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Porphyria Cutanea Tarda Presenting as Erythema-multiforme Like Lesions

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ABSTRACT

Porphyria cutaneatarda (PCT), is the most common type of porphyria.It is characterized by defective uroporphyrinogen III decarboxylase enzyme.It presents with erosion, bulla with milia formation and sometimes with hypertrichosis and abnormal pigmentation mostly on the photo-exposed sites. A urine fluorescence of coral red color helps in the diagnosis. Here, we present a rare case of porphyria cutanea tarda in a 15 years old male who presented with multiple targetoid plaques.

Keywords: Erythema-multiforme; porphyria cutanea tarda; targetoid.

INTRODUCTION

Porphyria cutanea tarda(PCT), is the most common type of porphyria, a group of metabolic diseases caused by defects in the heme biosynthetic pathway which results in an excess of porphyrins (red-brown pigments) and their precursors. PCT is characterized by presence of erosion, bulla with milia formation mostly on photo-exposed sites. Here, we present a case of PCT presenting as targetoid plaques over the trunk.

CASE REPORT

A 15 years old male from Makawanpur district presented with complains of multiple blistering eruption on the face for the last 9 months. The multiple vesicular eruptions were found to start abruptly and healed with post-inflammatory hyperpigmentation. There was slight burning and stinging sensation during the onset of the eruption.

Gradually, the lesions also started to appear on his chest and back region which presented with plaques rather than blisters.

For the past 9 months, patient had been treated couple of times as a case of otitis externa for eruption on the pinna. The skin eruption improved with topical combination of steroid and antibiotic. Yet, new eruptions occurred continuously. No systemic symptoms were noted. A detailed history could not elicit any use of medications prior to the eruption. No family history of similar eruption. No history of joint pains, oral ulcers or muscle weakness.

The patient was a student,who loved swimming during weekend. On examination, few tense vesicles along with multiple hyperpigmented macules were noted over the nose and bilateral mandible and pinna sparing the forehead (Figure 1). Multiple raised polymorphous targetoid plaques were present over the upper trunk (Figure 2a and 2b). There were few areas of hypertrichosis noted on bilateral tragus. No milia formation or scarring was noted. Nails and mucosa were both normal.

Complete blood count revealed total count of 11,800 with predominance of neutrophils (88%). Hemoglobin was 12.7 mg/dl and raised ESR of 26 mm/1st hour. Random blood sugar was within the normal limit. Urine turned slightly brown in color while on standing. Wood's lamp examination of the urine revealed coral red fluorescence (Figure 3). Urine porphobilinogen was positive. Liver function test showed only a slight rise in alkaline phosphatase. Serology for Hepatitis C was non-reactive. Coagulation profile was all within normal limits. Biopsy from the skin lesion showed subepidermal cleft.

Ultrasonography of abdomen and pelvis showed no abnormalities.

So, considering the presence of skin lesion with urine porphobilinogen along with coral red fluorescence of

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urine on standing without any systemic symptoms, we made a diagnosis of porphyria cutaneatarda.

Both patient and parents were on different measures of sun protection and its importance. Patient was also advised on regular follow-up.

DISCUSSION

The diagnosis of PCT is straightforward because of the characteristic clinical findings. Blisters and erosions develop acutely on sun-exposed skin, sometimes accompanied by hypertrichosis, abnormal pigmentation, and milia formation.[1-4] PCT is the most common porphyria and is characterized by defective uroporphyrinogen III decarboxylase enzyme.[1] Examination of urine is diagnostically crucial, which is red to brown in natural light and pink to red in fluorescent light. Diagnosis is confirmed by measurement of porphyrin in 24-hour urine.[5]

The mainstay of treatment is low dose twice weekly chloroquine. Venesection is reserved for the severe cases of iron-overload or hepatitis C-related liver disease.[2] Avoidance of sunlight exposure and wearing clothing designed to provide protection for conditions with chronic photosensitivity are essential.[6] The distribution of lesion in our case might be due to swimming habits of patients.

There is only a single case report published on PCT masquerading as epidermolysis bullosa acquisita.[7] But, there are no case reports to our knowledge of PCT that presented as erythema-multiforme like plaques.

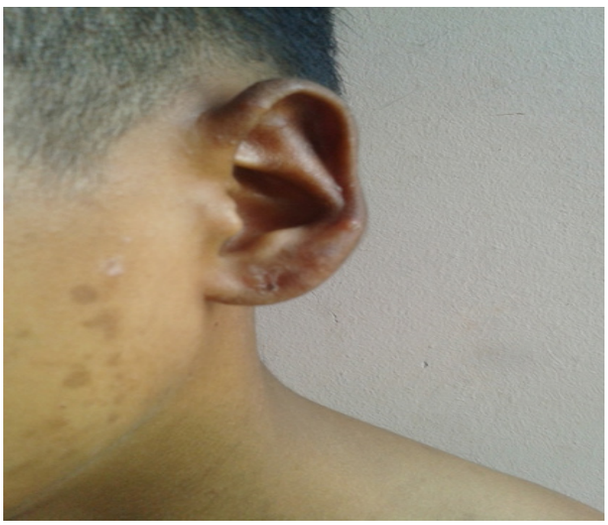

Figure 1. Few tense blister on left pinna with post-inflammatory hyperpigmented macules over the mandible.

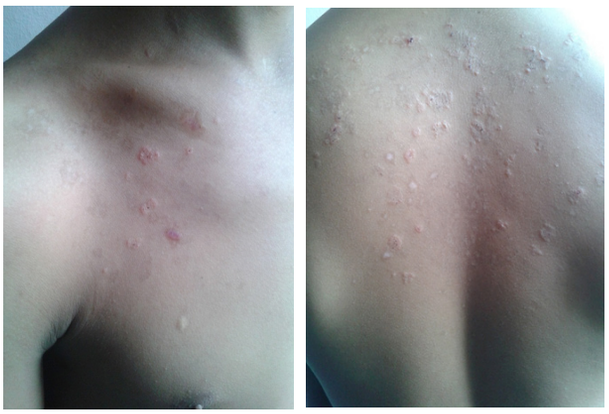

Figure 2a and 2b. Multiple targetoid plaques noted over the upper trunk.

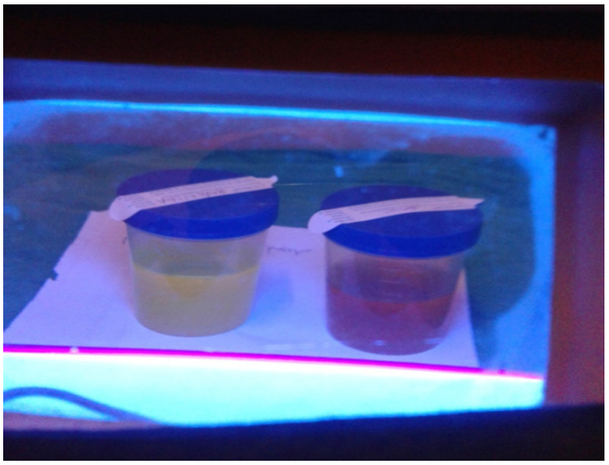

Figure 3. Wood's lamp examination of urine (Note: the urine on right with coral red fluorescence is from the patient while the other is from a normal subject)

CONCLUSIONS

Porphyria cutanea tarda is the most common type of porphyrias It is usually characterized by blisters and fragility of skin in light-exposed areas but rare presentations are not uncommon. Clinicians should be aware these presentations and any eruptions involving the sun-exposed regions should prompt proper investigations for porphyria.

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