Neuroradiology (1992) 34: 480–482

Neuro radiology

Marchiafava-Bignami disease: serial changes in corpus callosum on MRI*

K. H. Chang[1], S. H. Cha[1], M. H. Han[1], S. H. Park[2], D. L. Nah[2], and J. H. Hong[3]

[1] Department of Radiology, Seoul National University College of Medicine, Seoul, Korea
[2] Department of Neurology, Youngdeungpo City Hospital, Seoul, Korea
[3] Department of Radiology, Sungae Hospital, Seoul, Korea

Summary. Serial MRI findings of changes in corpus callosum lesions in two cases of Marchiafava-Bignami disease are presented. In both, MRI displayed diffuse swelling of the corpus callosum in the acute stage, thought to represent oedema and demyelination. In the chronic stage, in addition to atrophy of the corpus callosum with presumed focal necrosis, previously undescribed focal hypointensity on T2-weighted images, of unknown cause, was observed in the corpus callosum.

Key words: Degenerative disease – Magnetic resonance imaging – Corpus callosum – Marchiafava-Bignami disease

Marchiafava-Bignami (MB) disease is a rare neurological complication of chronic alcoholism, characterized by degeneration of the corpus callosum [1–5]. There are several reports on the CT and MRI findings [7–10]. To our knowledge, however, serial changes of the corpus callosum lesion on MR imaging have never been described. We present two cases of MB disease studied by MRI from acute to recovery stages.

Methods

Three MRI studies were carried out in each of two patients on a 0.5 T superconducting unit, except for the second study on case 1, performed on a 2.0 T superconducting unit. All images were obtained with spin echo pulse sequences; T1-weighted (T1WI) (500/30, TR/TE) sagittal, and proton-density-weighted (PDWI) (2000–3000/30) and T2-weighted (T2WI) (2000–3000/80–100) axial images were obtained in every study. Sagittal T2WI were sometimes obtained. The slice thickness/gap was 6–7 mm/2 mm for the 0.5 T unit and 5 mm/2 mm for the 2.0 T unit. The number of excitations ranged from two to four for T1WI on both 0.5 T and 2.0 T units, and was two for T2WI on the 0.5 T unit and one for T2WI on the 2.0 T unit. The acquisition matrix was 256 × 256 with spatial resolution of 1 mm × 1 mm.

* This work was supported by a grant from the Korea Radiologic Research Fund (1992)

Case reports

Case 1

A 56-year-old man with a long history of excessive alcoholic intake was admitted because of disorientation, progressive disturbance of consciousness and seizures. He complained of dysarthria and walking difficulty for 4 days before admission. Neurological examination showed no lateralizing signs. Blood and CSF studies were within normal limits. MRI imaging on admission demonstrated diffuse swelling of the entire corpus callosum, which was isointense with brain on T1WI (Fig. 1 a) and hyperintense on T2WI (Fig. 1 b). He recovered from stupour in a week, and showed signs of interhemispheric disconnection. On MRI obtained on a 2.0 T unit on the 11th hospital day, the corpus callosum had decreased slightly in both size and signal intensity, and there were focal areas of hypointensity in the genu and splenium on T2WI (Fig. 1 c). MRI on the 54th day showed severe atrophy of the corpus callosum with multiple focal areas of presumed necrosis in the genu, body and splenium (Fig. 1 d); multiple areas of marked hypointensity on T2WI were seen in these areas (Fig. 1 e, f). The patient was discharged in a static condition.

Case 2

A 45-year-old man with a long history of alcoholism presented with dysarthria, dysphagia and gait disturbance for 6 days prior to admission. Examination on admission revealed features of interhemispheric disconnection which suggested a lesion of the corpus callosum. Blood and CSF examinations showed no abnormality. MRI on admission showed diffuse enlargement and high signal intensity throughout the corpus callosum on both PDWI and T2WI. During the next week the patient gradually improved with supportive treatment. MRI on the 7th hospital day showed diminished signal intensity in the corpus callosum, with multiple residual foci of high signal intensity on T2WI (Fig. 2 a). Within a month the patient almost recovered from his neurological deficits, except for mild memory disturbance. MRI obtained on the 74th day demonstrated slight atrophy of the corpus callosum with a small area of presumed cystic necrosis in the splenium (Fig. 2 b, c) and with small foci of hypointensity in the genu and splenium on T2WI.

Discussion

MB disease was originally described in poorly nourished Italian males addicted to consumption of crude red wine. Subsequently, the disorder was reported in other popula-

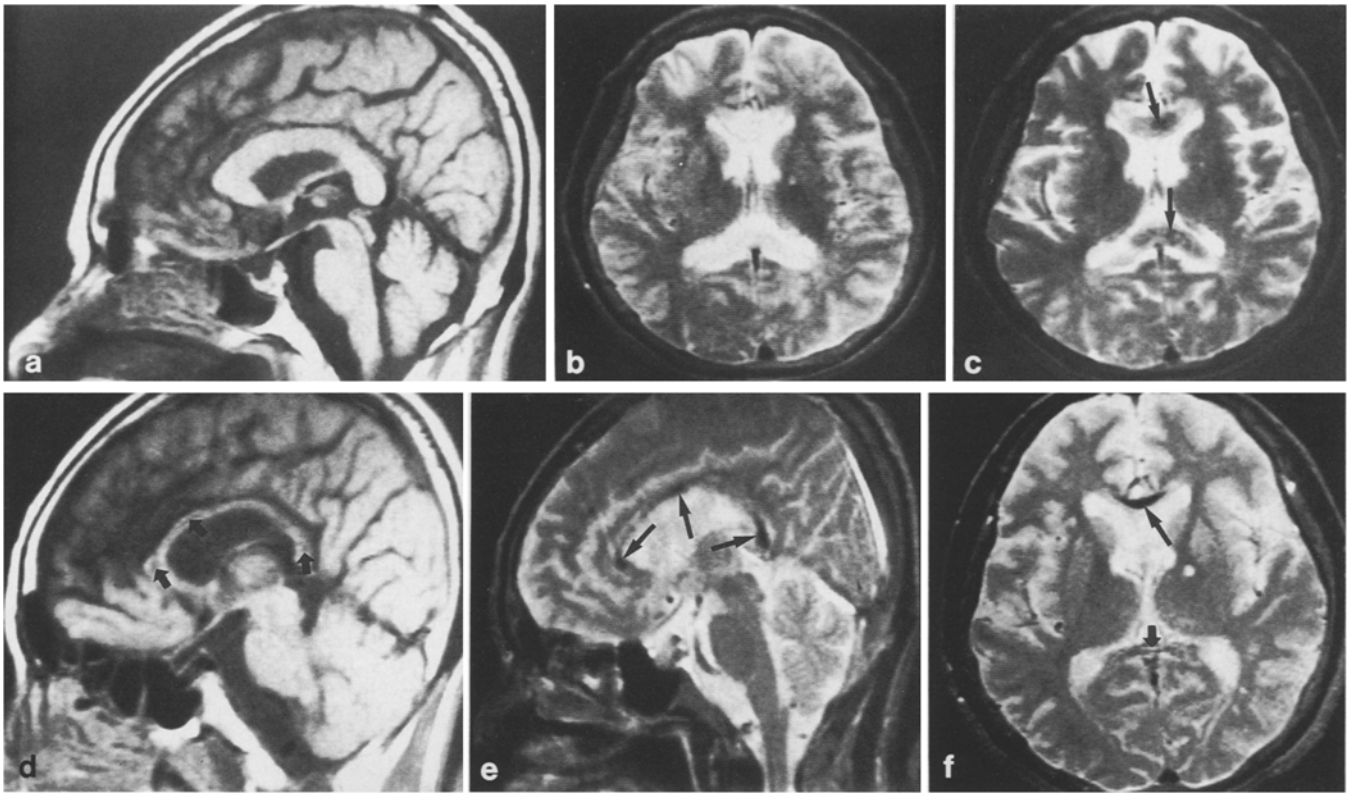

Fig. 1 a–f. Case 1. **a, b** Sagittal T1-weighted (500/30) and axial T2-weighted (2500/100) MRI on admission shows diffuse swelling of entire corpus callosum with high signal intensity in **b.** **c** Axial T2-weighted image (2500/100) on a 2.0 T unit 11 days later demonstrates generalized reduction of high signal from the corpus callosum, with areas of focal hypointensity in the genu and splenium *(arrows)*. **d–f** MRI on the 54th hospital day. Sagittal T1-weighted image (500/30) (**d**) reveals severe atrophy of entire corpus callosum with multiple areas of low intensity suggesting cystic necrosis in the genu, body and splenium *(arrows)*. Sagittal (**e**) and axial (**f**) T2-weighted (2500/100) images show multiple focal areas of marked hypointensity *(long arrows)* in the genu, body and splenium. A small transverse linear focus of hyperintensity *(short arrow)* in centre of the splenium (**f**), suggesting focal cystic necrosis. A small lacune is seen in the left globus pallidus presumably, unrelated to Marchiafava-Bignami disease

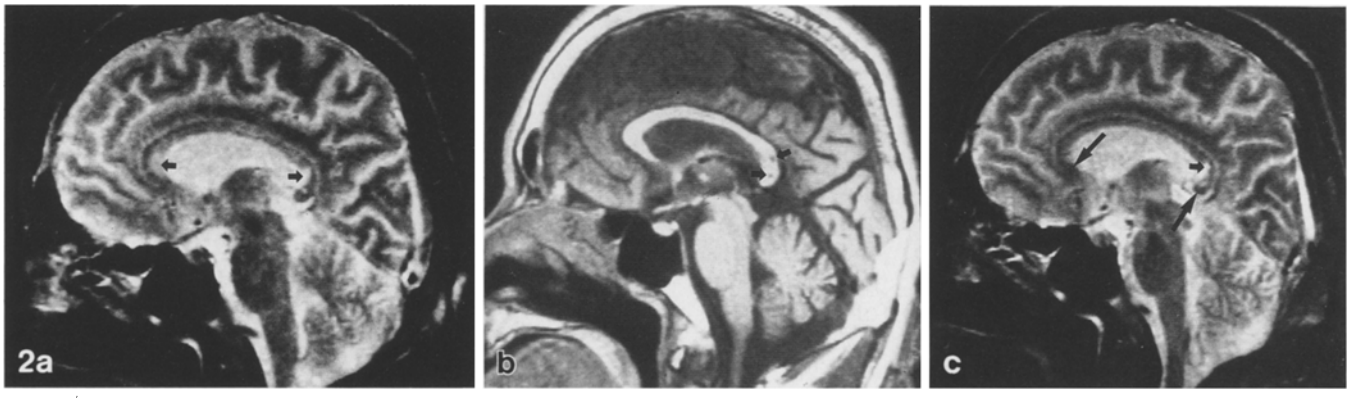

Fig. 2 a–c. Case 2. MRI on admission demonstrated diffuse swelling of entire corpus callosum (not shown). **a** Sagittal T2-weighted image (2500/100) 7 days after admission shows multiple foci of high intensity *(arrows)* in the genu and splenium. Sagittal T1-weighted (500/30) (**b**) and T2-weighted (2500/90) images (**c**) on 74th day reveal focal areas of presumed cystic necrosis in the splenium *(short arrows)* associated with small foci of hypointensity *(long arrows)* in the genu and splenium on T2-weighted image

tions with different preferences in alcoholic beverages [1]. It is not believed to be inherited; chronic alcoholism seems to be the principal aetiological consideration, although toxic factors are also occasionally implicated [5].

The main pathological findings consist of demyelination and necrosis of the corpus callosum, although cerebral hemispheric white matter and other interhemispheric commissural fibres may be involved. The lesion is most extensive in the central part of the corpus callosum, with relative sparing of the dorsal and ventral layers. The focal cystic necrosis is usually observed in the central parts of the genu, body and/or splenium [1–5]. Microscopically, the lesions display predominant demyelination with relative sparing of axis cylinders. Oligodendrocytes are

markedly reduced in number while lipid-laden macrophages are abundant [1]. The macrophages may be filled with scattered or clustered haemosiderin [3]. There may be proliferation and fibrous sclerosis of blood vessels within the lesions [2]. The exact pathogenesis is still uncertain.

The disease may present as two major clinical forms [10]: an acute form with severe disorders of consciousness and massive neurological disturbances, often resulting in death, and a chronic form, with interhemispheric disconnection, which may last several years [3, 6]. An intermediate form of acute onset of neurological disturbances followed by regression to the chronic form may also be observed, as in our cases [9, 10]. Though the corpus callosum lesion in MB disease causes the characteristic interhemispheric disconnection syndrome, correct diagnosis has not been easy, often being made only at autopsy, because of the complex clinical and neurological manifestations, particularly in the acute phase [3, 4, 6].

With modern imaging modalities such as CT and MRI, the disease can be more readily diagnosed in life. On CT the lesions of demyelination and necrosis are seen as hypodensity of the corpus callosum, particularly the genu and splenium [7–10]. However, small lesions in the body may not be visible on axial CT. They can be demonstrated far better on MRI. To our knowledge, MR findings in MB disease have been reported previously in only five cases, all studied in the chronic stage, more than 3 months after the onset of symptoms [7–10]. In this stage, MRI clearly shows diffuse atrophy of the corpus callosum, and focal necrosis, shown as hypointensity on T1WI and hyperintensity on T2WI, usually confined to the genu, body and/or splenium. These focal lesions are best demonstrated on sagittal T1WI or T2WI [7–10]. In the acute phase, demyelination and oedema of the corpus callosum are represented by diffuse enlargement, with hyperintensity on PDWI and T2WI and as isointensity or slight hypointensity on T1WI, as in our case 1.

The foci of hypointensity on T2WI in the corpus callosum, seen in our cases, have not previously been described. Their exact cause is unknown. They might be related to T2-shortening caused by haemosiderin deposition, abundant collections of lipid-laden macrophages, or both. It is known that some macrophages produce paramagnetic free radicals [11], presumably during active phagocytosis of lipid debris liberated because of the demyelinating process; these free radicals would be intracellular and therefore capable of producing T2-shortening. However, the susceptibility effect of T2-shortening was not confirmed by using a gradient echo technique in the present cases. Possible loss of mobile protons in the demyelinated fibres and fibrous sclerosis of blood vessels within the lesions [2] might also be contributable to hypointensity of the corpus callosum. Further investigation by MRI and pathological studies is needed to elucidate this.

Differentiation of MB disease from infarction of the corpus callosum may be difficult. However, selective involvement of the entire length of the corpus callosum (corresponding to the territories of both anterior and posterior cerebral arteries) and focal cystic necrosis confined to its central layer, with sparing of the ventral and dorsal layers, appear more likely to be due to MB disease. The combination of chronic alcoholism, the other clinical features and MRI findings facilitates the correct diagnosis.

References

1. Schochet SS (1985) Exogenous toxic-metabolic disease including vitamin deficiency. In: Davis RL, Robertson DM (eds) Textbook of neuropathology. Williams & Wilkins, Baltimore, pp 372–402
2. Smith WT (1976) Intoxications, poisons and related metabolic disorders. In: Blackwood W, Corsellis JAN (eds) Greenfield's neuropathology, 3rd edn. Arnold, London, pp 149–150
3. Lechevalier 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
4. Koeppen AH, Barron KD (1978) Marchiafava-Bignami disease. Neurology 28: 290–294
5. Ghatak NR, Hadfield MG, Rosenblum WI (1978) Association of central pontine myelinolysis and Marchiafava-Bignami disease. Neurology 28: 1295–1298
6. Lhermitte F, Marteau R, Serdaru M, Chedru F (1977) Signs of inter-hemispheric disconnection in Marchiafava-Bignami disease. Arch Neurol 34: 254
7. Kawamura M, Shiota J, Yagishita T, Hirayama K (1985) Marchiafava-Bignami disease; computed tomographic scan and magnetic resonance imaging. Ann Neurol 18: 103–104
8. Clavier E, Thiébot J, Delangre T, Hannequin D, Samson M, Benozio M (1986) Marchiafava-Bignami disease. A case studied by CT and MR imaging. Neuroradiology 28: 376
9. 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
10. Mayer JM, De Liege P, Netter JM, Danze F, Reizine D (1987) Computerized tomography and nuclear magnetic resonance imaging in Marchiafava-Bignami disease. J Neuroradiol 14: 152–158
11. Sze G, Zimmerman RD (1988) Magnetic resonance imaging of infections and inflammatory diseases. Radiol Clin North Am 26: 839–859

K. H. Chang, M. D.
Department of Diagnostic Radiology
Seoul National University Hospital
28 Yongon-dong, Chongno-gu
Seoul 110-744, Korea