



# PITYRIASIS LICHENOIDES

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Pityriasis lichenoides is an uncommon disease of the skin that can present in three different forms: pityriasis lichenoides et varioliformis acuta (PLEVA), pityriasis lichenoides chronica (PLC), and febrile ulceronecrotic Mucha-Habermann disease (FUMHD). These three forms represent a spectrum of disease presentation. Any of the three types can occur alone, but it is possible for one form to evolve from another. Pityriasis lichenoides affects roughly 1 in 2000 people per year. It is seen slightly more often in males and in late childhood to early adulthood. The disease can occur in people of all ages and races.

PLEVA starts as an acute eruption of bright red, flat to slightly raised, 2-10 mm oval spots. The lesions then evolve, developing small blisters and pustules, and eventually ulcerate and crust over. The lesions can appear singly or in groups, and can coalesce into large areas of rash. Successive crops appear over weeks, so all stages of lesions can be present at one time. The rash is typically seen on the trunk, thighs, upper arms, and flexural areas. In 10% of cases, the face, palms, soles and genitals are involved. There can be mild itching or burning, but usually no other symptoms are present in PLEVA. The rash can come and go, lasting for 1.5 to 18 months without treatment. Scarring and skin discoloration can result.

PLC is more common and has a more mild presentation compared with PLEVA. There is a subtle appearance of flat, red to brown oval spots over the trunk, thighs, and upper arms. Fine scale peels at the edges and is adherent to the center of each lesion. PLC can also relapse and remit over years. It typically does not scar.

FUMHD is very rare, and unlike PLEVA and PLC, it is considered a dermatologic emergency. It typically presents abruptly and dramatically with a wide-spread eruption of red to black ulcerated, necrotic plaques. There are systemic symptoms as well, which can include high fever, abdominal pain, diarrhea, joint pain, breathing difficulties, and changes in mental status. Patients with FUMHD require hospitalization. There is a 25% mortality rate in patients with FUMHD.

The cause of pityriasis lichenoides is not well understood. There is a proliferation of immune cells, called T-cells, in the skin. It is believed that genetically susceptible individuals mount an inappropriate immune response to a foreign agent, such as a virus or medication, which causes inflammation in the skin. Pityriasis lichenoides has been seen in association with many illnesses, including streptococcus, HIV, chickenpox, Epstein-Barr virus, cytomegalovirus, and hepatitis C. Some medications, such as **antihistamines**, estrogen-progesterone therapy, and the measles vaccine, have been implicated in pityriasis lichenoides. Most often, no cause for the disease is identified.

To make the diagnosis of pityriasis lichenoides, a dermatologist will **biopsy** a lesion to look for the characteristic pattern of inflammation in the skin. Blood tests are sometimes done to rule out other causes of rash or identify a triggering infection. PLEVA and PLC are not associated with any abnormal blood tests. However in FUMHD, there can be increases in the white blood cell count and markers of inflammation. PLEVA can look very similar to some types of cutaneous lymphoma, so it is very important to exclude malignant or premalignant conditions during the diagnosis.

PLEVA and PLC will both eventually resolve without treatment, but it can take months or years. A prolonged course of antibiotics, such as erythromycin or **tetracycline**, is often given to decrease the duration of the disease. **Oral steroids** are sometimes used with the antibiotics to speed clearance of lesions. **Light therapy** is also helpful treating PLEVA and PLC. FUMHD is treated in hospital with medications including IV gamma globulin, **dapsone**, **cyclosporine**, and **methotrexate**.

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