



CLINICAL CASE

Pellagra: An old, new differential diagnosis in dementia patients?

Pelagra: un antiguo y nuevo diagnóstico diferencial en pacientes con demencia

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Case

An 85-year-old male was seen in outpatient care and affected by severe mixed dementia (Alzheimer's disease coexisting with vascular dementia) associated with mixed anxiety-depressive disorder, anorexia, and capricious intakes that triggered a caloric-protein malnutrition. As for the comprehensive geriatric assessment, she presented moderate dependence for the basic activities of daily living (Barthel Index 45/100), did not perform instrumental activities, and had a 24-h caregiver. His usual treatment included vortioxetine, quetiapine, amlodipine, atorvastatin, furosemide and acetyl salicylic acid.

In the previous months the patient had presented a worsening of cognitive functions, with somatic complaints such as epigastralgia which were attributed to progression of Alzheimer's disease. Consulted due to a symmetric, scaly, sunburn-like, hyper-pigmented erythematous plaques extending from his dorsal parts of arms and hands and around his neck (Fig. 1a).

The differential dermatological diagnosis included lupus, polymorphous light eruption, photocontact dermatitis, pseudoporphyria cutanea tarda and pellagra. The family reported that the patient had not been exposed to any new chemicals, that no changes had been made to his usual medical treatment, and that he had no personal or family background of photodermatitis. A blood test was performed, no alterations were found in the acute phase reactants, nor in the liver function markers. A decreased vitamin B6 levels (<9 nmol/L, normal 23–173 nmol/L) was detected, niacin determination was not available in our laboratory. No skin biopsy was performed because the patients' families refused. The onset of new psychological and behavioral symptoms associated with dementia, such as hetero-aggressive behavior, and depression in a previously clinically stable patient; and the presence of digestive symptoms such as epigastralgia without objectionable diarrhea contributed to the possibility of pellagra. We started nicotinamide with dermatological improvement 2 weeks after the start of treatment (Fig. 1b).



Fig. 1. Clinical image at presentation (A) and after 2 weeks of treatment (B).

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Pellagra is a nutritional disorder characterized by the four Ds: diarrhea, dermatitis, dementia and death as a result of a severe deficiency of niacin (B3).¹ The symptoms evolve at different intervals over time making it hard to see the pattern and apply diagnostic.¹

Low levels of niacin lead to deficiencies of the coenzymes nicotinamide adenine dinucleotide (NAD) and NAD phosphate (NADP). These are involved in cell anabolism, that is why pellagra mainly affects cells with a high replication rate, such as skin or enterocytes; or in those cells with high energy consumption, such as neurons. This explains the symptomatology of “the 4 Ds”, impairment of enterocyte function can cause diarrhea, epigastralgia or achlorhydria; as for the effects on central nervous system cells, manifestations such as cognitive impairment, depression, or distal tremor are among other possible ones described.³

The dermatological histopathology of the disease is nonspecific, consisting of dilated blood vessels and mild inflammation.²

In developed countries pellagra continues to occur but is limited to high-risk groups such as HIV infected patients, alcoholism and malnourishment.^{2,4} Some case series speak of an approximate incidence of 0.5 cases per 100,000 population, however it could be higher due to underdiagnosis.² In this case, eating habits that were conditioned by dementia which created a niacin deficiency. This is the reason why pellagra could be considered in the differential diagnosis of sun-exposed areas lesions in neurocognitive disorders.

Authors' contributions

Dr. Miralles takes responsibility for the paper as a whole. Drafting of the manuscript: Dr. Miralles, Dr. Betancor. Critical revision of the manuscript for important intellectual content: All authors.

Funding

This manuscript was not sponsored or funded.

Conflict of interest

None reported.

Acknowledgement

We thank the patient's family for grating permission to publish this information.

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