WERNICKE’S ENCEPHALOPATHY PRESENTING WITH SEVERE DYSPHAGIA: A CASE REPORT

MIKAEL TRUEDSSON*, BODIL OHLSSON and KLAS SJÖBERG

Department of Medicine, Lund University, University Hospital, S-20502 Malmö, Sweden

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Abstract — A 62-year-old man developed dysphagia 4 weeks before the classic symptoms and signs of Wernicke’s encephalopathy appeared. Adequate treatment with parenteral thiamine resulted in complete recovery of all symptoms, including his dysphagia. This extraordinary presentation with dysphagia, and the unusual course of the disease, encouraged us to present this case history.

INTRODUCTION

Wernicke’s encephalopathy is caused by a deficiency of thiamine and is characterized by the triad of ophthalmoplegia, ataxia and mental confusion. Pathological lesions of Wernicke’s encephalopathy are characteristically distributed symmetrically in structures surrounding the third and fourth ventricles, including the mammillary body, dorsomedial thalamus, periaqueductal grey matter, anterior superior cerebellar vermis, oculomotor nuclei and vestibular nuclei (Charness, 1993). Most frequently, the encephalopathy is a consequence of alcohol misuse, but it may occur in any disease which causes sufficient malnutrition, such as gastric carcinoma and pyloric obstruction (Barrie, 1947), hyperemesis gravidarum (Chaturachinda and McGregor, 1968), prolonged parenteral feeding (Nadel and Burger, 1976) and hunger strike (Pentland and Mawdsley, 1982). Wernicke’s encephalopathy is curable if thiamine is administered parenterally early in the course of the disease, but it may cause neurological deficits, or, in the worst case, death, if therapy is delayed (Shikata et al., 2000).

A patient who presented with dysphagia and later developed a classical Wernicke’s encephalopathy prompted us to present his interesting case history for discussion.

CASE HISTORY

A 62-year-old man was admitted to our hospital with increasing dysphagia of 3 weeks duration, a swollen neck and a 10 kg weight loss. Physical examination revealed a normal heart rate of 72 beats/min. His blood pressure was 140/90 mmHg and there was no thyroid enlargement or palpable lymph nodes. His medical history consisted of hypertension and chronic bronchitis.

He had elevated levels of plasma ASAT 2.65 μkat/l (aspartate aminotransferase; normal level: <0.80 μkat/l), ALAT 2.02 μkat/l (alanine aminotransferase; normal limit <0.80 μkat/l), amylase 0.88 μkat/l (normal range: 0.20–0.80 μkat/l), sedimentation rate 75 mm/h (normal level <22 mm/h), serum α1-antitrypsin 1.93 g/l (normal range: 0.87–1.68 g/l), serum orosomucoid 1.61 g/l (normal range: 0.54–1.17 g/l), serum c-reactive protein 8.9 mg/l (normal limit: <3.0 mg/l), serum ceruloplasmin 0.53 g/l (normal range: 0.22–0.38 g/l), serum immunoglobulin IgA 6.30 g/l (normal range: 0.70–3.65 g/l) and plasma creatinine 106 μM (normal range: 63–105 μM), while plasma sodium 134 mM (normal range: 136–146 mM) was slightly decreased.

Initially, the investigations concentrated on excluding malignancy of the oesophagus or the surrounding tissue. Chest X-ray showed evidence of emphysema. Endoscopy of the oesophagus and stomach was normal. A barium meal revealed some dysfunction of the oesophagus with slightly reduced motility; an ultrasound of the liver, bile ducts and pancreas showed signs of a fatty liver. The patient was examined by an ENT physician, who could not find any explanation for the dysphagia.

Because of his inability to swallow, the patient initially received a glucose infusion intravenously and after a few days an intestinal feeding tube was placed into the descending duodenum for enteral nutrition.

Five days after admission, the patient developed diplopia and staggering gait. The neurological signs consisted of gait ataxia, horizontal nystagmus, lateral rectus palsy, absence of deep tendon reflexes and positive Babinski’s sign on the right side. It was not until then that it was known that he had been drinking ~225 cl of 40% alcohol (i.e. 720 g) per week for the last 8 months and consequently it was suspected that the patient was suffering from Wernicke’s encephalopathy. The patient received two injections of thiamine (50 mg/ml, 100 mg per dose) intravenously. The time period between the two doses was 16 h. The neurological symptoms and signs, including the dysphagia, completely disappeared 24 h after the first injection.

DISCUSSION

Our patient developed signs compatible with progressive Wernicke’s encephalopathy. The unique finding in this patient was his dysphagia, which developed 4 weeks before any other neurological symptoms occurred. We believe that dysphagia is an extremely rare symptom in this disease, and has only been briefly mentioned in two Japanese articles (Sakakibara et al., 1997; Kikuchi et al., 2000). Our patient had difficulties in initiating swallowing, suggesting an oropharyngeal dysphagia. The oropharyngeal stage of deglutition is controlled by the n. glossopharyngeus, n. vagus and n. hypoglossus. The motor nuclei of these nerves are located in the floor of the fourth
ventricle, an area that may be affected in Wernicke’s encephalopathy (Charness, 1993), suggesting that his dysphagia was an early manifestation of Wernicke’s encephalopathy.

Endoscopy, a barium meal and examination by an ENT physician did not provide any alternative explanation for the dysphagia. The swollen neck was initially considered to be due to subcutaneous emphysema, but chest X-ray showed no pneumothorax, nor any mediastinal tumour.

When the neurological symptoms and signs developed a few days later, the differential diagnoses considered were hypophosphataemia, central pontine myelolysis, Miller–Fisher’s syndrome and Wernicke’s encephalopathy.

Alcoholism and alcohol withdrawal in combination with carbohydrate infusion after starvation are known to contribute to hypophosphataemia (Subramanian and Khardori, 2000). The neurological symptoms and signs resemble Wernicke’s encephalopathy, including ophthalmoplegia, ataxia, areflexia, confusion, dysphagia and muscular weakness (Zurkirchen et al., 1994; Subramanian and Khardori, 2000), but the plasma phosphate concentration in our patient was normal (1.20 mM, reference value 0.70–1.30 mM).

Central pontine myelinolysis is a disease predominantly observed in alcoholics but also non-alcoholics with liver diseases, malnutrition, cancer, congestive heart failure, adrenal insufficiency and renal disease (Ashrafian and Davey, 2001). The most common cause is rapid correction of hyponatraemia. The symptoms are weakness in extremities, diplopia, dysarthria, changes in corticospinal reflexes, dysphagia and confusion (Charness, 1993; Ashrafian and Davey, 2001). This diagnosis was excluded by the almost normal plasma sodium concentration (134 mM, reference value 136–146 mM).

Miller–Fisher’s syndrome, a variant of Guillaum–Barré’s syndrome, is characterized by ophthalmoplegia, ataxia and areflexia (Fisher, 1956), but weakness and sensory disturbances of the limbs may also occur. The syndrome is associated with upper respiratory tract infection and its incidence appears to be highest in Asia. The nosological position of the symptoms is not clear, but it seems that the pathological process affects both the peripheral and central nervous system. The prognosis is good and full recovery or very few symptoms are seen after ~6 months (Mori et al., 2001). Our patient had the classical triad of Miller–Fisher’s syndrome, but no history of infection. Furthermore, full recovery after treatment with thiamine ruled out this diagnosis, as well as the alternative ones suggested.

The development of Wernicke’s encephalopathy in our patient was due to heavy alcohol intake for several months in combination with insufficient food. A relative thiamine deficiency caused dysphagia, which led to further inadequate dietary intake. Our inappropriate glucose infusion aggravated the thiamine deficiency even more and the patient developed classic symptoms of Wernicke’s encephalopathy. Complete recovery occurred 24 h after treatment with adequate doses of thiamine, which confirms the accuracy of the diagnosis.

Notably, only a subset of alcoholics with thiamine deficiency develops Wernicke’s encephalopathy. The reason is perhaps inherited (Blass and Gibson, 1977) or acquired (Jeyasingham et al., 1987) abnormalities of transketolase, a thiamine-dependent enzyme, leading to reduced affinity for thiamine. It is not known why the structures around the ventricles are most susceptible to thiamine deficiency. In contrast to the majority of patients with Wernicke’s encephalopathy, our patient’s first symptom was dysphagia, which indicates that there must be a varying susceptibility for symptoms of lack of thiamine in different brain regions between individuals.

In summary, we report a patient, where the classic signs of Wernicke’s encephalopathy developed after a glucose infusion, a well-known complication that is easily avoided. The exceptional feature was that our patient developed dysphagia several weeks before the other symptoms manifested themselves. This finding has rarely been described and suggests that Wernicke’s encephalopathy should be suspected in patients with dysphagia of unknown cause, especially in the presence of alcohol misuse.

REFERENCES


