although there is little agreement as to the location. Wik [28] found that the parietal cortical areas were most affected, while Eckardt [11] found a mediofrontal hypometabolism.

Finally, more frequent use of functional brain imaging in MBD could improve our knowledge about pathogenesis and prognosis of this rare complication of chronic alcoholism.

Acknowledgements. The authors thank Professor JP Bureau for help and advice, and Mr. R. Hunzinger for technical assistance.

References

1. Alavi A, Hirsch LJ (1991) Studies of central nervous system disorders with single-photon emission computed tomography and positron emission tomography: evolution over the past 2 decades. Sem Nucl Med 21:58–81
2. Barbizet J, Degos JD, Lejeune A, Leroy A (1978) Syndrome de disconnexion interhémisphérique avec dyspraxie diagnostique au cours d'une maladie de Marchiafava-Bignami. Rev Neurol (Paris) 134:781–789
3. Baron R, Heuser K, Marioth G (1989) Marchiafava-Bignami disease with recovery diagnosed by CT and MRI: demyelinisation affects several CNS structures. J Neurol 236:364–366
4. Bourrat C, Tomassi M, Bochu M, Kopp N, Malsch S (1984) X-ray scanning in Marchiafava-Bignami disease, Rev Neurol (Paris) 140:426–431
5. Bracard S, Claude D, Vespignani H, Almeras M, Carsin M, Lambert H, Picard L (1986) Computerized tomography and MRI in Marchiafava-Bignami disease. J Neuroradiol 13:87–94
6. Brion S, Marchiafava-Bignami syndrome. In: Vinken PJ, Bruyn GW (eds) Handbook of clinical neurology, vol 28. North Holland, Amsterdam, pp 317–329
7. Castaigne P, Buge A, Escourolle R, Rancurel G (1971) La maladie de Marchiafava-Bignami. Etude anatomo-clinique de dix observations. Rev Neurol (Paris) 125:179–196
8. Delangre T, Hannequin D, Clavier E, Denis P, Mihout B, Samson M (1986) Maladie de Marchiafava-Bignami d'évolution favorable. Rev Neurol (Paris) 142:933–936
9. Delay J, Brion S, Escourolle R, Sanchez A (1959) Rapports entre la dégénérescence du corps calleux de Marchiafava-Bignami et la sclérose laminaire corticale de Morel (à propos de 5 observations anatomo-cliniques). Encéphale 4:281–311
10. D'Antona R, Baron JC, Samson Y, Serdaru M, Viader F, Agid Y, Cambier J (1985) Subcortical dementia. Frontal cortex hypometabolism detected by positron tomography in patients with progressive supranuclear paralysis. Brain 108:785–799
11. Eckardt MJ, Rohrbaugh JW, Rio De, Martin RP (1990) Positron emission tomography as a technique for studying the chronic effects of alcohol on the human brain. Ann Med 22:341–345
12. Heepe P, Nemeth L, Brune F, Grant JW, Kleihues P (1988) Marchiafava-Bignami Disease. A Correlative Computed Tomography and Morphological study. Eur Arch Psychiatriy Neurol Sci 237:74–79
13. Ironside R, Bosanquet FD, McMenemey WH (1961) Central demyelination of the corpus callosum (Marchiafava-Bignami disease). Brain 84:212–230
14. Kempinsky WH (1958) Experimental study of distant effects of acute focal brain injury. A study of diaschisis. Arch Neurol Psychiatry 79:376–389
15. Laplane D, Morel-Maroger A (1973) Complications nerveuses de l'alcoolisme. Encycl Med Chir (Paris) Neurologie 17045 g–10
16. Lechevallier B, Andersson JC, Morin P (1977) Hemispheric disconnection syndrome with a 'crossed avoiding' reaction in a case of Marchiafava-Bignami disease. J Neurol Neurosurg Psychiatry 40, 483–497
17. Lopez-Arlandis JM, Burquera JA, Hernandez M, Garcia F, Coret F, Catala J (1990) Hallazgos de la tomografia computerizada en la forma aguda de la enfermedad de Marchiafava-Bignami. Neurologia 5, 62–4
18. Luaute JP, Bidault E, Sanabria E (1990) Etat athymhormique après accident vasculaire cérébral du postpartum. Apport de l'imagerie cérébrale. Ann Médico Psychol (Paris) 532–538
19. Marchiafava E, Bignami A (1903) Sopra un alterzione del corpo calloso osservata in sogetti alcoolisti. Riv Patol Nerv Ment 8, 544–549
20. Mathew RJ, Wilson WH (1990) Anxiety and Cerebral Blood Flow. Am J Psychiatry 147, 7 838–849
21. Meyer JS, Shinohara Y, Kanda T, Fukuuchi Y, Ericsson AD, Kok NK. Diaschisis resulting from acute unilateral cerebral infarction. Arch Neurol, 1970, 23, 241–247
22. Meyrignac C, Bokobza M, Cesaro P, Degos JD (1987) Contribution de l'imagerie par résonnance magnétique au diagnostic de la maladie de Marchiafava-Bignami. Presse Médicale 16, 37, 1863–1864
23. Shiota J, Kawamura M, Hirayama K, Isono O, Makit K (1989) Antemortem diagnosis of Marchiafava-Bignami disease. Clin Neurol 29, 701–706
24. Tikofsky RS, Heliman RS (1991) Brain single-emission computed tomography: never activation and intervention studies. Sem Nucl Med XXI: 40–57
25. Torvik A, Lindboe CF, Rodge S (1982) Brain lesions in alcoholics. A neuropathological study with clinical correlations. J Neurol Sci 56:233–248
26. Van Heertum RL, O'Connell RA (1991) Functional brain imaging in the evaluation of psychiatric illness. Sem Nucl Med 2:24–39
27. Weinberger DR, Berman KF (1988) Speculation on the Meaning of cerebral metabolic hypofrontality in schizophrenia. Schizophr Bull 14:157–167
28. Wik G, Borg S, Sjorgen L (1988) Positron emission tomography determination of regional cerebral glucose metabolism in alcohol-dependent men and healthy controls using ^{11}C-glucose. Acta Psychiatr Scand 78:234–241