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Case Report

Pellagra: An uncommon disease in the modern era - A case report

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Abstract Pellagra is a state of niacin deficiency, a vitamin required for various metabolic processes, cell signaling and DNA repair. It is characterized by diarrhea, dermatitis and dementia that can ultimately lead to death. 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 with a view to highlight that a strong clinical suspicion and careful socio-nutritional history is important to avoid missing such cases. Absence of facial and neck involvement also adds rarity to our case.

Key words
Dermatitis, diarrhea, niacin deficiency, pellagra.

Introduction

Pellagra is a nutritional disorder caused by the deficiency of niacin and is characterized classically with three 3 Ds, namely dermatitis, diarrhea and dementia.[1] These classical manifestations are rarely seen nowadays, especially due to dietary deficiency alone.[2] We report a case of pellagra with the classical dermatological manifestations on the hands and feet but no involvement of the facial and neck region. Absence of facial and neck involvement has been rarely reported thus obliging us to present this case.

Case Report

A 32-year-old female patient presented to us with darkening of the hands and feet for 6 months and darkening of the oral cavity for 2 months. The patient initially developed darkening of the right foot, which gradually increased to involve the both feet up to the ankles and both hands including the inner wrists. The lesions were associated with itching. There was history of fluid-filled and pus-filled lesions over the feet around 2 months ago and which was present till 15 days prior to presentation. The skin lesions developed burning sensation on exposure to sunlight. The patient developed oral ulcers and darkening of the oral cavity around 1 month back. She was otherwise healthy and had regular diet consisting of vegetables, pulses and wheat. The bowel habits were normal. She denied any change in her behavior. There was no history of similar disease in the family. There was no history of intake of any medication or addiction. On examination, the patient had pallor. Bilaterally symmetrical, sun burnt like blackish hyperpigmented plaques were present over the dorsal aspect of hands and feet more prominent over the knuckles, and extending over

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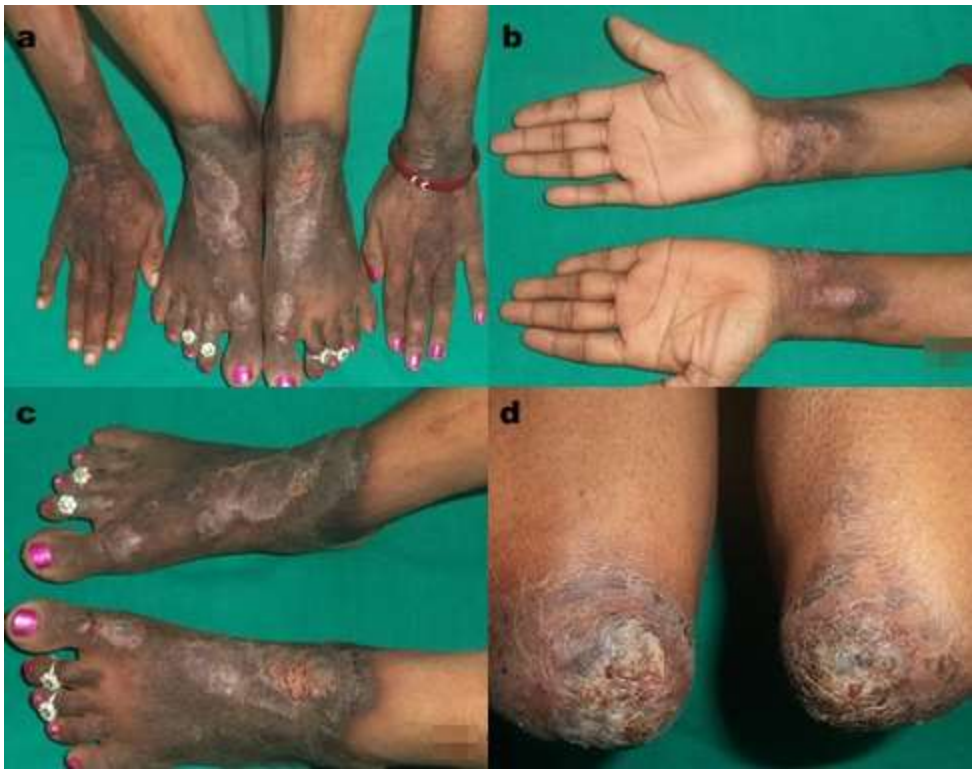

Figure 1 Well-marginated, blackish hyperpigmented plaques present bilaterally over the dorsal aspects of both hands and feet with a burnt out appearance (a). Involvement of hands and feet with a clear-cut demarcation resembling gauntlet and boots respectively (b, c). Hyperkeratotic, hyperpigmented and crusted plaques over elbows (d).

Figure 2 Oral mucosa showing pigmentation, superficial erosions and whitish patch like deposits (a, b).

to the lower part of the leg and medial 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**). There were similar plaques over both the knees, elbows and upper medial buttocks (**Figure 1**). Some areas over the plaques showed erosions and crusting. At some places, the plaques had atrophic changes. The oral cavity showed pigmentation of the buccal mucosa and 2-3 ulcers with a red halo on the lateral aspect of the tongue with whitish deposits over them (**Figure 2**). Rest of the cutaneous, systemic and mental state examination was within normal range.

The patient was found to have hemoglobin of 6.3% with a microcytic and hypochromic red blood cell indices. Rest of the blood biochemistry was within normal range. HIV ELISA was nonreactive. Histopathology showed hyperkeratosis, parakeratosis, elongation of rete ridges and mild perivascular mononuclear infiltrate. Serum niacin estimation could not be done due to financial constraints. 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 nutritional supplementation. The patient responded dramatically and the lesions disappeared in 4 weeks.

Discussion

Pellagra was first described in 1735 by Don Gaspar Casal by the name 'mal de la rosa' due to the reddish and glossy rash that he noted amongst the poor of Spain. Pellagra is caused by the cellular deficiency of niacin (vitamin B3) or its precursor tryptophan. This vitamin is important for the metabolism of carbohydrates, fats, proteins and alcohol, detoxifies various drugs and plays a role in cell signaling and DNA repair.[3] The recommended daily allowance of niacin is 15-20 mg. 60 mg of tryptophan is required to make 1mg of niacin apart from vitamin B2 and B6.[4] Primary pellagra is said to occur due to dietary deficiency of niacin or tryptophan. Secondary pellagra is due to processes interfering with metabolism or absorption of niacin or tryptophan. This occurs in chronic alcoholism, anorexia nervosa, ileocolitis, prolonged diarrhea, Hartnup's disease and carcinoid syndrome. Drugs like isoniazid, phenobarbitone, azathioprine, 5-fluorouracil and 6-mercaptopurine can lead to pellagra.[1,5] Poverty, alcoholism and consumption of corn are the most commonly observed risk factors associated with pellagra.

Clinical features of pellagra begin with gastrointestinal symptoms followed by dermatitis and then dementia, ultimately leading to death. In Italian, pellagra means rough skin. Early cutaneous lesions mimic sunburn and presents as erythema of sun-exposed areas with vesiculation that leave areas of denuded epithelium that heals with dusky brown pigmentation. Sun exposure leads to exacerbation of the lesions.[6] Later these lesions turn hyperkeratotic, dry, scaly, parchment like and pigmented, with a yellow brown hue. Healing occurs centrifugally with desquamation of the center of the lesions and persistence of erythema at the margins. Though pellagra can present on any site, the dorsum of the hands, feet, face, neck and arms are the usual sites of involvement.[7] The most common site is the dorsum of hands with the lesions extending up the arm in the form of gauntlet. Lesions on the face follow the distribution of the fifth nerve. A well-marginated eruption of the front of neck has been termed as Casal's necklace. Recurrent erythema occurs on photoexposed skin. The oral mucosa is involved in a third of all cases and shows inflammation, edema, furring, erosions and ulcers. Gastrointestinal features include diarrhea, nausea, vomiting, decreased appetite, gastritis and achlorrhydria.[6] Neuropsychiatric features include headache, irritability, poor concentration, apathy, depressed mood, psychomotor unrest, ataxia, photophobia and spastic paresis.[8] Death may occur due to the malnourished state of the patient.

Diagnosis is based on a suggestive clinical setting. Serum niacin levels may be used to estimate deficiency or urinary levels of pyridone or N-methyl nicotinamide may be used to confirm the disease.[9] Histopathology is nonspecific and includes hyperkeratosis, parakeratosis, pallor of upper epidermis, capillary dilatation, dermal edema and perivascular lymphohistiocytic infiltrate. These

can help differentiate pellagra from other differentials like lupus erythematosus, porphyria cutanea tarda, polymorphic light eruption, drug-induced eruptions, other nutritional deficiencies like zinc and eczematous dermatitis.

Since patients of pellagra are often deficient in other nutrients as well, a high protein diet with B-complex vitamins is needed.[10] Niacinamide is preferred over nicotinic acid for treatment as high doses of nicotinic acid may precipitate flushing, itching and burning. WHO recommends 300 mg of oral nicotinamide or 100 mg parenterally daily in divided doses. Neurologic symptoms disappear in 2-3 days but cutaneous lesions take around a month to resolve. Maintenance with 50-100 mg per day is recommended.[11] Photoprotection is advisable. Dietary and lifestyle modifications like abstinence from alcohol, change of drugs, and avoidance of maize may be needed.

Conclusion

Pellagra due to dietary causes is rare nowadays. We report this case of pellagra with classical cutaneous features to highlight the fact that pellagra is down but not yet out. This case also highlights the fact that pellagra can occur without affecting face and neck. A careful social and dietary history, proper clinical examination and a strong clinical suspicion is needed to prevent missing the diagnosis of this disease.

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