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Maurice Victor


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MR in the Diagnosis of Wernicke-Korsakoff Syndrome

Maurice Victor\(^1,2\)

Wernicke disease, or Wernicke encephalopathy, is a neurologic disorder of abrupt or saltatory onset, characterized by nystagmus, abducens and conjugate gaze palsies, unsteadiness of stance and gait, confusion, and apathy. These manifestations may occur singly or in various combinations. Korsakoff psychosis (better termed Korsakoff syndrome or Korsakoff amnesic state) refers to an abnormality of mentation in which learning and memory are affected out of proportion to other cognitive functions; the patient is otherwise alert and responsive. Most patients who present with the manifestations of Wernicke disease and survive the acute illness are left with an enduring amnesic state—in which case the term Wernicke-Korsakoff syndrome is appropriate. The syndrome is common; autopsy studies in disparate parts of the world (Oslo, Boston, Cleveland, and Western Australia) have shown a prevalence of between 2 and 3% of the adult population [1].

The Wernicke-Korsakoff syndrome is due to nutritional deficiency, more specifically to a deficiency of vitamin B\(_1\), or thiamine. In the Western world, it occurs most often in alcoholics. Alcohol displaces food in the diet and also adds its own carbohydrate calories, thus increasing the need for thiamine. However, the syndrome is observed with some regularity in a wide variety of other medical settings: prolonged infectious-febrile conditions, carcinoma, hyperemesis gravidarum, small-bowel obstruction, anorexia nervosa, and prolonged voluntary starvation—to name the better documented ones.

Increasingly, in recent years, attention has been drawn to iatrogenic factors in the causation of the Wernicke-Korsakoff syndrome. From time to time, the syndrome is a complication of a prolonged course of dialysis or hyperalimentation. In a significant proportion of patients who have gastric partitioning or plication, for the treatment of morbid obesity, persistent vomiting and the Wernicke-Korsakoff syndrome develop. In confused alcoholics and otherwise nutritionally depleted patients, particularly those with other serious medical illnesses, the long-term administration of dextrose and water without supplemental vitamins may precipitate the syndrome or cause an early form of it to worsen. This is not an uncommon clinical event, even on the wards of teaching hospitals [2] and is all the more distressing because it can be prevented by the simple expedient of adding B vitamins to the parenteral fluids.

Patients who die in the acute stages of Wernicke disease show symmetrical lesions in the paraventricular regions of the thalamus and hypothalamus, in the mammillary bodies, periaqueductal region of the midbrain, floor of the fourth ventricle (particularly in the regions of the dorsal motor nuclei of the vagus and vestibular nuclei), and midline structures of the cerebellum. Histologically, the lesions are characterized by various degrees of necrosis, with prominence of blood vessels and a proliferation of astrocytic and microglial cells. Hemorrhages are found in only 20% of autopsied cases; they are usually petechial in size, and many of them appear to be agonal.

In this issue of the AJNR, two separate groups of neuroradiologists [3, 4] describe for the first time the MR appearance of the acute lesions of the Wernicke-Korsakoff syndrome. Bilaterally symmetrical diencephalic (medial thalamic) and mesencephalic (periaqueductal) lesions stand out with remarkable clarity, much more so than with CT scanning [5, 6]. This is a matter of more than theoretical interest. If early signs and symptoms of the syndrome are not recognized,
and specific treatment (parenteral administration of large doses of thiamine) is not undertaken immediately, the disease invariably will progress to a state of stupor, coma, and death. Conversely, early diagnosis and treatment halts the progression of the disease. The manifestations of Wernicke disease can be reversed partially or completely, depending on their duration before the institution of treatment, and the development of a Korsakoff amnesic state can be prevented, or an early form of the state can be kept from worsening. Thus, MR may be of critical importance, particularly in the cases that do not conform to the classic clinical picture.

The classic triad of signs described by Wernicke—ophthalmoplegia, ataxia, and an apathetic-confusional state—is still diagnostically useful, but it will be found on initial examination in only one third of patients. In the remainder, the signs occur singly or in some other combination. Moreover, the ocular and ataxic signs are so sensitive to thiamine that they can be attenuated greatly by a meal or two (e.g., before hospital admission) and then can be detected only if carefully sought. Thus, ophthalmoparesis can be reduced to a fine horizontal nystagmus on far lateral gaze, and ataxia may be so restricted as to be elicited only on tandem walking. The occurrence of coma as the initial event in Wernicke disease is rare; however, apathy, drowsiness, confusion, and profound fatigue are commonplace and will progress to coma if untreated, as they did in Wernicke’s original cases [1]. Once coma has supervened, identification of the disease becomes more difficult; the presence of serious medical disease (e.g., sepsis, hepatic failure) compounds this difficulty.

In summary, an awareness of the commonality of the Wernicke-Korsakoff syndrome and the clinical settings (not only alcoholism) in which it occurs and some knowledge of the incomplete or attenuated forms of the syndrome and how to elicit them permit a diagnosis to be made in most cases. Even a suspicion of the disease calls for the immediate parenteral administration of large doses of thiamine, after which MR logically could be used to substantiate the diagnosis.

Finally, MR (and CT) may serve another purpose in the diagnosis of Wernicke-Korsakoff syndrome—namely, to identify the nature of a chronic Korsakoff amnesic state in patients who have never shown the ocular and ataxic signs of Wernicke disease or in whom these signs have been abolished. Occasionally, such an amnesic state may be the only manifestation of Wernicke-Korsakoff syndrome, or a clinically indistinguishable state may be due to other forms of diencephalic disease (infarction, tumor) or to discrete lesions in other parts of the brain (basal forebrain, hippocampal formations). Atrophy of the mamillary bodies, medial thalamic nuclei (widening of third ventricle), and periaqueductal gray matter (dilated aqueduct) mark the amnesic state as alcoholic-nutritional in origin. In cases of Korsakoff amnesic state due to basal forebrain or bilateral hippocampal lesions, MR and CT show characteristic abnormalities in these structures [7–10].

REFERENCES