Wernicke’s encephalopathy with atypical cortical damage∗

N. Alberti a,∗, J. Bocquet a, S. Molinier a, A. Veron a, S. Roggerone a, L. Pinsard a, F.-X. Arnaud b, V. Dousset a

a Service de Neuro-imagerie, CHU Pellegrin, place Amélie-Raba-Léon, 33000 Bordeaux cedex, France
b Service d’Imagerie, HIA Val-de-Grâce, 74, boulevard de Port-Royal, 75230 Paris 05, France

Case report

A 32-year-old man was found unconscious at home (GLASGOW estimated at 4). The rest of the physical examination did not provide very much information, in particular, there was no sign of neurological focalisation. The pupils were also reactive in semi-mydriasis.

According to those close to him, his daily consumption of alcohol was judged to be ‘‘excessive’’. He supposedly described a sensation of ‘‘double vision’’ over the last three days and could not reach for the last 24 hours. The first laboratory tests did not reveal anything specific and, in particular, the glycaemia was normal. The toxicology assessment was also negative. The emergency X-ray computed tomography was found to be normal as was the lumbar puncture. The EEG was not contributory, revealing a non specific diffuse slowing down. An encephalic MRI was carried out (Figs. 1 and 2).
Wernicke’s encephalopathy with atypical cortical damage

What is your diagnosis?

After reading the case report, what diagnosis would you choose from the following proposals:

- atypical Wernicke’s encephalopathy;
- epilepsy;
- cerebral hypoxia;
- thrombophlebitis of the right sinus and the upper longitudinal sinus;
- variant of Creutzfeld–Jacob’s disease.

Diagnosis

Atypical Wernicke’s encephalopathy.

Description of the images

The FLAIR sequences provided reveal a bilateral and symmetrical peri-ventricular hypersignal of the fourth ventricle...
and bi-thalmics (median nuclei) in diffusion restriction as well as atypical signal anomalies of the vertex such as cortical hypersignals in diffusion restriction, in a ribbon-like fashion (Figs. 3 and 4). These anomalies were all integrated in a Wernicke’s encephalopathy with atypical cortical damage. The patient came out of the coma quickly after vitamin therapy and adapted reanimation measures. The cortical anomalies of the vertex regressed in a noteworthy manner on the control MRI, as did the bi-thalmic and periventricular lesions. They totally disappeared after six months.

Discussion
Wernicke’s encephalopathy is a metabolic disease related to a thiamine (vitamin B1) deficiency. It mainly affects alcoholic and especially undernourished patients. This disease is also found in cases of malnutrition (anorexia nervosa, prolonged fast, parenteral nutrition without the addition of thiamine) or even in case of prolonged vomiting (sometimes pre-enclampsia) [1].

The mechanisms underlying this disease are still not completely clear. Thiamine plays a role in the metabolism of

Figure 3. Axial sections of MRI in FLAIR sequence and diffusion sequence B1000 (a, b). Bilateral and symmetrical cortical hypersignals of the vertex (arrows). These signal anomalies are clearly in diffusion restriction on diffusion sequence B1000 (arrow).

Figure 4. Axial section of MRI in FLAIR sequence passing in thalamic region as well as at the height of the fourth ventricle respectively (a, b). Bilateral and symmetrical hypersignals of the thalami in their dorso-median portions (arrows) as well as the peri-ventricular substance of the fourth ventricle (arrow).
Wernicke's encephalopathy with atypical cortical damage

alcohol and glucose. In fact, thiamine acts like a coenzyme of three essential enzymes in the intermediate metabolism of carbohydrates: transketolase (that mainly acts in the synthesis of DNA), α-ketoglutarate dehydrogenase (that plays a role in neuron excitotoxicity) and the pyruvate dehydrogenase complex. A deficiency in one of these enzymes in case of a thiamine deficiency leads to an osmotic deregulation between the extra- and intracellular media, directly resulting in sequence of diffusion anomalies. The metabolism of the periventricular regions particularly depends on thiamine, accounting for the predominance of lesions at this level [1,2].

Suddenly occurring in its most common form, Wernicke's encephalopathy is classically characterised by a clinical triad including reduced consciousness of variable importance, a cerebellar syndrome (in particular ataxia and dysarthria) as well as oculomotor disorders (ophthalmoplegia involving three or six, nystagmus). The symptomatology is rarely complete. The diagnosis is therefore based on the imaging and, more specifically, the MRI. Wernicke's encephalopathy specifically impairs the hippocampo-mamillo-thalamic circuit (Papez circuit). It also affects the grey substance in contact with the iter of Sylvius (grey periaque ductal substance) and the fourth ventricle. These classically bilateral and symmetrical lesions are specific to this disease. In 1998, the work of Antunez et al. demonstrated that simultaneous impairment such as T2-FLAIR hypersignal of the periaque ductal regions and the medial dorsal thalamic nuclei was in favour of such an encephalopathy with a specificity of 93% [3]. Moreover, these lesions take up the contrast after the injection of Gadolinium in a non constant manner [1]. No enhancement is noted in the aforementioned zones in our case. However, in our patient, the cortical impairment in vertex ribbon was more outstanding. This aspect is widely described in the versive disease (and especially in the postcritical state), in reversible posterior leuko-encephalopathy or in case of hypoglycaemia. More rarely, it may be found in the sporadic form of Creutzfeld–Jakob's disease. The context was not in favour of any of the aforementioned aetiologies. Nevertheless, this cortical impairment of the vertex may be integrated in Wernicke's encephalopathy. The team of Philip J. Langlais and Shu-Xing Zhang demonstrated the cortical vulnerability of the vertex in a great number of mice with a thiamine-poor diet [4]. Most often reversible, they may turn out to be permanent in some of them. In man, these anomalies of signal have been described although a great many grey zones persist, in particular in the prognosis of the latter.

It is interesting to note that these lesions do not necessarily have an obvious clinical translation. For this reason, we are reporting the case of a patient with Wernicke's encephalopathy with cortical impairment of the vertex similar to our patient and presenting a high level of consciousness enabling an in-depth clinical examination without noteworthy clinical motor impairment [5]. The consciousness disorders of our patient did not allow for the detection of this radio-clinical conflict. Other atypical topographies may be revealed during such a disease: the head of the caudate nuclei and lenticular nuclei, red nuclei, nuclei of the facial, abducen and vestibular nerves are rare targets possibly impaired during Wernicke's encephalopathy [1]. Our patient came out of his coma quickly after vitamin therapy and adapted reanimation measures. The cortical anomalies of the vertex regressed in a noteworthy manner in the control MRI as did the bi-thalamic and periventricular lesions. They totally disappeared after six months.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References