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Marchiafava-Bignami Disease

First Case in Germany

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Summary. The first case of Marchiafava-Bignami disease in a German is reported. The female patient was a heavy drinker and died after a fortnight of coma. Necropsy showed a typical degeneration of the corpus callosum and of the subcortical white matter of the right cerebral hemisphere. Furthermore, this case is the fourth of the cases of Marchiafava-Bignami disease in a woman described up to now.

Key words: Marchiafava-Bignami disease – Chronic alcoholism – Necrosis – Demyelination – Corpus callosum.

Zusammenfassung. Es wird über den ersten Fall einer Marchiafava-Bignamischen Krankheit bei einer Deutschen berichtet. Die Patientin war schwere Trinkerin und verstarb nach einem zweiwöchigen Koma. Bei der Obduktion zeigte sich eine typische Degeneration des Balkens und der subcorticalen weißen Substanz der rechten Großhirnhemisphäre. Weiter ist dieser Fall unter den bisher beschriebenen Fällen von Marchiafava-Bignamischer Krankheit der vierte bei einer Frau.

Schlüsselwörter: Marchiafava – Bignamische Krankheit – Chronischer Alkoholismus – Nekrose – Demyelination – Corpus callosum.

Introduction

It was long supposed that Marchiafava-Bignami disease was restricted to Italians and effected men exclusively. Now many cases of this disease, characterized by primary degeneration of the corpus callosum, in non-Italians have been reported and reviewed excellently by Seitelberger and Berner (1955), Ironside et al. (1961) and more recently by Castaigne et al. (1971), who counted 105 cases, only three of them in women (Schwob et al., 1953; Delay et al., 1956; Castaigne et al., 1971). However, the great majority of those patients living in Mediterranean countries is striking.

The present case of Marchiafava-Bignami disease is the first case in a native German who always lived in Germany and the fourth case in a woman.

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Case Report

A female patient (case no. GZ 202/77) of 55 was admitted to the Klinikum Steglitz, West Berlin. Because the patient could not be asked herself and did not live in a family, it was difficult to get anamnestic information. However, we learned from her daughter-in-law that the patient showed no symptoms at all up to one month before her death. Then her husband, who also was a heavy drinker, died. After the funeral of her husband the patient began to refuse food. She was even unable to swallow Lasixtablets administered for her leg edemas. Two weeks later she could no longer speak and she finally fell, which led to admission to the hospital. She was in a deep coma and showed no reaction to pain stimuli. She showed cachexy (weight 45 kg, 170 cm tall), severe exsiccosis and Cheyne-Stokes respiration. Cirrhosis of the liver was also ascertained. After infusions and antibiotic therapy her state of consciousness improved and she reacted temporarily to words spoken to her. Wernicke's encephalopathy was suspected and vitamin B_1 was administered. Three days later she developed a heart insufficiency with bradycardiac dysrhythmias. She suddenly died 15 days after admission with total atrioventricular block and respiratory arrest.

At necropsy cirrhosis of the liver, bronchiectases, chronic bronchitis and dilatation of the right heart ventricle were found.

As regards the kind of alcoholic beverages, the daughter-in-law of the patient said that the patient regularly liked to drink a fruitwine made of red currants, which can be bought directly from private producers in Berlin. She also drank two or three glasses of normal wine each evening.

Neuropathological Findings

Gross examination showed a normally configurated brain 1235 g in weight with slight fibrosis of the leptomeninges. The arteries of Willis' circle were free of sclerotic changes. Coronal sections through the brain revealed a punched-out symmetrical greyish lesion of the central corpus callosum which was restricted to the middle part of the truncus and did not encroach on the genu or splenium corporis callosi. Nowhere in the midline was the lesion discontinuous as seen by Merritt and Weisman (1945) and Seitelberger and Berner (1955). The corpora

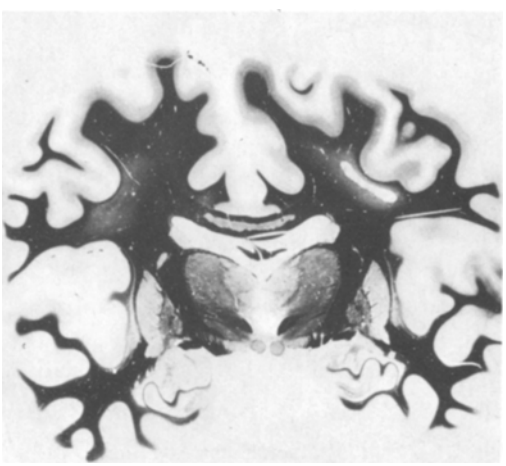

Fig. 1. Coronal section through the corpora mamillaria showing the two demyelinated lesions (Heidenhain-Wölcke)

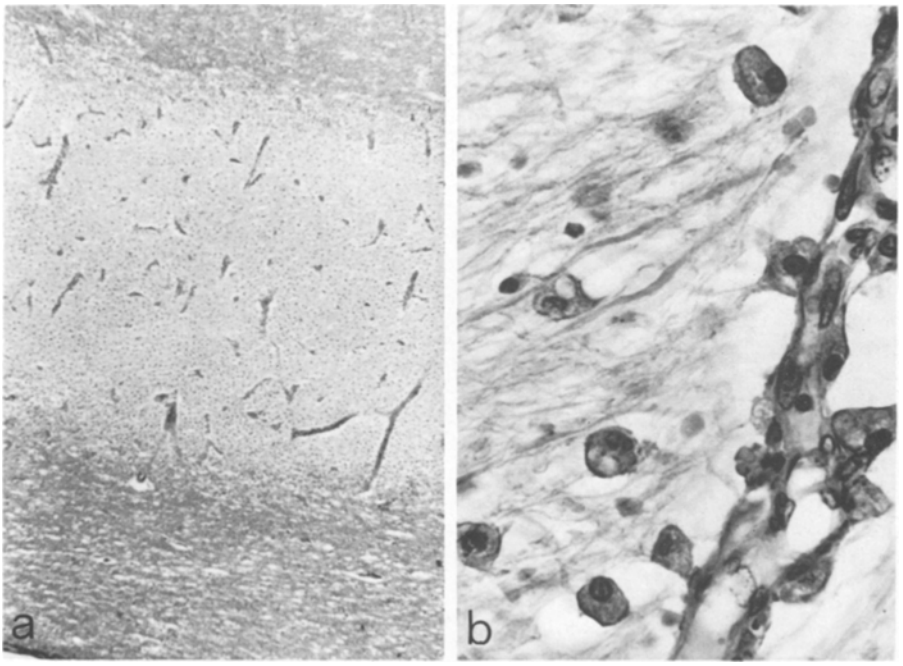

Fig. 2. a The lesion in the corpus callosum with proliferated bloodvessels. **b** Detail of **a**: Lipid phagocytes and bloodvessel in the centre of the lesion (Hematoxylin-eosin)

mamillaria were pale and macroscopically inconspicuous like all other parts of the brain. Microscopically the lesions showed a demyelination (Fig. 1) and an almost total necrosis of oligodendrocytes and astrocytes. The entire lesions were packed with innumerable lipid phagocytes and many proliferated, fibrotic bloodvessels with marked hypertrophy of the vesselwalls could be seen (Fig. 2). Axis cylinders were preserved within the lesions and in the surrounding slightly vacuolated edematous neuropil some swollen axons could be observed. In the subcortical lesion exactly the same changes were found. Apart from this a certain gliosis of the corpora mamillaria was to be seen, but no other pathological findings could be made, there was especially no evidence of Wernicke's encephalopathy.

Discussion

Obviously chronic alcoholism in combination with undernutrition led to the characteristic feature of Marchiafava-Bignami disease. It is possible that ingredients in red currant wine have a toxic effect similar to that of the ingredients in some sorts of Italian red wine, although it has not yet been possible to prove which ingredients are responsible. On the other hand a vascular participation in the pathogenesis of this disease seems to be most probable.

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