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Hurricane Katrina evacuee develops a persistent rash

Seven months after Hurricane Katrina, a 52-year-old African American woman who was evacuated from New Orleans came to our office with a hypopigmented rash on both her upper thighs and arms. Immediately following the 2005 hurricane, she was forced to wade through the polluted waters of New Orleans for many hours before being rescued by boat. Four days passed before she had access to a shower. It was after this shower that she first noticed a single erythematous spot the size of a silver dollar on her left thigh, that several weeks later faded to hypopigmented macules and plaques. Over time, the rash spread to both thighs and arms (**FIGURES 1 AND 2**). She did not have any itching, pain, bleeding, fever, chills, weight changes, or gastrointestinal symptoms after the rash appeared.

The patient reported that she was married and had worked as a chef for 20 years. She smoked cigarettes, drank alcohol occasionally, and was obese. She had synovitis of her left ankle, which led to surgery. She had no known drug allergies and was taking ibuprofen. Her mother, age 79, had glucose intolerance; her father, age 82, had a renal cell carcinoma removed.

Due to a confluence of situational, economic, and medical problems, she did not seek care for this rash until April

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She waded through the polluted waters of New Orleans for many hours before she was rescued by boat

FIGURE 1

Rash on patient's legs

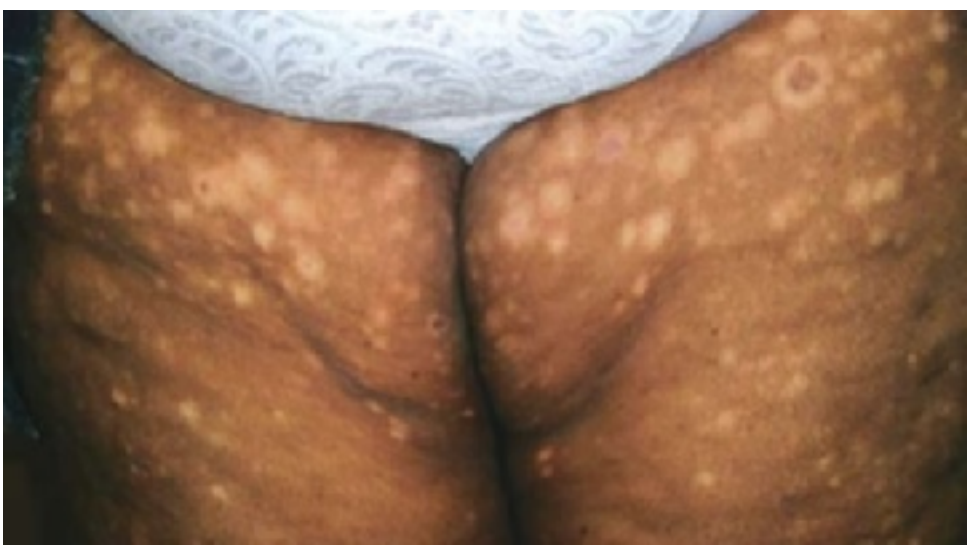

PHOTO BY RICHARD P. USATINE, MD.

Front view of the patient's lower extremities, exhibiting hypopigmented macules and plaques to the upper thighs.

2006. At that time, a punch biopsy revealed the findings in **FIGURE 3**. Physical exam revealed her skin to have symmetrically distributed hypopigmented macules and plaques to all 4 extremities. She had no lymphadenopathy.

- **What is the diagnosis?**
- **Were Katrina's flood waters to blame?**

FIGURE 2

Rash on right arm

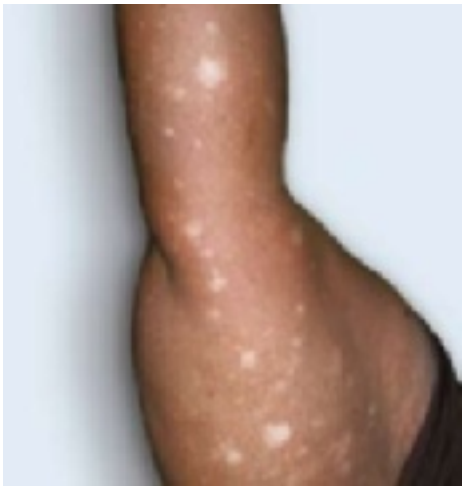

PHOTO BY RICHARD P. USATINE, MD.

Distribution of hypopigmented macules and plaques is symmetrical when compared with macules and plaques on the left arm.

FIGURE 3

Hematoxylin/eosin stain

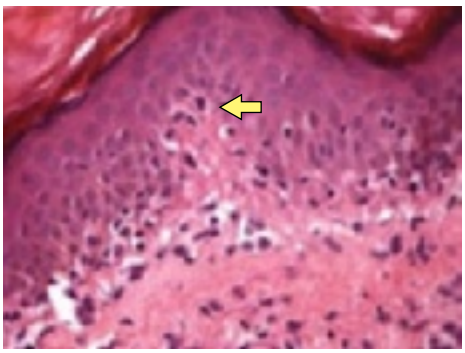

IMAGE PROVIDED BY JENNIFER BLANKENSHIP, MD.

Hematoxylin and eosin stain (at 400x power) of punch biopsy showing "cerebriform" lymphocytes at the dermal-epidermal junction, migrating into the epidermal layer.

TABLE

Staging for cutaneous T-cell lymphoma based on the Tumor, Node, Metastasis (TNM) system

STAGE	TNM STAGING	RECOMMENDED TREATMENTS
IA IB	**T1, N0, M0:** Patch or plaque <10% body surface area **T2, N0, M0:** Patch or plaque >10% body surface area	Topical high-potency steroids, PUVA, topical nitrogen mustard, carmustine, or bexarotene
IIA	**T1–2, N1, M0:** Patch or plaque with palpable but pathologically normal lymph node	Same as Stage I; if refractory, use total skin electron beam therapy
IIB	**T3, N0–1, M0:** Tumor/nodule	Chemotherapy or photophoresis, refer to medical oncologist, radiation oncologist, and dermatologist
IIIA	**T4, N0, M0:** Generalized erythroderma	
IIIB	**T4, N1, M0:** Erythroderma and palpable but pathologically normal lymph node	
IVA	**T1–4, N2–3, M0:** Pathological lymph node	
IVB	**T1–4, N0–3, M1:** Visceral (M1) or blood involvement	

T: 0–4 = indicates size or direct extent of the primary tumor

N: 0 = tumor cells absent from regional lymph nodes; 1 = tumor cells spread to closest or small number of regional lymph nodes; 3 = tumor cells spread to most distant or numerous regional lymph nodes

M: 0 = no distant metastasis; 1 = metastasis to distant organs (beyond regional lymph nodes)

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Mycosis fungoides may be caused by environmental hazards, but we saw no other cases in Katrina evacuees

Dx: T-cell lymphoma—timing is coincidental

The biopsy revealed cutaneous T-cell lymphoma (CTCL)—specifically a type known as mycosis fungoides, named after the mushroom-like skin tumors seen in severe cases. CTCL is a malignant lymphoma of helper T-cells that have an affinity for the skin.

Normal life expectancy likely if diagnosed early

In about 10% of cases, the T-cells spread via the lymphatic system to metastasize to the liver, lung, and bone marrow, but more often remain confined to the skin and lymph nodes, and most patients diagnosed early have a normal life expectancy.[1]

The disease is rare, with about 1000 new cases per year in the US,[1] and is more common in African Americans,[2] although the photos in dermatological atlases overwhelmingly show Caucasians. CTCL presents in numerous ways, but patients usually have a long course of persistent rash that is often pruritic and usually erythematous or hyperpigmented.

No one knows what causes CTCL, but current theories point to exposure to environmental hazards such as Agent Orange.[2] And while the number of environmental hazards in the floodwaters of Katrina were vast, we have no scientific evidence that an environmental exposure was to blame. In fact, while we cared for a number of Katrina evacuees—many of whom had skin infections—this was the only case of mycosis fungoides.

It's likely that the start of the visible mycosis fungoides lesions after the flood was purely coincidental.

Diagnosis hinges on palpation and biopsy

To diagnose CTCL, you need to palpate for enlarged lymph nodes and perform a full-thickness punch biopsy of the lesion. If the biopsy is negative and the rash persists, take another biopsy—results can be falsely negative at the beginning stages of the disease.

There are no guidelines on which studies should be used for staging CTCL (**TABLE**), but some sources recommend that if lymph node involvement is suspected on physical exam, lymph node biopsies should be done in addition to a chest radiograph.[3] In more advanced stages, consider a computed tomography (CT) scan of the abdomen and pelvis. A recent study concluded that CT and positron-emission tomography (PET) scans used together were more sensitive in staging, but this may not be cost-effective.[4]

Late-stage mycosis fungoides is usually associated with immunocompromise. Therefore, HIV testing should be performed in all patients with CTCL. Other important laboratory studies include complete blood count—with differential—as well as peripheral smear looking for Sézary cells, lactic dehydrogenase (LDH), liver function tests, and uric acid.

A disease that's easy to mistake for vitiligo

The hypopigmented spots of CTCL look so much like vitiligo, it is frightening to think how easy it would be to miss the diagnosis. Complicating matters: All vitiligo does not need a biopsy to confirm the clinical impression.

So what made this case suspicious enough to warrant a biopsy? First, the hypopigmented spots on our patient were not a typical distribution for vitiligo, which has a predilection for the hands and face. Second, our patient had a hypopigmented patch that had a dark, slightly raised plaque within it, which also would not be typical for vitiligo. (See the patient's left upper thigh, just below the inguinal area, in **FIGURE 1**.) Finally, our patient had a rapid onset of multiple discrete macules that did not coalesce into larger hypopigmented areas; in vitiligo you would expect otherwise.

The differential diagnosis also includes idiopathic guttate hypomelanosis, a benign condition that can cause multiple small hypopigmented macules. The size of immunoglobulin H macules, however, is tiny compared with what you'll see with CTCL. The absence of scales makes eczema, psoriasis, or tinea corporis very unlikely.

Treatment starts with topical steroids

This is a rare disease that lacks the data needed to support an evidence-based approach to treatment. Topical steroids are recommended for stage I when the disease is local to the skin (see **TABLE** for recommended treatments of other stages). Ultraviolet light (PUVA and UVB) is also used; a recent study suggests the PUVA is a good treatment alternative for stages IA and IB (SOR: **C**).[1]

Our patient's course

Initially, we prescribed a high-potency generic steroid (clobetasol 0.05% cream) for this patient, to be used twice daily to the affected areas.

The patient reported no improvement with this approach, while she awaited her appointments with dermatology and oncology specialists.

Her blood tests were essentially normal, including a negative HIV test.

She is currently receiving narrowband UVB treatment twice weekly. ■

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When the disease is local to the skin, topical steroids and UV light are helpful

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