Unusual presentation of Wernicke Encephalopathy without clear risk factors

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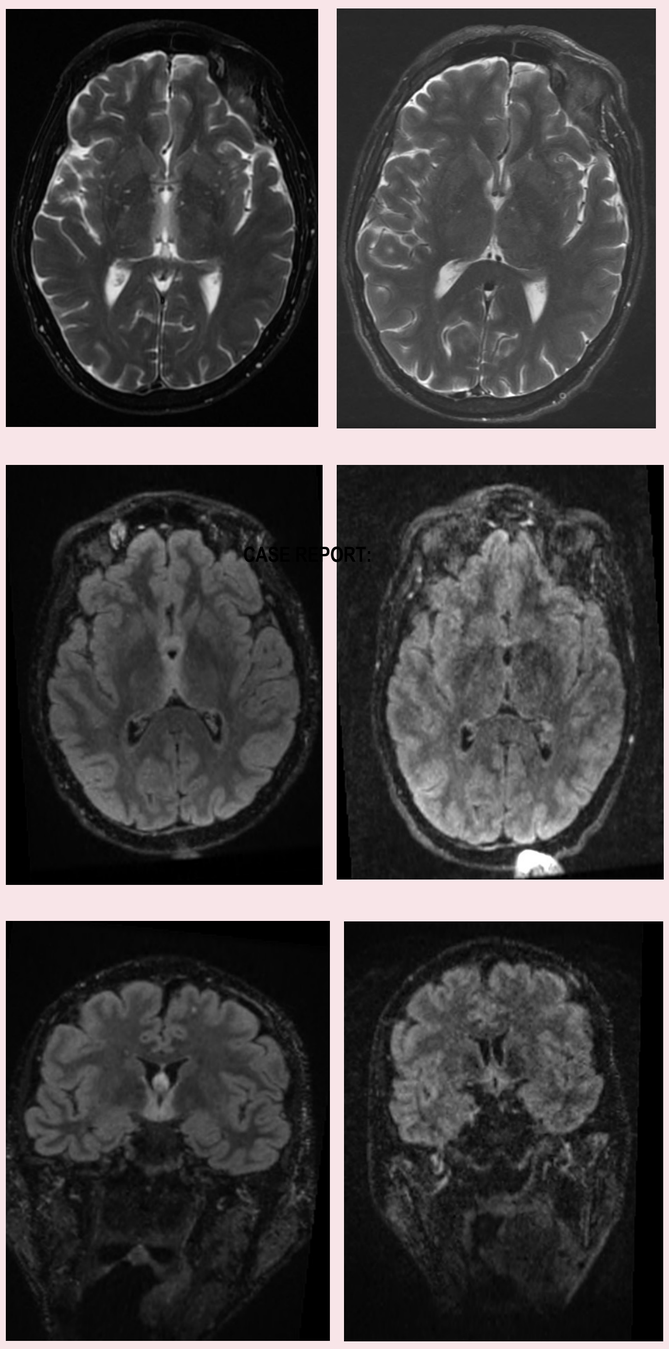
INTRODUCTION

Wernicke encephalopathy (WE) is a medical emergency characterized by ataxia, confusion, nystagmus and ophtalmoplegia, related to a thiamine depletion, mainly caused by alcoholism or by a reduced thiamine intake or absorption. If untreated, WE can lead to Korsakoff syndrome and even to death. We describe a case with an unusual presentation.

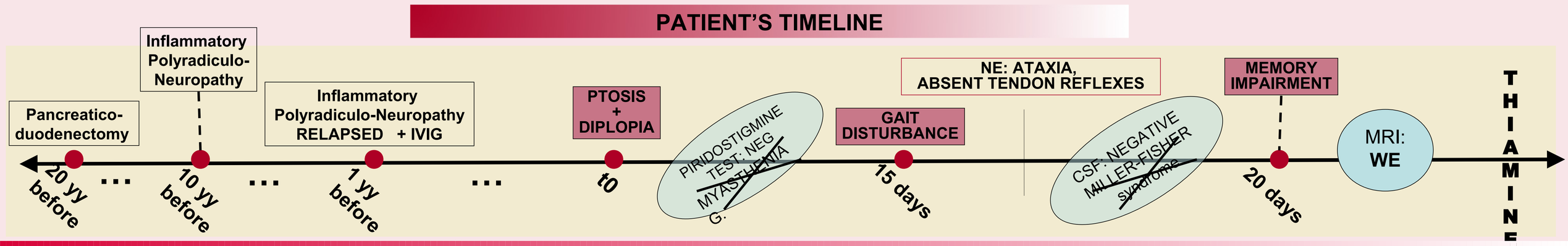
CASE REPORT:

A 53-year-old man came to our attention, as an out-patient, with **bilateral palpebral ptosis** and a **slight horizontal diplopia**, with onset and progressive worsening in the previous 7 days. In his medical history there were a pancreatico-duodenectomy due to ampullary cancer performed 20 years before and an inflammatory polyradiculoneuropathy occurring 10 years before and relapsed in the current year, after a gastrointestinal infection and improved with IVIG.
He was submitted to CT scan (negative for focal lesions) and tensilon Test with a weakly, subjective, response of ptosis; thus we asked for AchR Ab and Musk Ab assay and we prescribed a trial with Piridostigmine and corticosteroids. Two weeks later he returned to clinic complaining the worsening of ptosis and diplopia, and the onset of gait disturbances; blood tests showed serum negativity for Myasthenia. Neurological examination showed: bilateral ptosis, sideward gaze palsy and vertical ophtalmoparesis, absent tendon reflexes and ataxic gait.


CASE REPORT:

We hospitalized him and performed a lumbar puncture, suspecting another poliradiculo-neuropathy relapse, in the variant of a Miller-Fisher syndrome. CSF analysis was normal. In the next days he started to lose the ability of standing and became confused, less oriented, with memory impairment.
MRI revealed bilteral lesions in thalamus, hypothalamus, mammillary bodies and periventricular gray matter, supporting the diagnosis of WE. Thiamine iv at high doses led to partial resolution of his ptosis, gaze palsy and gait ataxia. After 10 days he was discharged: at that time he was oriented but not completely recovered as to memory. MRI, repeated one month later, showed a significant improvement. In parallel his memory had further improved as well. The patient and his relatives denied any alcohol consumption. In this case, Thiamine depletion was likely due to the pancreasectomy performed 20 years before which, for unknown reasons, caused such a depletion only after a long time

PATIENT'S TIMELINE



CONCLUSIONS:

This case report can be usefull for clinicians dealing with WE, because it contains some unusual aspects including bilateral ptosis as first symptom complained by the patient, and a significant time delay between gastrointestinal surgery and clinical presentation of the disease.

Bibliography: Simon J Scalzo, Stephen C Bowden, Margaret L Ambrose, Greg Whelan, Mark J Cook "Wernicke-Korsakoff syndrome not related to alcohol use: a systematic review" J Neurol Neurosurg Psychiatry 2015;0:1–7

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