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Case Study

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PELLAGROUS DERMATITIS, STILL NOT UNCOMMON: A CASE REPORT

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Abstract
Pellagra usually results from niacin deficiency and presents with the classic triad of dermatitis, diarrhea, and dementia and eventually death, if it is untreated. The disease can occur due to dietary deficiency or due to interference of metabolism or absorption of niacin as in alcoholism, drug intake or gastrointestinal disease. Pellagra due to primary dietary deficiency is considered rare nowadays. We describe an adult female with classical features of pellagra but lacking involvement of face and neck, possibly due to dietary deficiency.

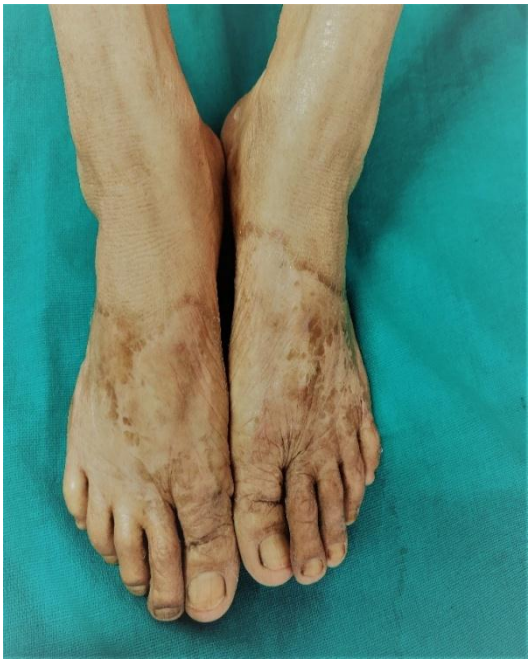
INTRODUCTION

Pellagra is a nutritional disorder caused by a cellular deficiency of niacin or its precursor amino acid, tryptophan. Niacin, also known as nicotinic acid or vitamin B3, is essential for carbohydrate, fat, protein and alcohol metabolism, detoxification of drugs and reactive oxygen species, cell signalling and DNA repair.[1,2] It is classically known as the disease of 4 D's dermatitis, dementia, diarrhoea and death .[3] The term pellagra - from *pelleagra*, Italian for *rough skin* - was first used by Frappoli in 1771 due to its dermatologic manifestations.[4]

Niacin forms into nicotinic acid and nicotinamide, which serve as precursors of two important coenzymes of cellular metabolism, namely nicotinamide adenine dinucleotide (NAD) and NAD-phosphate.[5] NAD is involved in the catabolism of carbohydrates, fats, proteins and alcohol. NADP functions in anabolic reactions, for the synthesis of fatty acids and cholesterol. The reduced form NADP is converted to NADPH and is used in reactions to detoxify reactive oxygen species and drugs. In addition, NAD is also involved in non-redox reactions such as cell signalling and DNA repair.[6]

CASE REPORT

A 61-year-old female patient presented to dermatology opd with darkening of the hands and feet for eight months. The patient initially developed darkening of the right foot, which gradually increased to involve the both feet and both hands. The lesions were associated with mild itching. There was history of fluid-filled and pus-filled lesions over the feet around 2 months ago.The skin lesions developed burning sensation on exposure to sunlight. She was otherwise healthy and hadregular diet consisting of vegetables, pulses and wheat. The bowel habits were normal. She denied any change in her behaviour. There was no history of similar disease in the family. There was no history of intake of any medication or addiction. On general physical examination, the patient had pallor. On cutaneous bilaterally symmetrical, slight hyperpigmented plaques were present over the dorsal aspect of hands and feet extending to the middle part of the dorsum of foot and dorsal aspect of the wrist on the forearm with a clear-cut demarcation from the more proximal normal looking skin resembling the 'Gauntlet' over the wrist and 'boots' over the lower legs (Figure 1,2). Rest of the cutaneous, systemic and mental state examination was within normal range. The patient was found to have hemoglobin of 9g/dl with a microcytic and hypochromic red blood cell indices. Her renal function test and liver function test are with in normal limits. Based on the typical clinical features and lab investigations the patient was diagnosed to be a case of pellagra and was started on nicotinamide 100 mg thrice a day apart from iron and other nutritionalsupplementation. The patient responded dramatically and the lesions disappeared in 3 weeks.



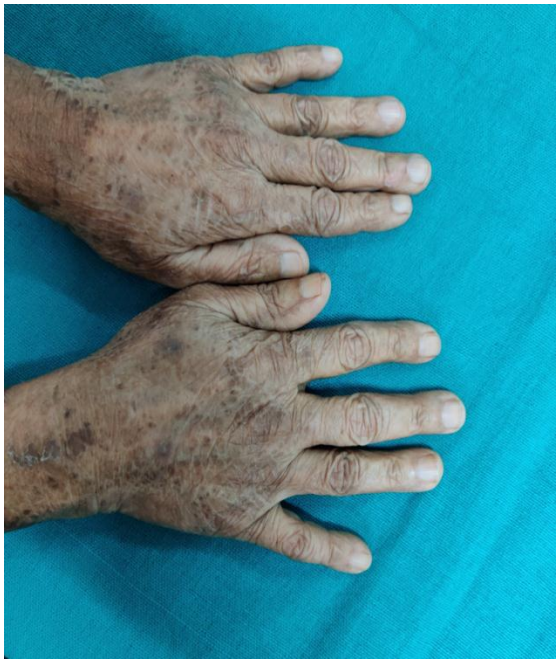


Figure 1&2: shows cutaneous bilaterally symmetrical, slight hyperpigmented plaques were present over the dorsal aspect of hands and feet.

DISCUSSION

The first medical report of pellagra is credited to Gaspar Casal in Spain in 1735 7. He described an exposed-site dermatitis on the dorsum of the hands and feet with a collar-like rash on theupper part of the neck in Asturian peasants.[7]

Pellagra is a chronic disease affecting the skin, nervous system, and gastrointestinal tract usually due to a deficiency of nicotinic acid (Niacin -VitB3) or its precursor tryptophan.[1] Primary pellagra occurs in dietary deficiency of tryptophan or niacin. Secondary pellagra is caused by conditions characterised by interference with absorption or metabolism of tryptophan and niacin, including anorexia nervosa, chronic alcoholism, prolonged diarrhoea, ileitis, colitis, cirrhosis, carcinoid syndrome, Hartnup disease and HIV. Medicines including azathioprine, 5-flurouracil, 6-mercaptopurine and phenobarbitone inhibit the conversion of tryptophan to niacin. Isoniazid is an analogue of niacin and suppresses endogenous formation.[1,2]

The pellagrous dermatitis rash is typically bilateral, symmetrical and limited to exposed sites of solar exposure.[8] It is well-defined and usually most prominent on the dorsum of hands, 'V' of the neck, face, radial aspects of the forearms, and exposed skin on legs and feet.[9] These sites initially become red and swollen; patients may complain of burning and pain.[10] The clinical features at this stage closely resemble sunburn with erythema and skin oedema as dominant signs.[10] Skin changes on the arms may extend proximally with a characteristic line of demarcation at the distal margin of clothing to form the 'glove' or 'gauntlet' of pellagra.[11]

Feet and legs up to the distal edge of trousers or skirt are also commonly affected sites. The sharply demarcated hyperpigmentary changes of pellagra on the lowerlegs and feet have been likened to 'boots'.[11] A broad hyperpigmented band or collar like appearance has been reported for pellagra on the neck. This is commonly referred to as 'Casal's necklace' in recognition of the first doctor to describe this characteristic sign of pellagra.[10,12]

The pathogenetic mechanism of photosensitivity in pellagra remains unclear. Four theories have been proposed: (i) cutaneous deficiency in urocanic acid;[12,13] (ii) accumulation of kynurenic acid;[10,12] (iii) deficiency of NAD /NADP;[14] and (iv) altered porphyrin metabolism 57.[15]

About one-third of patients with pellagra have involvement of the lip, tongue and oral mucous membrane.[2]

Bilateral symmetrical skin lesions over the bony prominences of the body sites include the knees, ankles, elbows and spinous processes.[10,16] These lesions are hyperkeratotic and hyperpigmented. In contrast to the exposed-site photosensitivity skin changes, these skin changes are slow in onset.[16]

Patients with pellagra may develop sebaceous gland hyperplasia and prominent seborrhoea.[17]

Clinical diagnosis is confirmed by rapid response to oralnicotinamide when up to 500 mg daily in divided doses is given. Nicotinamide is the preferred supplement, as niacin causes flushing and headaches.[2,6,18]

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/ their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed

Conflict of Interest

nil

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