Wernicke encephalopathy and Systemic sclerosis: rare association of rare conditions

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Background: Wernicke encephalopathy (WE) is a neurological syndrome due to thiamine deficiency. The association between WE and Systemic Sclerosis (SS) has not previously described. The case of a patient affected by SS developing WE is reported.

Case report: A 67-year-old woman with SS complained of nausea, vomit and progressive weakness for 1 month. She was not a drinker and previous infections were not reported. A week before hospitalization she experienced acute onset of diplopia, disorientation and personality changes. Neurological examination disclosed complete bilateral external ophthalmoplegia, distal-dominant weakness and dysmetria in all four limbs, hypo-elicitable lower limb reflexes. Nerve-conduction study revealed sensory-axonal polyneuropathy. Gastrointestinal endoscopy showed gastrectasia with absence of propulsive waves and consequent gastric fluid stasis (Figure 1). Brain MRI depicted hyperintense bilateral symmetrical lesions involving peri-aqueductal region and extending superiorly to thalami and mammilary bodies and inferiorly to the floor of fourth ventricle, cerebellar vermis and medulla (Figure 2). A diagnosis of WE was established. Patient was treated with intravenous thiamine 100 mg/day, with recovery and disappearance of brain lesions in 7 days.

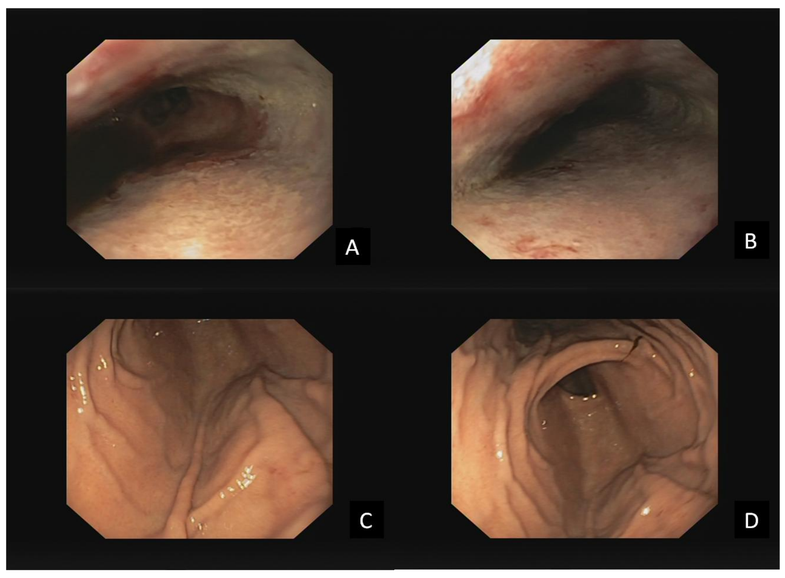

Figure 1. Endoscopic examination of esophagus showed parietal sclerosis (A) causing dilatation and erosive lesions (B). Endoscopic view of the gastric walls documented gastrectasia with absence of propulsive waves (C) and consequent gastric fluid stasis (D).

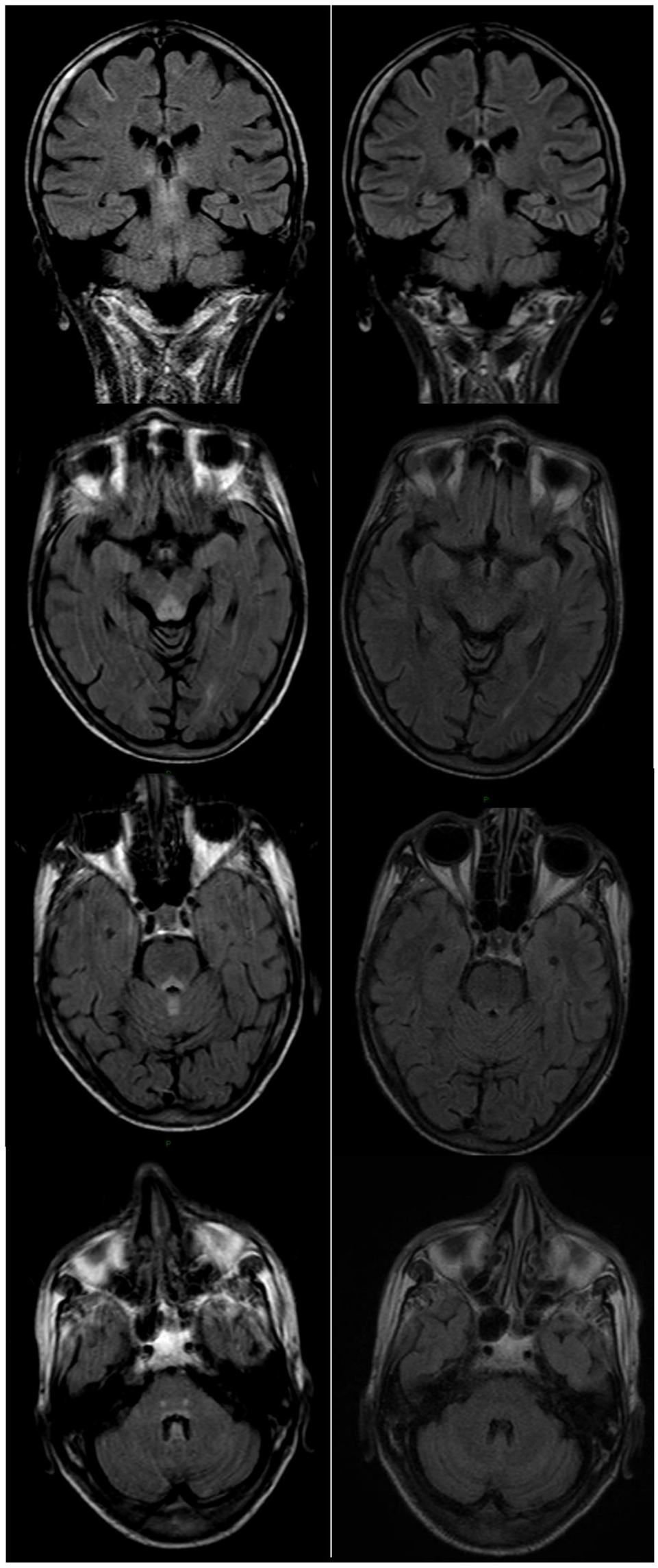

Figure 2. Brain T2-FLAIR MR scan at admission showed a symmetrical bilateral hyperintensity area surrounding the Sylvian Aqueduct and extending also to the thalami and the mammilary bodies. Pathological areas were also visible around the floor of the forth ventricle, involving the cerebellar vermis, the pons and the medulla (**left**). MR scan performed seven days after starting intravenous thiamine administration demonstrated the disappearance of hyperintensity areas (**right**).

Discussion: WE cardinal signs are ophthalmoplegia, ataxia and disorientation. Causes of thiamine deficiency are alcoholism, orofacial, gastric and colon cancers, gastric-surgery, drug-induced gastritis, peptic ulcer, biliary colics, Crohn's disease, pancreatitis, hyperemesis gravidarum, sepsis, coma, renal diseases, acquired-immunodeficiency disease, chronic febrile infectious diseases, thyrotoxicosis, anorexia, bulimia and binge-eating disorder (1). Diagnosis is mainly supported by the response of neurological signs to parenteral thiamine, that should be immediately started in order to avoid irreversible brain damage. SS is an autoimmune chronic disease characterized by deposition of connective tissue in skin and internal organs, particularly esophagus, stomach and bowel, with consequent dysphagia, emesis and malabsorption due to delayed gastric emptying (2). In our case, SS gastrointestinal manifestations and related malabsorption are responsible for thiamine deficiency. The case of a patient complained of SS, pneumoperitoneum and cystoides intestinalis, developing Korsakoff's psychosis has been previously reported (3). In this case malabsorption syndrome was due to different gastrointestinal disorders. SS should be considered as a clinical condition affecting the correct absorption of thiamine, when clinical signs and neuroradiological features lead to diagnosis of WE, in absence of other causes of malabsorption.

Conclusion: To our knowledge, this is the first case describing the pure association between WE and SS, without other related medical conditions. Acute onset of altered consciousness and impaired ocular motility in a SS patient should rise the diagnostic suspicious of WE to start precociously the appropriate therapy.

References

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