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"Syndrome in syndrome": Wernicke syndrome due to afferent loop syndrome. Case report and review of the literature

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SUMMARY: **"Syndrome in syndrome": Wernicke syndrome due to afferent loop syndrome. Case report and review of the literature.**

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Wernicke syndrome is a rare neurological pathology due to a deficit in vitamin B1. The syndrome is common among alcohol abusers, patients with malignant tumor or gastrointestinal diseases, those who undergo hemodialysis or long-term peritoneal dialysis, pregnant women with hyperemesis, women who breast-feed, patients with hyperthyroidism or anorexia nervosa or gastric or jejunal-ileal bypass surgery for obesity, patients submitted to gastric surgery or prolonged total parenteral nutrition or prolonged intravenous therapy.

We report a case of Wernicke syndrome due to afferent loop syndrome characterized by incoercible vomiting.

RIASSUNTO: **"Sindrome in sindrome": sindrome di Wernicke causata da sindrome dell'ansa afferente. Case report e revisione della letteratura.**

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La sindrome di Wernicke è una malattia neurologica rara causata dalla mancanza di vitamina B1. E' comune negli alcolisti, nei pazienti con tumori maligni o malattie gastrointestinali, in quelli sottoposti da lungo tempo a emodialisi o a dialisi peritoneale, nelle donne gravide affette da iperemesi, nelle donne che allattano, nei pazienti con ipertiroidismo o anoressia nervosa e nei pazienti obesi sottoposti a by-pass chirurgico digiuno-ileale, nei pazienti sottoposti a chirurgia gastrica o a nutrizione parenterale totale per lungo tempo o a prolungata terapia endovenosa. Riportiamo un caso di sindrome di Wernicke causata da sindrome dell'ansa afferente, caratterizzata da vomito incoercibile.

KEY WORDS: Wernicke syndrome - Afferent loop syndrome.
Sindrome di Wernicke - Sindrome dell'ansa afferente.

Introduction

Wernicke's encephalopathy (WE) was originally reported in 1881 by Carl Wernicke and first called "polioencephalitis hemorrhagica superior". It was a fatal syndrome characterized by ophtalmoplegia, ataxia and confusion. The first reported patients were two alcoholics and a woman with persistent vomiting after ingestion of sulfuric acid (1). Wernicke syndrome is a rare neurological pathology due to a deficit in vitamin B1. The syndrome is common among alcohol abusers, patients with malignant tumor or gastrointestinal diseases, those who undergo hemodialysis or long-term peritoneal dialysis, pregnant women with hyperemesis, women who breast-feed, patients with hyperthyroidism (2,3) or anorexia nervosa (4) or gastric or jejunal-ileal bypass surgery for obesity (1), patients submitted to gastric surgery (1,5-8) or prolonged total parenteral nutrition (9) or prolonged intravenous therapy (10).

Wernicke's encephalopathy commonly refers to the complex of symptoms of ophtalmoplegia, ataxia and acute confusional state. It is due to thiamine deficiency that shifts the pyruvic acid metabolism towards lactate production with lactic acidosis and anion gap.

Case report

Male patient, aged 56 years, was hospitalized in Neurology Department for confusion due to suspected toxic-metabolic encephalopathy and intractable vomiting. Two months earlier, in another hospital, he had undergone surgery for adenocarcinoma of the papilla of Vater, with "ampullectomia and reimplantation of the papilla on stent of the common bile duct (Cattel tube) and stent of Wirsung, and gastrodigiunoanastomosis". The onset of emetic symptoms was reported within a few days after surgery.

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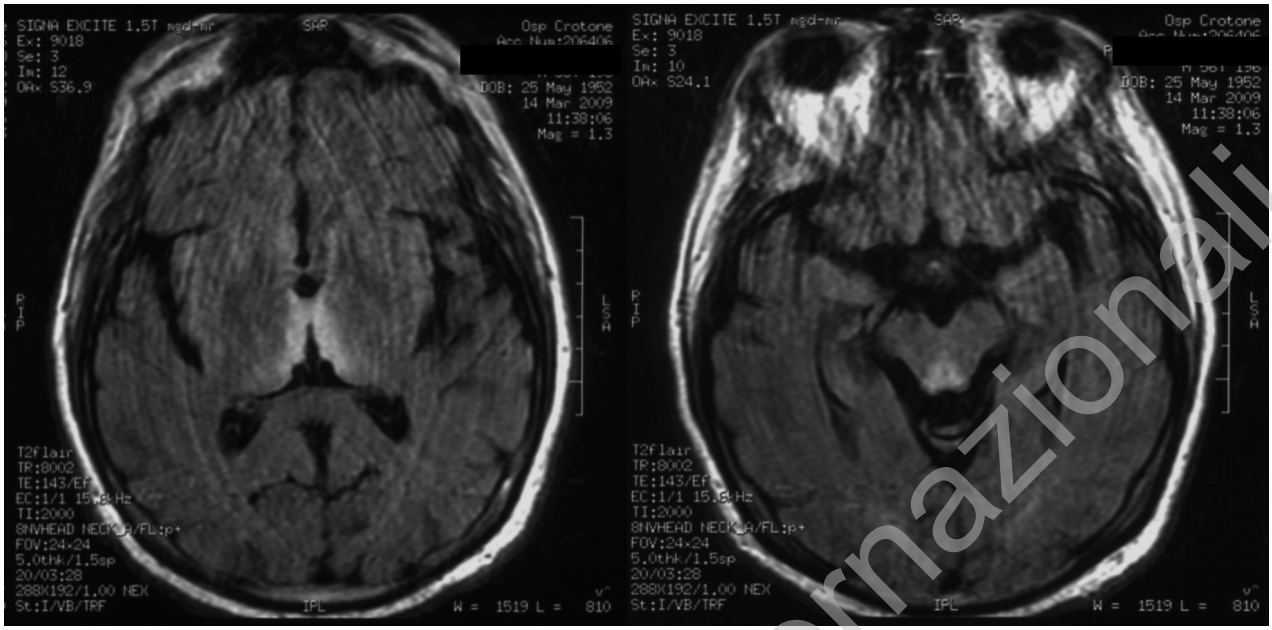

Fig. 1 - MRI hyperintensity of signal in long TR sequences in the posterior pons, periaqueductal midbrain, quadrigeminal lamina, thalamus and hypothalamus and extended up behind the walls of the third ventricle, compatible with toxic-dysmetabolic Wernicke's syndrome.

The clinical neurological examination showed an alert bedridden patient, conscious, fairly cooperative, mildly disoriented in time and amnesic compared to recent events, with lateral gaze nystagmus to the right and left, horizontal conjugate gaze deficits in both directions, mild dysphasia, drift of upper limb to the Mingazzini test (unenforceable the same by lower limbs), dysmetria to the index-nose test, severe bilateral flaccid paraparesis, osteotendineous reflexes everywhere absent, superficial and deep hypoesthesia of the lower limbs, slowed speech.

Brain MRI was performed (Fig. 1), documenting "hyperintensity of signal in long TR sequences in the posterior pons, periaqueductal midbrain, quadrigeminal lamina, thalamus and hypothalamus and extended up behind the walls of the third ventricle, compatible with toxic-dysmetabolic Wernicke's syndrome". EEG described a "marked slowdown and widespread brain bioelectrical activity". An upper digestive tract endoscopy highlighted "cardial incontinence and esophagitis, bile reflux gastritis with plenty of stagnation gastric as result of ampullectomia".

After surgical consultation, for the suspected diagnosis of afferent loop syndrome with uncontrollable vomiting and multivitamins deficiency complicated by Wernicke encephalopathy, it was decided to transfer the patient to our unit surgery for undergoing him to another surgical procedure. Opening the abdominal cavity is highlighted the presence of anisoperistaltic gastrojejunostomy with afferent loop very short and dilated. We therefore decided to pack entero-enteric latero-lateral anastomosis (Braun's procedure). The postoperative was uncomplicated, with disappearance of emetic symptoms, gradual recovery of oral feeding and discrete recovery of cognitive function and mnesic confusional state, decreased upper limb weakness.

Brain MRI (Fig. 2) performed in the 9th postoperative day documented a "normal ventricular-cisternal system with normal morphology and size, with no more obvious areas of altered signal dependent on the brain parenchyma". Resigned in the 18th postoperative day, the patient performed physical therapy cycles with fair recovery of motor function whereas cognitive functions were alternated (shiny and amnesic with confusion phases). The patient died 9 months after the first surgery for metastasis from the ampullar tumor.

Discussion

Today thiamine deficiency is a re-emerging problem in non-alcoholic patients and it may develop in surgical patients with risk factors such as malnutrition, prolonged vomiting and long-term high glucose concentration parenteral nutrition (11).

Afferent loop syndrome

Afferent loop syndrome is an uncommon complication of gastroenterostomy reconstruction after gastrectomy. It's caused by an intermittent obstruction of the afferent loop after a Billroth II resection or a gastric-jejunostomy, also a long time after surgery (12).

Most cases of afferent loop syndrome are due to obstruction caused by adhesions, stomal stenosis, kinking at the anastomosis, intussusception, internal hernia, malignancy or inflammation surrounding the anastomosis. Food passes in the afferent loop causing the liberation of colecistochinina-pancreozima and secretina that, in turn, stimulate the bilious and pancreatic secretion which pours in jejunal loop. The afferent loop is place of stagnation up to a maximum of fullness (about 150-200 cc) untill suddenly it empties, pouring the enteric juice with bile and pancreatic secretion in the stomach. Often the cause of this syndrome is due to excessive lenght of the afferent loop or to its shortness with the presence of angles.

The abrupt depletion of the loop in the remnant of the stomach is followed by bilious vomiting, generally 1-2 hours after the meal, usually preceded by nausea and sometimes headaches. Sometimes you can add sudden

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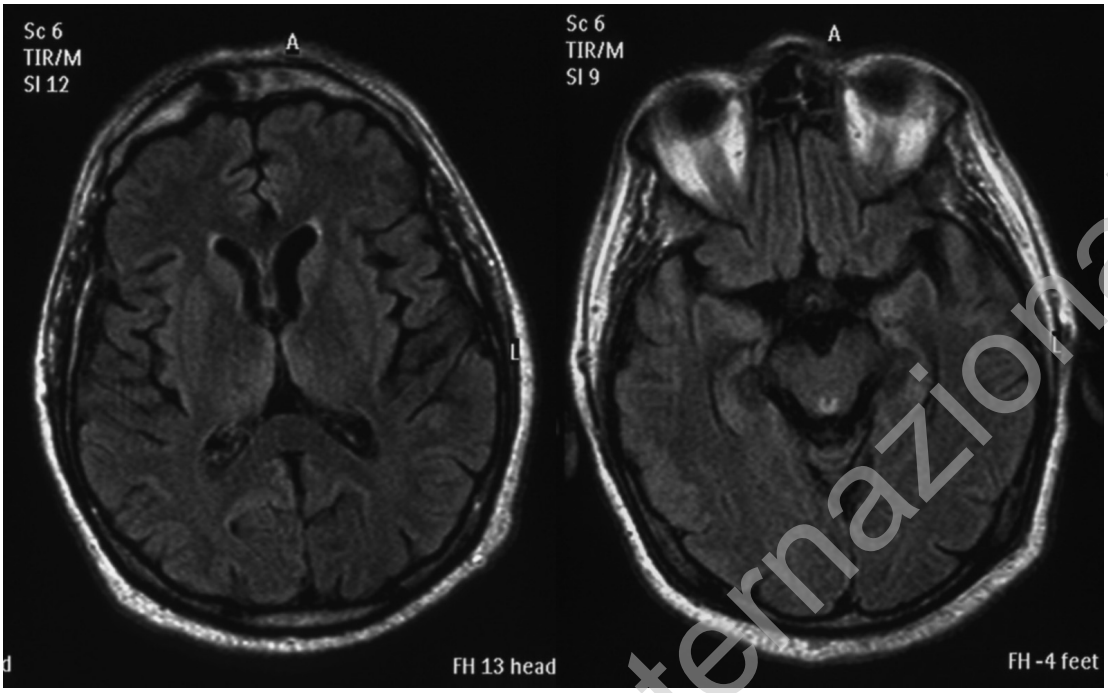

Fig. 2 - MRI after surgery for afferent loop syndrome. Normal ventricular-cisternal system, with no areas of altered signal.

fatigue, pallor, sweating, dizziness and, as outstretched loop's sign, also pain irradiated in epigastrium. The obstructed afferent loop appeared on CT as a fluid-filled tubular mass with a diameter ranging between 4 and 8 cm (13-15). Often the diagnosis is not suspected clinically since patients may presents symptoms many year after the initial surgery too (12).

Wernicke's syndrome

Free thiamine, which is converted into thiamine diphosphate after going through the blood-brain barrier, is known as one of the cofactors which aid in energy metabolism within the brain. Thiamine deficiency causes deprivation of transketolase which is a thiamine-dependent enzyme in pentose phosphate pathway. As a result, pyruvic acid and lactic acid accumulate within the brain, causing activation of cellular membranes and concentration of nerve conduction molecules, thereby damaging the nervous system.

Thiamine deficiency is known to cause Wernicke's syndrome, but the pathophysiology is still uncertain. The cerebellar vermis is sensitive to thiamine deficiency, in particular the Purkinje cells. The Purkinje cells have widespread connections to the cerebral hemispheres and the extrapyramidal connections. The damage to the Purkinje cells can result in motor problems, including ataxia and also impairment of executive functions, learning and memory. Wernicke's encephalophaty (WE) can also alter sympathetic outflow. It can result in postural hypotension, syncope and mild hypothermia. When learning and memory deficits are associated with WE the syndrome is called Wernicke-Korsakoff Syndrome (WKS). Confusion is the most common initial presentation of WE. Patients can be apathetic, inattentive, indifferent to their sorroundings or agitated. MRI can confirm the diagnosis by hypersignal images most frequently along the bulbo-pontine tegmentum, the midbrain's tectum, the periacqueductal location, the thalamus and mammillary bodies (11,16).

When Wernicke encephalopathy is correlated to gastric surgery, it usually develops weeks or months after the procedure and in rare cases also after many years. The development of Wernicke encephalopathy more than 20 years after surgery is extremely rare and all the reported cases pertain to Japanese patients (17,18). A work of 2006, published on *Archives of Neurology* (19), describes the first case of a white patient who developed WE 28 years after gastric surgery. He had undergone partial gastrectomy with gastrojejunostomy because of a peptic ulcer and had been receiving monthly intramuscular injections of vitamin B12.

Conclusion

The incoercibile vomiting and the hydro-electrolytic and multivitaminic depletion due to Billroth II gastrectomy or gastrojejunostomy anastomosis are the cause of Wernicke's syndrome development described in our patient.

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In accordance with these observations, we consider opportune in these cases to re-establish the gastro-intestinal continuity in association with an entero-enteric Braun anastomosis. This second anastomosis favours the afferent loop's outflow directly to the intestine avoiding the gastric outstretching and the vomiting.

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