

Answer to “What is Your Diagnosis?” on p.79

Diagnosis: Pellagra

Pellagra is a nutritional disease due to a deficiency of niacin, or its precursor amino acid, tryptophan and characterized by photosensitivity, gastrointestinal symptoms and neuropsychiatric disturbances leading to even death, deserving the mnemonic of 4 “D”s: dermatitis, diarrhea, dementia, and death (1,2). This patient had a typical cutaneous presentation associated with gastrointestinal and neurologic findings which were consistent with pellagra.

Skin changes are pathognomonic

The most characteristic cutaneous finding is a photosensitive rash affecting symmetrically the dorsal surfaces of the hands, forearms, face, neck, upper chest and dorsal surfaces of the feet (3). In the acute stage erythema, edema and vesicobullae with burning and pain develops after sun exposure, resembling a sunburn reaction. Recurrent events lead to a chronic, scaly rash with a thickened, violaceous or hyperpigmented skin. The dorsal hands and forearms are the most commonly involved areas forming the “gloves” of pellagra. There is striking clear demarcation from normal skin. Facial lesions show a “butterfly” eruption similar in appearance to that of lupus erythematosus. Lips, tongue and oral mucous membranes are frequently involved, manifesting cheilitis, angular stomatitis, and glossitis (1-3). Neuropsychiatric findings include emotional disturbances such as depression, anxiety, or irritability, headaches, insomnia, mental confusion, ataxia, and peripheral neuritis. In later stages of the disease hallucinations, seizures, paralysis of the extremities, psychosis, dementia, or even coma may develop (1-4).

The most frequent gastrointestinal symptoms are loss of appetite, abdominal pain, vomiting, and later watery diarrhea (1-3).

In developed countries, pellagra occurs usually in alcoholics

Pellagra is classically associated with dietary deficiency, subsisting on corn based diets on impoverished populations. Other vitamin deficiencies or malnutrition, which interfere with the conversion of tryptophan to niacin, often exists. In the developed world, it is a rare condition mostly seen in alcoholics, food faddists, or recluses with psychiatric disorders (1-3). In alcoholics pellagra develops due to poor diet and malabsorption. Moreover, hepatocytes are also inefficient in utilizing niacin (1,5).

Gastrointestinal malabsorption diseases, carcinoid tumour, Hartnup disease, prolonged intravenous supplementation, hepatic cirrhosis, HIV infection and anorexia nervosa may also cause pellagra. Similarly, drugs which interfere with the tryptophan-niacin pathway including isoniazid, pyrazinamide, ethionamide, 6-mercaptopurine, hydantoins, phenobarbital, azathioprine, and chloramphenicol may induce pellagra (1-3).

Pellagra is primarily a clinical diagnosis. The most reliable test is urinary excretion of niacin metabolites

Cutaneous biopsy findings are not specific. Although their correlation with body stores are not so strong, low serum levels of niacin, tryptophan, nicotinamide adenine dinucleotide, or a reduced urinary excretion of metabolites may indicate niacin deficiency. In our patient, low serum levels of niacin (6.5 ug/L, N: 8-52) supported the clinical diagnosis of pellagra.

The differential diagnosis of pellagra includes phototoxic and photoallergic skin diseases, porphyrias and pseudoporphyrias, lupus erythematosus, and polymorphous light eruption.

Rapid clinical response to niacin treatment provides an important additional clue to diagnosis

Specific treatment is usually with oral niacin 100-400 mg/day, until resolution of symptoms and healing of all the skin lesions. In severe cases niacin can be initially given by parenteral route. The recommended daily maintenance dose of niacin is 5-20 mg. We treated our patient with 300 mg/day niacin along with B complex vitamins and a high quality protein diet. Regular application of sunscreens on exposed areas was recommended. After 6 weeks, the eruption was completely healed (Figure 2), and gastrointestinal and neuropsychiatric symptoms were dramatically regressed.



In conclusion, pellagra should be suspected when symmetric photosensitive skin eruptions develop in patients with chronic alcoholism.

References

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