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# A case of vomiting in an anorexic achalasic patient

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**Abstract. – Wernicke's encephalopathy is a neurological disorder caused by thiamine (vitamin B1) deficiency characterized by vertigo, ataxia, and mental confusion. Wernicke's encephalopathy has a causative association with alcoholism but recently there has been an increased prevalence also in other clinical conditions. In literature potentially fatal Wernicke's encephalopathy onset in an advanced achalasia has been previously reported only once. We describe for the first time an improvement of achalasic symptoms in a young patient affected by end-stage achalasia and anorexia nervosa (coming from ineffective Heller-Dor myotomy) after vitamin B1 supplementation. This case report suggest a potential positive impact of B1 supplementation on end-stage achalasic patients and requires systematic studies to confirm this observation.**

*Key Words:*
Achalasia, Anorexia nervosa, Wernicke syndrome, Thiamine.

## Introduction

Vomiting is one of the more common upper gastrointestinal symptoms in the clinical practice either of central, peripheral or metabolic origin. We report of a young woman initially referring to our hospitals' Emergency Department for upper gastro-intestinal symptoms impairment in history of anorexia nervosa syndrome not responding to the pharmacotherapy; it was subsequently made radiologic diagnosis of subsiding severe achalasic condition ineffectively treated by surgery with further worsening of the symptoms. A Wernicke's syndrome was finally diagnosed by biochemical and MRI imaging and successfully treated by thiamine supplementation with the consensual unexpected achalasic dismotility pattern improvement.

### Case Presentation

A 26 year-old woman was referred to the Emergency Department of our Hospital for the acute worsening of a long-lasting dyspepsia with recurrent vomiting, advanced dehydration with defeated nutritional status associated with anorexia nervosa. The patient was previously treated by anti-psychotic drugs both with a continuative psychological support without any clinical improvement.

During the diagnostic evaluation endoscopy revealed an abnormal dilatation of the esophagus with a tight stenosis at the distal third of the organ. No pathological intraluminal lesions were observed. The endoscopic pattern was deemed to be indicative of achalasia[1]. Barium swallow gastric examination confirmed the abnormal oesophageal and gastric transit and showed the distension of the oesophagus with complete aperistalsis. In addition a tight cardial stenosis was confirmed with the typical "rat's tail" radiological features (Figure 1A). So far the patient was candidate for surgery. Before the surgical procedure an abdominal CT-scan was performed. The esophagus appeared extremely enlarged in all its intra-thoracic course and in particular in the distal pre-cardial tract (up to 8 cm from the cardia). At the level of the diaphragmatic hiatus a grinding "halt of caliber" was clearly detected without any pathological external lesions. Due to the defeated conditions of the patient an oesophageal manometric investigation was not underwent. All the routine laboratory's tests were within the normal range.

According with the poor clinical status a total oesophagectomy, considered as the gold standard for this advanced stage of achalasia, was considered not feasible. Thus an extramucosal miotomy of the distal esophagus (up to 4 cm from the cardia) with Heller-Dor fundoplicatio was performed. No complications occurred after the pro-



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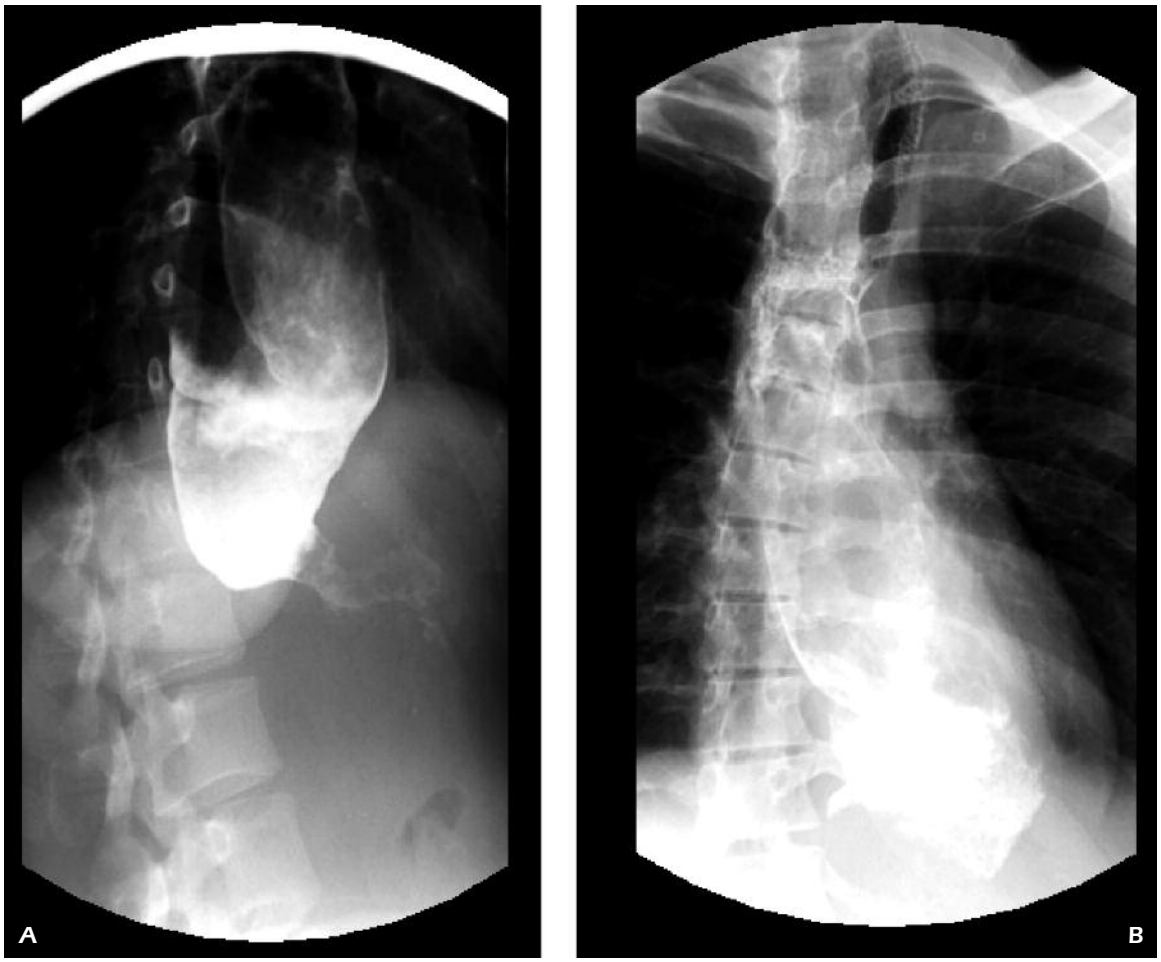


**Figure 1.** Transit before *(A)* and after *(B)* the extramucosal myotomy sec. Heller-Dor and thiamine supplementation.

cedure until the fifth post-operative day when the patient showed a progressive decay of the mental status associated with dyplopia, nystagmus, dysmetria, ataxia, deep asthenia and a loss of short-term memory over than vomiting not responding to metoclopramide and/or ondansetron intravenous administration. Respiratory (hypoxic encephalopathy) and metabolic causes were initially excluded while the neurologic status rapidly impaired turning from a mild confusion to coma. Occult post-operative septic status was also excluded according to the laboratory tests and a new abdominal CT scan. The brain CT scan excluded intracranial bleeding or stroke with the evidence of a bilateral thalamic hypodensity. After neurologic counseling brain MRI showed hyper-intensity of both thalamic system and either the mamillary bodies as well as the white periaquaeductal matter on long TR and FLAIR images (Figure 2). The combination of MRI imaging and neurological signs was indicative of a Wernicke's syndrome (WE) diagnosis, as lately also confirmed by detection of low plasma thiamine concentration [5 mg/dl high performance liquid chromatography (HPLC)][2,3]. Intramuscular administration of thiamine chlorohydrate (300 mg daily for 2 weeks) together with intravenous nutritional support (1700 Kcal/day) were promptly started with a significant clinical improvement since the second treatment day. The patient restarted the oral feeding irrespective of severe achalasic condition, and a significantly improved oesophago-gastric motility pattern was also observed at the radiographic study (Figure 1B).

## Discussion

Thiamine is crucial for several biochemical processes such as intermediate carbohydrate and

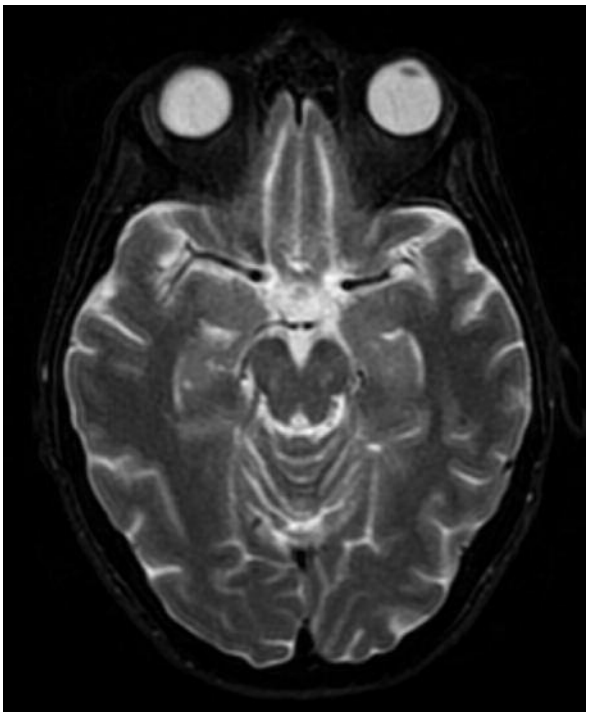

**Figure 2.** Brain MRI long TR and FLAIR images showing hyper-intensity of both thalamic system and either the mamillary bodies as well as the white peri-aqueductal matter.

lipid metabolism over than amino acids and glucose-derived neurotransmitters synthesis. Any condition of unbalanced nutritional status lasting for 2-3 weeks can lead to thiamine depletion and brain lesions usually located in vulnerable regions with high thiamine content and turnover (e.g. diencephalic and brainstem areas)[2]. WE is a medical emergency thus thiamine supportive therapy should be immediately initiated in order to prevent irreversible brain damages invariably leading to death (acute mortality rate of about 20%) or to the chronic form of the encephalopathy (Korsakoff's syndrome) in up to 85% of survivors[2]. The presumptive diagnosis of WE can be confirmed by assessing the thiamine status by the direct measurement in whole blood or by performing the erythrocyte transketolase activation test although these measurements are limited by low specificity and technical difficulty[3,4]. MRI is currently considered the most valuable method to confirm the WE diagnosis[2]. Gastrointestinal causes of WE include: peptic ulcer, acute pancreatitis, oesophageal metastasis, and gastric or oesophageal carcinoma[5]. A combination of ophthalmoplegia, nystagmus, and ataxia, considered to reflect a Wernicke-Korsakoff syndrome, has already been described in a patient with anorexia nervosa after laparoscopic cardiomyotomy for achalasia[6,7].

The association between achalasia and anorexia nervosa and/or misdiagnosis of the two disorders have been already described[8]. Dysphagia is the initial and main clinical feature of achalasia. Other aspecific symptoms, such as vomiting and weight loss are also common. There is an usual elapse before the disease is diagnosed[6] throughout achalasia can be easily confounded with anorexia nervosa. Nevertheless differential diagnosis between achalasia and anorexia nervosa is not always obvious as long oesophageal motor disorders are common in patients with a primary diagnosis of anorexia nervosa[9]. For example patients with eating disorders frequently have gastric emptying abnormalities causing bloating, postprandial fullness, and vomiting. These symptoms usually improve with refeeding but sometimes prokinetics administration may be necessary. In addition wilful avoidance of food and spontaneous or self-induced vomiting have been reported in patients with echolalia[10,11]. Thus, many gastrointestinal diseases may present like eating disorders. Rosenzweig et al[12] consider this misdiagnosis to be related to delay in obtaining appropriate investigations or misinterpretation of their results. Other authors suggest that a careful clinical history can localize gastrointestinal motility disorders together with appropriate diagnostic tests use[13] according to the recognition of two main groups of symptoms. First, dysphagia, odynophagia, heartburn, and reflux have oesophageal origins and occur in achalasia. In this case the appropriate tests are barium-swallow endoscopy and oesophageal manometry and/or scintigraphy. The second group of symptoms includes nausea, vomiting, anorexia, bloating, and abdominal pain, all symptoms of motility disorders of the stomach and small intestine.

So far evidences from the literature do not include a clear causal relationship between achalasia, anorexia, malnutrition and consequent vitamin deficiency.

The clinical case above reported support for the first time the possible relationship between Wernicke syndrome due to chronic vitamin deficiency and "nervosa/achalasic" anorexia as common cause of vomiting in achalasic patients. Paradoxically an impaired achalasia may be both a "clinical sign" of vitamin deficiency due to an organic and nervosa anorexia and a cause of it turning all together in these pathophysiologic vi-

cious circle. In fact the non-functional recovery after surgical oesophageal myotomy of the dysphagia above reported asked for further and systematic investigation about metabolic conditions possibly underlying the dyspepsia.

In conclusion, we can be consequence and cause of advanced status achalasia and anorexia nervosa. Vitamin B1 supplementation could represent an "add-on" future source for the treatment of advanced achalasia together with the surgery staying the gold standard if this observation will be supported by systematic studies.

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