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Case Report

PELLAGRA-LIKE DERMATITIS - A CASE REPORT

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Abstract

Pellagra-like dermatitis refers to a skin condition resembling the dermatologic manifestations of pellagra, which is a systemic disease resulting from niacin (vitamin B3) deficiency. Vitamin B3 is needed for several metabolic processes, cell signaling and DNA repair. This condition is characterized by a photosensitive rash that typically affects sun-exposed areas. The classic skin changes include symptoms known as the 4Ds: dermatitis, diarrhea, dementia and death. The main causes of pellagra are: nutritional deficiency of niacin, restrictive diets, chronic alcoholism, gastrointestinal malabsorption, metabolic disorders and certain medications.

We present the case of a 55-year-old farmer, with megaloblastic anemia and a seven-month history of skin changes, including erythematous lesions on sun-exposed areas such as the face, neck, bilaterally on forearms, dorsum of the hands and feet, along with itching and desquamation. Gastrointestinal symptoms included intermittent diarrhea, weight loss, neurological symptoms, and anxiety. Laboratory tests revealed anemia, hypoproteinemia, low levels of folic acid and serum iron. Skin biopsy results were consistent with pellagra, but did not exclude contact dermatitis. Treatment with niacin, folic acid, vitamin B12 and iron led to significant clinical improvement. Regular follow-up visits with hematology, gastroenterology, neurology specialists, and nutritional therapy were recommended.

While pellagra is rare in modern clinical practice due to better nutrition, it still occurs sporadically. Diagnosis relies on the classic 4D features, laboratory findings and histopathological features. Despite its rarity, pellagra-like dermatitis should be considered in differential diagnoses for dermatological and gastrointestinal symptoms. Early diagnosis and treatment can result in significant improvement.

Keywords: pellagra-like dermatitis, pellagra, deficiency of niacin (vitamin B3), diarrhea, dementia

Introduction

Pellagra-like dermatitis refers to a skin condition resembling the dermatologic manifestations of pellagra, which results from niacin (vitamin B3) deficiency. First recognized by Gaspar Casal in 1735, pellagra remained a major widespread cause of death until the early 20th century [1,2]. The name ‘pellagra’ is derived from the Italian ‘pelle agra’ meaning sour or rough skin. It is a systemic nutritional disorder resulting from niacin (vitamin B3) deficiency

and/or its precursor tryptophan[1,3], which can have widespread effects on the skin, gastrointestinal system and central nervous system.

The main causes of pellagra are: nutritional deficiency of niacin; restrictive diets, chronic alcoholism; gastrointestinal malabsorption (inflammatory bowel disease - Crohn disease, ulcerative colitis, jejunoileitis, prolonged diarrhea, chronic colitis, cirrhosis), gastrointestinal surgery including gastroenterostomy or subtotal gastrectomy; certain medications (antidepressants, azathioprine, chloramphenicol, 5-florourqacil, sulfonamides, carbamazepine and antituberculotics), metabolic disorders, carcinoid syndrome [3,4,5].

Vitamin B3 is crucial for the functioning of cells in the human body and its deficiency manifests itself with symptoms that affect the skin, the digestive and the neurologic systems[6].

Pellagra classically occurs as a set of four symptoms (as 4D): dermatitis, diarrhea, dementia and death, although dementia may not always be present [5,6,7]. Skin changes are often the first sign, appearing in spring and summer, after sun exposure.

The characteristic dermatitis begins like a photosensitive rash that appears in areas exposed to sunlight, such as the face, neck, arms, and hands. It starts as redness (erythema) and swelling (edema), with an acute or intermittent onset, gradually changing into an exudative eruption, with itching and burning. It resembles sunburn in the early stages, sometimes with the presence of vesicles, bullae, erosions and crusts. Then, the skin becomes thickened, lichenified, hyperpigmented, rough with dark scales, crusts and cracks, as well as a sharp demarcation from normal skin [5]. The distribution of changes is typical: the facial region in the form of a butterfly, a clear demarcation in the form of a band around the neck ("Casal's necklace"), extended down over the sternum ("cravat sign), the dorsal part of the hands and fingers extending proximally to forearms ("pellagra gauntlet"), the dorsal part of the feet, up to the ankle with sparing of the palms and the heels ("gaiter of pellagra" or "pellagra boots") [4,8]. Mucous membrane involvement may manifest as cheilitis, angular stomatitis, a red tongue and ulceration of buccal mucosa and vulva [5]. Gastrointestinal symptoms are diarrhea (develops in 50% of cases, watery, mucoid, or bloody, persists longer than 4 weeks), nausea, vomiting, abdominal pain. Neuropsychiatric features such as initial apathy and weakness, headache, insomnia, confusion, irritability, anxiety, tremor, depression, can progress to psychosis, dementia and encephalopathy in later stages. Without treatment pellagra leads to death from multiorgan failure [5].

Diagnosis is primarily made based on clinical features and rapid response to vitamin supplementation. Histhopathological changes in the skin are not specific; there are no chemical tests available for a definitive diagnosis of pellagra. However, low levels of urinary excretion of N-methylnicotinamide and pyridone indicate a niacin deficiency [3].

Treatment of pellagra consists of drugs with niacin or nicotinamide, other B vitamins, zinc and magnesium, as well as a diet rich in calories. Locally, skin lesions are treated with emollients. Prevention is based on nutritional education (food sources of niacin: eggs, peanuts, meat, fish, red meat, and seeds), photoprotection and abstinence from alcohol [9].

Case report

We present the case of a 55-year-old male farmer with multiple systemic and dermatologic symptoms that suggest a diagnosis of pellagra-like dermatitis. He had a seven-month history of painful, pruritic dermatitis, primarily involving sun-exposed areas. The skin lesions presented with erythematous, livid discoloration of the face, with distinctive patterns such as Casal's collar – an erythema surrounding the base of the neck - and the "cravat sign" extending to the sternum, accompanied by discrete desquamation, all of which were highly suggestive of pellagra (Figure 1).

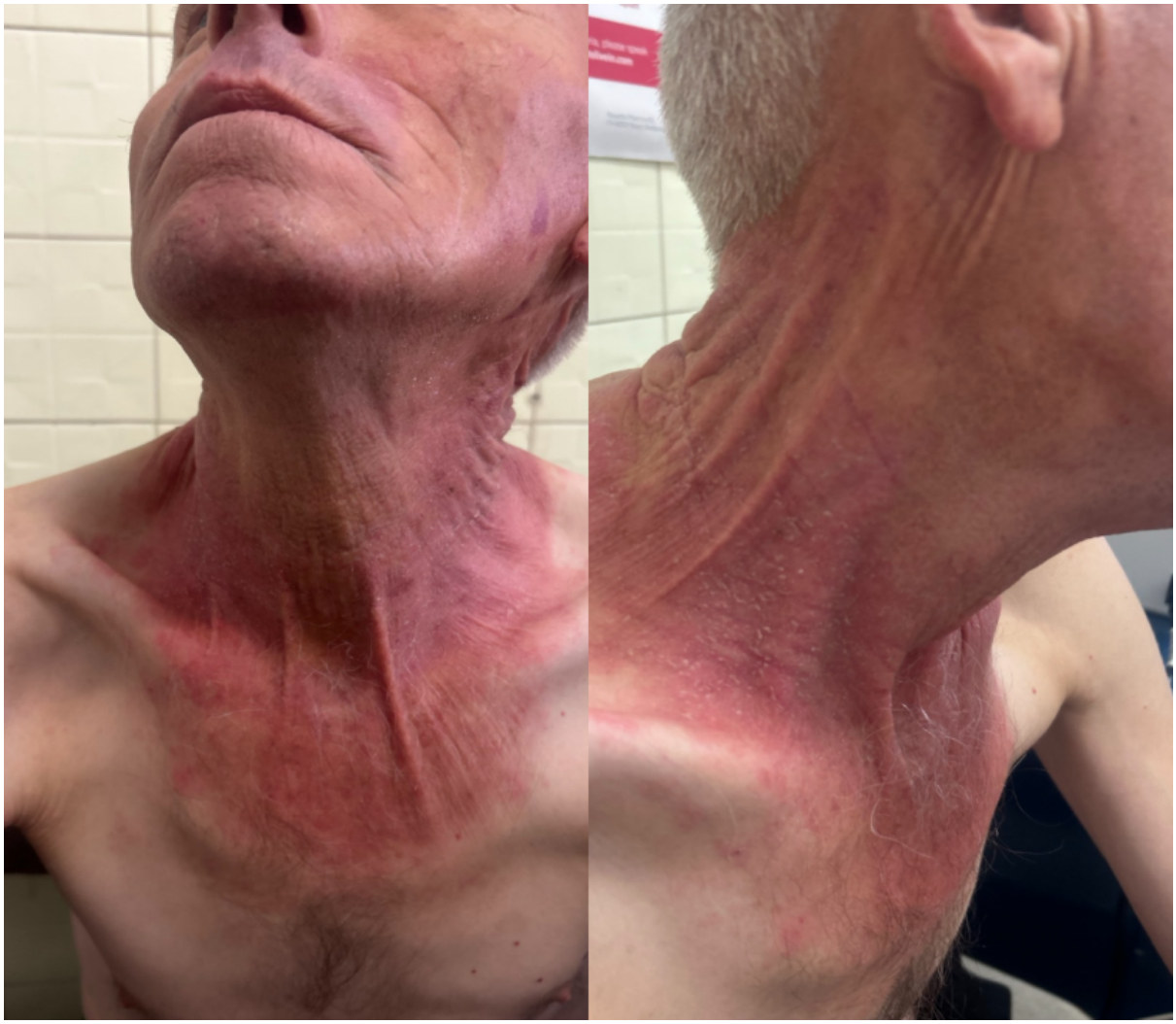

Fig. 1. Erythematous livid skin lesions on the face, the Casal's collar (erythema around the neck), cravat sign (extending to the sternum), with discrete desquamation

Additionally, there were bilateral, symmetrical, erythematous pruritic patches with edema on the forearms and the dorsal surfaces of his hands and feet, while the palmar regions and heels remained unaffected. The affected areas showed papules, erosions, crusts and lichenification with hyperpigmentation (Figure 2).

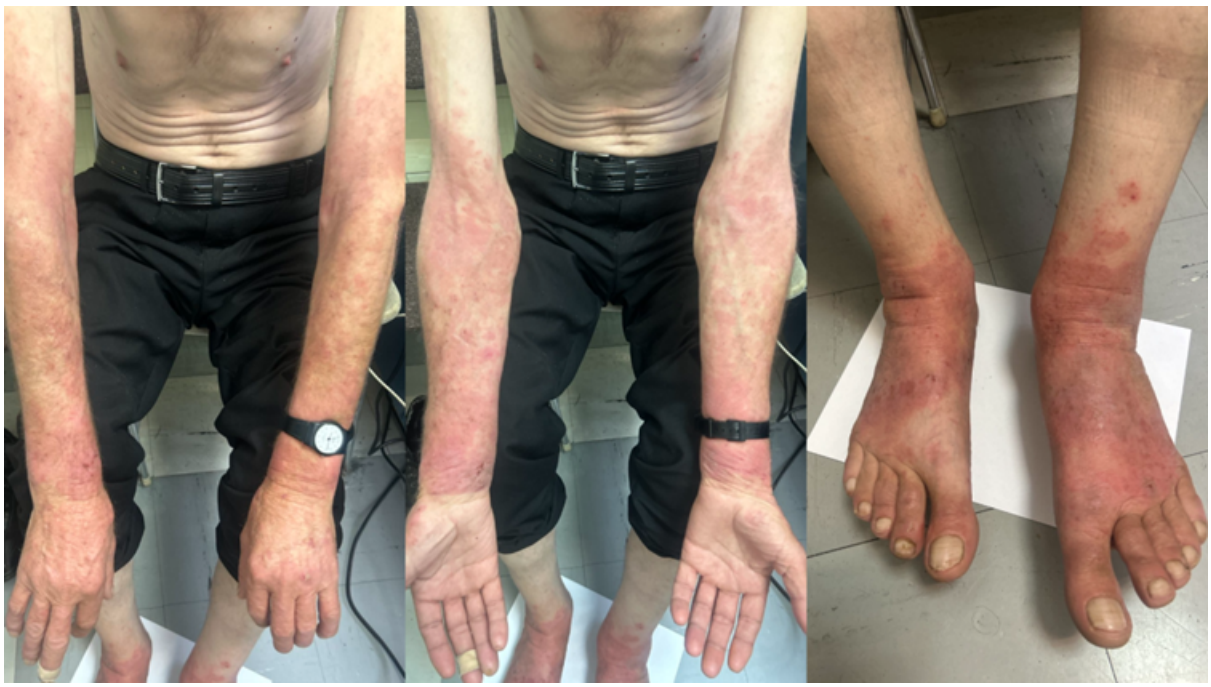

Fig. 2. Bilateral, symmetrical skin lesions on the forearms and dorsal surface of hands with sparing of the palmar region ("pellagra gauntlet") and bilateral, symetrical edema and erythema with lichenified and hyperpigmented skin on feet ("pellagra boots")

The patient reported pruritus and burning, worsened by sunlight exposure. He had a history of megaloblastic anemia and chronic obstructive pulmonary disease. In recent months, he has reported intermittent diarrhea (occasionally with blood), weight loss, anxiety, suicidal ideas. He denied alcohol use.

Laboratory investigations showed several abnormalities in favor of anemia and hypoproteinemia, while the urinary status was normal.

Laboratory findings: anemia with low RBC count (3.62), low hemoglobin (11.30), low HCT 34.60↓; low serum iron (Fe): 2.2 µmol/L; low folic acid: 0.869 ng/mL, hypoproteinemia: low total protein (63.8); elevated liver enzymes (AST 61, ALT 71, LDH 598). Deviations in the values also were in: Neu 9.90 ↑; Mon 0.89↑ ; Limf 15.60%↓; serum creatinine 75↓

Pulmonary radiography (RTG) and abdominal ultrasound were normal.

Histopathological examination of a skin biopsy showed numerous eliminated serosites, mild spongiosis, lymphocytic infiltration and a pseudo-pautrier pattern consistent with inflammatory skin conditions like pellagra, but did not rule out contact dermatitis.

The patient's history of anemia, hypoproteinemia, and vitamin deficiencies (B12, folate, iron) supported this diagnosis. The skin changes, particularly the Casal's collar and cravat sign, along with symptoms worsening with sun exposure, are typical of pellagra. Based on clinical, laboratory and histopathological findings, a diagnosis of pellagra-like dermatitis was established.

Treatment with niacin (B3), folic acid, vitamin B12, vitamin C, and iron resulted in significant improvement in the dermatologic manifestations. Further monitoring and follow-up by a hematologist for anemia management, neurologist, nutritionist as well as gastroenterohepatology specialists for possible gastrointestinal malabsorption or other underlying issues (e.g., Crohn's disease, celiac disease, tumour) is essential. Nutritional counseling, photoprotection and ongoing supplementation were recommended to prevent recurrence.

Discussion

Niacin ingested with food is absorbed through the small intestine into tissues, where it is converted into a coenzyme called nicotinamide adenine dinucleotide (NAD), which, together with enzymes, catalyzes chemical reactions[6].

The coenzyme NAD is involved in the metabolism of carbohydrates, fats and proteins, as well as DNA repair and cell signaling [6,7,10]. Therefore, tissues with high energy needs or rates of cell turnover, such as the skin, intestines and brain, are those affected by pellagra.

The skin changes in pellagra are distinctive and typically manifest as a symmetrical erythematous rash in sun-exposed areas. A hallmark sign, known as the necklace of Casal, presents as a ring of erythema and hyperpigmentation around the neck. Gastrointestinal symptoms, including diarrhea, anorexia, and nausea, along with malabsorption, anemia, and neurological changes, further emphasize the systemic nature of the disorder. Diagnostic confirmation is supported by laboratory findings such as anemia, hypoproteinemia, hypocalcemia, hypokalemia, and hypophosphatemia. While pellagra is rare today, it may be overlooked in populations with chronic alcoholism, liver cirrhosis, anorexia, tumors, or those on certain medications. However, with the rise in restrictive diets like veganism, which lack adequate supplementation, there has been an increase in cases in recent years.

In this case, malnutrition, chronic diarrhea, and megaloblastic anemia likely contributed to niacin deficiency. Gastrointestinal malabsorption compounded this deficiency. The presence of cutaneous signs, gastrointestinal symptoms and laboratory abnormalities strongly supported the diagnosis.

In conclusion, this case demonstrates the importance of recognizing pellagra-like dermatitis as a potential sign of underlying systemic disease, especially in patients presenting with chronic skin lesions, progressive weight loss and neuropsychiatric symptoms.

Pellagra, typically associated with malnutrition or malabsorption disorders, requires a high degree of clinical suspicion due to the absence of specific diagnostic tests. A comprehensive evaluation of dietary intake, gastrointestinal function and neuropsychiatric status is crucial for accurate diagnosis.

The dermatologic findings, combined with the patient's psychiatric symptoms emphasize the need for a multidisciplinary approach involving dermatology, hematology, gastroenterology, and psychiatry. Early diagnosis and niacin supplementation can lead to rapid clinical improvement, preventing potentially irreversible complications and highlighting the vital role of dermatologists in identifying systemic nutritional deficiencies through skin manifestations.

Conflict of interest statement. None declared.

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