Case report

Magnetic resonance imaging in a case of Wernicke's encephalopathy

L. Pagnan¹, G. Berlot², R. S. Pozzi-Mucelli¹

¹ Institute of Radiology, University of Trieste, Cattinara Hospital, Strada di Fiume, I-34149 Trieste, Italy ² Department of Anaesthesia and Intensive Care, University of Trieste, Cattinara Hospital, Strada di Fiume, I-34149, Trieste, Italy

Received 7 March 1997; Revision received 28 August 1997; Accepted 14 November 1997

Abstract. Wernicke's encephalopathy is an uncommon disorder caused by a thiamine deficiency which is clinically characterized by the triad of ophthalmoplegia, ataxia and disturbances of consciousness, each finding being variably present. The disease is caused by malnutrition or malabsorption, and is often associated with prolonged alcohol intake, neoplasm and extensive inflammatory processes of the digestive tract and parenteral hyperalimentation-induced gastrointestinal mucosal atrophy. Clinical diagnosis can be elusive and MRI may be the only imaging technique able to detect the cerebral lesions, whose type and distribution are characteristic of the Wernicke's encephalopathy, whereas CT is positive only in exceptional cases. We report a case of a 56-yearold woman who developed a Wernicke's encephalopathy 1 month after a colonic resection with signal intensity changes located in the mammillary bodies and in the medial thalamic nuclei.

Key words: MR imaging – Wernicke's encephalopathy – Thiamine deficiency

Introduction

In circumstances associated with an increased need for vitamins, such as during a postoperative phase, the sustained shortage of nutrients eventually leads to clinically relevant consequences [1]. Among vitamins, thiamine plays an important role as a coenzyme both in the Krebs and in the pentose-phosphate cycles. In its absence, glucose metabolism is impaired, causing pyruvate and lactate accumulation and adenosine triphosphate stores depletion [2].

Clinically, this condition is associated with a highoutput haemodynamic status, known as shoshin-beriberi, and with the occurrence of Wernicke's encephalopathy (WE) which is characterized by the triad of ophthalmoplegia, ataxia and consciousness disturbances.

Neuropathologically, blood-brain-barrier dysfunction and oedema-associated local hypoperfusion cause a neuronal loss that can be demonstrated in selected brain areas including the periventricular regions, the medial thalamic nuclei, the massa intermedia, the floor of the third ventricle and the mammillary bodies [3, 4].

At present, MR imaging is considered the most valuable imaging technique to diagnose WE, since cranial CT normally fails to demonstrate significant density alterations and is positive only in exceptional cases [5].

We describe the case of a patient who developed a full-blown WE due to a prolonged post-operative starvation.

Case report

A 56-year-old woman was transferred from the surgical ward to our intensive care unit 1 month after right haemicolectomy for cancer, because her state of consciousness was progressively decreasing. The clinical diagnosis was coma of unknown origin. Possible clinical differential diagnoses were brain neoplasm, possibly metastatic, or vascular disease.

Her past history revealed a mild-to-moderate alcohol intake over a period of several years. At surgery, the liver appeared fatty. In the postoperative course there was a persistent ileus, with poor response to intravenous prostygmin; however, repeated barium enemas failed to demonstrate any abnormality.

The ileus prevented any oral intake, but the patient refused the positioning of a central venous catheter (CVC) to supply fluids and total parenteral nutrition.

Small amounts of glucose and aminoacids were then peripherally supplied, which were, however, unable to prevent a weight loss of more than 12 kg.

Upon admission to the intensive care unit, moderate hypotension (arterial pressure 90/60 mm Hg) and tachy-

Correspondence to: R.S.Pozzi-Mucelli

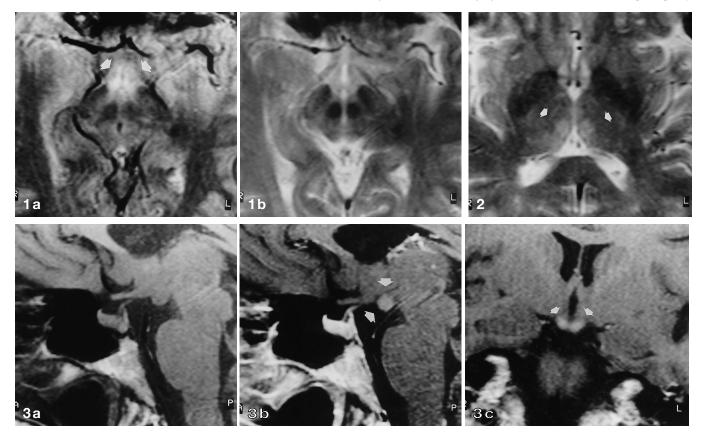


Fig. 1. a Axial proton-density image (TR = 3000, TE = 20); **b** Axial T2-weighted image (TR = 3000, TE = 80): mild hyperintensity of the floor of the third ventricle, better shown in **a** (arrows)

Fig. 2. Axial T2-weighted image (TR = 3000, TE = 80): mild symmetrical signal hyperintensity of the dorsal thalamic nuclei (*arrows*)

Fig. 3. a Sagittal T1-weighted image (TR = 450, TE = 19); **b–c** sagittal and coronal T1-weighted images after gadolinium-DTPA: marked symmetrical enhancement of the mammillary bodies (*arrows*)

cardia (heart rate 130 bpm) were present but subsided rapidly after administration of fluids via a CVC inserted on an emergency basis. Blood chemistry demonstrated a moderate anaemia (Hb = 7.2 g/dl) associated with hyponatremia (Na⁺ = 120 mEq/l) and hyperlactataemia (6 mEq/l). Despite the elevated blood lactate values, both the blood and the intramucosal gastric pH (ipH) were normal. Anaemia was treated with packed red blood cells and hyponatraemia was slowly corrected.

Hyperlactataemia did not change even after the patient was fully volume resuscitated and a total parenteral nutrition (TPN) was started.

The patient appeared drowsy but arousable; peripheral reflexes were bilaterally reduced; the pupils reacted sluggishly to light, and a bilateral horizontal nystagmus was present. Heel-to-knee and finger-to-nose tests were abnormal.

The MRI images were obtained at 1.5 T (Gyroscan ACS II, Philips, Best, The Netherlands), using spinecho T1-, proton-density and T2-weighted sequences with axial, coronal and sagittal sections. T1 sequences were repeated after gadolinium-DTPA injection.

Proton-density images showed high signal intensity (Fig. 1 a) in the suprasellar cistern, at the level of the hypothalamus. This finding was less striking in the T2-weighted image (Fig. 1 b). Bilateral and symmetrical mild hyperintensities were seen in the medial thalamus (Fig. 2).

Sagittal and coronal T1-weighted scans, after gadolinium-DTPA injection, clearly showed bilateral and symmetrical enhancement of the mammillary bodies (Fig. 3).

Wernicke's encephalopathy was then confirmed and the patient was given 100 mg thiamine intravenously as a bolus, and the same daily dose was administred with the TPN. After 2 days, hyperlactataemia fully subsided and the neurological conditions started to improve. One week after the beginning of the treatment, the patient was fully alert, but both the nystagmus and the signs of cerebellar dysfunction were still present, although reduced. Bowel movements reappeared and the patient was placed on enteral nutrition (EN) and then transferred to the surgical ward and discharged home free of symptoms.

Discussion

According to some investigations, findings characteristic of WE can be demonstrated in 1-2% of all autopsies. The same findings are found in 12.5% of autopsies made in alcoholic patients [6], although WE is L. Pagnan et al.: MR imaging in a case of Wernicke's encephalopathy

clinically diagnosed only in 20% of these subjects [2, 7].

Although it was initially considered a consequence of the alcohol-related malnutrition, WE can occur in all conditions associated with a reduced thiamine intake and/or absorption, including prolonged starvation [8], hyperemesis gravidarum [9], anorexia nervosa, Wipple's disease, malignancies [10] and extensive inflammatory disorder of the gastrointestinal tract [11, 12].

In post-operative patients, thiamine deficiency can occur either due to surgical reduction of the absorptive surface, or due to functional conditions such as protracted ileus or vomiting. The occurrence of WE is increasingly reported also in patients undergoing banded gastroplasty for morbid obesity [13, 14].

The clinical diagnosis of WE is not always straigthforward mainly for two reasons:

1 Its signs and symptoms may not be present altogether, or they can be too subtle to be recognized early.

2. Its onset can be either abrupt or gradual, with symptoms developing over days or weeks [1].

Early diagnosis is essential in order to avoid irreversible neuronal changes associated with a persisting shortage of thiamine. Moreover, a high index of suspicion should be maintained in patients with known risk factors, because the supply of thiamine-free parenteral glucose can precipitate WE [13].

In our patient, the simultaneous occurrence of neurological disturbances associated with systemic hyperlactataemia unrelated to gastrointestinal acidosis [15] suggested the clinical diagnosis of WE, which was radiologically confirmed by an MRI examination.

Computed tomography is not considered a reliable technique diagnosing WE. Computed tomography is positive only in exceptional cases. The CT findings include mild hypodensity in the selected areas already mentioned [2, 4].

Thanks to its multiplanar imaging capability and its intrinsically higher contrast resolution, it is easier to detect the lesions on MRI. The abnormalities are typically bilateral, symmetrical and involve selected and pathognomonic zones (periventricular regions, medial thalamic nuclei, massa intermedia, third ventricular floor and mammillary bodies) [2, 4, 16].

However, the distribution of the lesions may vary widely. In fact, some authors report that in the chronic form, lesions can be restricted to mammillary bodies and the thalamus, as in our case [3], but the periaqueductal region, midbrain, reticular formation and tectal plate can also be commonly involved [4].

Unusual symmetrical hyperintense lesions involving selected cortical areas, along the central and precentral sulci, have also been reported [17]. Our MRI study was obtained using spin-echo sequences. A T1-weighted spin echo showed mild focal signal hypointensity with an important enhancement in the mammillary bodies, after contrast administration, due to the blood-brainbarrier breakdown [18–21], whereas proton-density and T2-weighted sequences showed diffuse signal hyperintensity in the same areas.

The signal characteristics and the site of these lesions are considered virtually pathognomonic of Wernicke's encephalopathy [2]. However, some authors reported a case with typical WE imaging findings that autopsy revealed cytomegalovirus encephalitis and primary cerebral lymphoma in a 29-year-old HIV-positive patient [22].

Some authors have reported the utility of FLAIR sequence in detecting lesions and also in monitoring them [23].

In conclusion, the distinctive findings of this case were the very selective location of the MR abnormalities in the mammillary bodies, the floor of the third ventricle and the dorsal nuclei of the thalamus and the great efficacy of MR imaging which, due to the typical findings, enabled early diagnosis which could not be predicted on a clinical basis. Therefore, the quick onset of thiamine therapy led to regression of symptoms and prevented permanent brain lesions modifying the prognosis.

References

- Dempsey DT, Mullen JL, Buzby GP (1988) The link between nutritional status and clinical outcome: Can nutritional intervention modify it? Veteran administration cooperative trial of perioperative total parenteral nutrition in malnourished surgical patients, background, rationale and study protocol. Am J Clin Nutr 47: 351–356
- Brody BA (1996) The Wernicke-Korsakoff syndrome. Neuropathology and pathogenetic basis. Int J Neuroradiol 2: 216–230
- Heye N, Terstegge K, Sirtl C, Mc Monagle U, Schreiber K, Meyer-Gebner M (1994) Wernicke's encephalopathy: causes to consider. Intensive Care Med 20: 282–286
- Osborne AG (1994) Acquired metabolic, white matter, and degenerative diseases of the brain. In: Osborne AG (ed) Diagnostic neuroradiology. Mosby, St. Louis, pp 748–781
- Gallucci M, Bozzao A, Splendiani A, Masciocchi C, Passariello R (1990) Wernicke encephalopathy: MR findings in five patients. Am J Neuroradiol 11: 887–892
- Torvik A, Lindboe CF, Rogde S (1982) Brain lesions in alcoholics. A neuropathological study with clinical correlation. J Neurol Sci 56: 233–248
- Schroth G, Wichmann W, Valavanis A (1991) Blood-brain-barrier disruption in acute Wernicke encephalopathy: MR findings. J Comput Assist Tomogr 15: 1059–1061
- Doraiswamy P, Massey E, Enright K, Palese VJ, Lamonica D, Boyko O (1994) Wernicke-Korsakoff syndrome caused by psychogenic food refusal. MR findings. Am J Neuroradiol 15: 594–596
- Omer SM, Al-Kawi MZ, Al-Watban J, Bohlega S, McLean DR, Miller G (1995) Acute Wernicke's encephalopathy associated with hyperemesis gravidarum: magnetic resonance imaging findings. J Neuroimaging 5: 251–253
- Mizutani T (1995) Neurologic complication of cancer. Their pathogenesis and management. Biotherapy 9: 1472–1479
- 11. Reuler JB, Girard GE, Cooney TG (1985) Current concepts: Wernicke's encephalopathy. N Engl J Med 312: 1035–1038
- 12. Girault JM, Dousset V, Andrieu De Levis P, Viaud B, Berge J, Carlier P, Caillé JM (1995) Quid? J Radiol 76: 297–299
- Abarbanel JM, Berginer VM, Osimani A, Solomon H, Charuzi I (1987): Neurologic complication after gastric restriction surgery for morbid obesity. Neurology 37: 196–200

- L. Pagnan et al.: MR imaging in a case of Wernicke's encephalopathy
- 14. Shiozawa T, Shiota H, Shikata E, Kamei S, Mizutani T (1995) Development of Wernicke'encephalopathy during the period of oral food intake after a subtotal colectomy for ulcerative colitis. Clin Neurol 36: 169–174
- Ruokonen E, Takala J, Kari A, Saxen H, Mertsola J, Hansen EJ (1993) Regional blood flow and oxygen transport in septic shock. Crit Care Med 21: 1296–1303
- Lee HK, Chang KH, Na DO, Han MH (1996) MR imaging findings of Wernicke's encephalopathy in non-alcoholic patients. Int J Neuroradiol 2: 210–215
- 17.Yamashita M, Yamamoto T (1995) Wernicke's encephalopathy with symmetric pericentral involvement. MR findings. J Comput Assist Tomogr 19: 306–308
- Harter SB, Nokes SR (1995) Gadolinium-enhanced MR findings in a pediatric case of Wernicke's encephalopathy. Am J Neuroradiol 16: 700–702

- Shogry MEC, Curnes JT (1994) Mammillary body enhancement on MR as the only sign of acute Wernicke's encephalopathy. Am J Neuroradiol 15: 172–174
- D'Aprile P, Gentile MA, Carella A (1994) Enhanced MR in the acute phase of Wernicke encephalopathy. Am J Neuroradiol 15: 591–593
- Schroth G, Wichmann W, Valavanis A (1991) Blood-brain-barrier disruption in acute Wernicke encephalopathy: MR findings. J Comput Assist Tomogr 15: 1059–1061
- 22. Brechtelsbauer DL, Urbach H, Sommer T, Blumcke I, Woitas R, Solymosy L (1997) Cytomegalovirus encephalitis and primary cerebral lymphoma mimicking Wernicke's encephalopathy. Neuroradiology 39: 19–22
- 23. Maeda M, Tsuchida C, Handa Y, Ishii Y (1995) Fluid attenuated inversion recovery imaging in acute Wernicke's encephalopathy. Radiat Med Med Imaging Radiat Oncol 13: 311–313

Book review

De Salles, A., Lufkin, R.: Minimally Invasive Therapy of the Brain. New York, Stuttgart: Thieme 1997. 278 pp., 325 illustrations, (ISBN 3-13-106541-9), DM 375.00.

The book is edited by a neurosurgeon and a neuroradiologist and has contributions from 41 different authors, mainly from the United States and Germany. In their preface the authors state that minimal invasive therapy of the brain is a powerful technique for lowering the costs of medical care. This text arose from the need to document the new techniques available for the treatment of brain pathologies that can compete with the traditional large surgical approaches.

The text has 21 chapters and covers all surgical pathologies of the brain: brain tumors, neurovascular, and CNS functional disease.

The first chapter is written by the editors and gives a short overview starting with the history of imaging techniques and ending with neurosurgical endoscopy.

The following chapters deal with frameless stereotaxis, graphic workstations, and databases. This last subject is dealt with in too much detail and hard to understand for a non-computer freak.

The fifth chapter is an excellent introduction to functional MRI and its implications for preoperative mapping, together with helpful references. Unfortunately the most recent references are from 1995, but this chapter is nevertheless a good appetizer for all those wanting to start with functional MRI.

Chapter 6 deals with MR spectroscopy over 14 pages and I still did not grasp why MRS is an important tool for minimal invasive procedures. It would have been more effective to have a longer chapter on "Endoscopy of the cranial base" which is, with 6 pages, clearly too short.

Gary Duckwiler and Fernando Vinuela wrote the part of endovascular therapy in their usual excellent style with illustrative figures and well-chosen references. They remark that neurointerventional procedures are not really minimally invasive despite their small needle puncture approach. They carry a substantial risk of morbidity and mortality of up to 5%. In total, this is an excellent overview of all endovascular approaches from transarterial tumor embolization to angioplasty.

European

Radiology

The next five chapters cover the field of MR-guided treatment techniques, starting with a technical introduction, the instrumentation, clinical application of interstitial thermotherapy, and different ablation techniques. Despite the fact that larger clinical results are still lacking all these chapters are well written and understandable. It remains for the reader to extrapolate what the clinical role of these techniques might be in the future. Sometimes I felt the figures could have been a little smaller, specifically in the chapter on clinical application of laser-induced interstitial thermotherapy. The graphs of the temporal evolution of the temperature-dependent phase shift cover two pages and are hard to understand.

Chapters 13–17 deal with radiosurgery: principles, AVMs, benign and malignant brain tumors, and functional disease. And again, the reader gets a good state-of-the-art overview and many answers to questions of well-informed patients. As in previous chapters the figures are sometimes to space-occupying, specifically in the chapter on benign tumors.

The last four chapters cover the field of neuroendoscopy from instrumentation to interactive image-guided and ventricular and cranial base endoscopy. These could have been longer and more extensive.

In summary, "*Minimal Invasive Therapy of the Brain*" is a valuable resource for all those involved in the therapy of neurological diseases. It provides everybody with a well-written overview of all minimal invasive techniques under one cover. It is probably beyond the scope of radiology residents, but clearly belongs in the departmental library and in the personal library of neurosurgeons, neuroradiologists, and radiation oncologists. Its disadvantage is that due to the rapidly evolving nature of the field this work of reference will soon need up-dating. So we have to look for the second edition or go into the recommended internet website "www.radiology.com" for further information. M. Forsting, Essen