OASIS DERMATOLOGY GROUP

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Pityriasis Lichenoides

Pityriasis lichenoides is a rare, chronic, and inflammatory skin disorder characterized by distinctive eruptions that manifest in three distinct forms: pityriasis lichenoides et varioliformis acuta (PLEVA), pityriasis lichenoides chronica (PLC), and febrile ulceronecrotic Mucha-Habermann disease (FUMHD). These variants represent a spectrum of disease severity and clinical progression, with PLEVA and PLC being the most commonly observed forms. While the overall incidence of pityriasis lichenoides is approximately 1 in 2,000 people per year, the condition primarily affects males and typically presents in late childhood to early adulthood. However, it can occur in individuals of all ages and ethnic backgrounds.

Clinical Presentation and Subtypes

Pityriasis Lichenoides Et Varioliformis Acuta (PLEVA)

PLEVA is the acute form of pityriasis lichenoides, presenting as bright red, flat to slightly raised, 2-10 mm oval lesions. These spots often evolve into blisters and pustules, which can later ulcerate and form a crust. PLEVA lesions may appear individually or in groups and can merge to form large, confluent areas. Successive crops of lesions can emerge over a period of weeks, leading to the presence of lesions in various stages of development at once. The trunk, thighs, upper arms, and flexural areas are the most commonly affected sites, with 10% of cases involving the face, palms, soles, and genital area. Symptoms may include mild itching or burning, though many patients report no additional symptoms. The lesions can persist for 1.5 to 18 months, typically resolving spontaneously, but scarring and hyperpigmentation may occur as a result of prolonged inflammation.

Pityriasis Lichenoides Chronica (PLC)

PLC is a more chronic and milder form of pityriasis lichenoides, characterized by the appearance of flat, red to brown oval spots. These lesions are usually smaller than those seen in PLEVA and may have a fine scale that is adherent at the center of the lesion while peeling at the edges. The lesions typically appear on the trunk, thighs, and upper arms, and the condition often has a relapsing-remitting course that can last for years. Unlike PLEVA, PLC typically does not result in scarring or significant pigmentation changes. Most patients experience spontaneous resolution of the lesions, but relapses are common.

Febrile Ulceronecrotic Mucha-Habermann Disease (FUMHD)

FUMHD is the rarest and most severe form of pityriasis lichenoides, and it is considered a dermatologic emergency. FUMHD presents suddenly and dramatically with widespread red to black ulcerated plaques, which may become necrotic. In addition to the cutaneous manifestations, patients often exhibit systemic symptoms, including fever, abdominal pain, diarrhea, joint pain, and respiratory distress, with some individuals developing neurological symptoms such as altered mental status. Due to the severity of systemic involvement, hospitalization is required, and FUMHD carries a 25% mortality rate in affected individuals.

Etiology and Pathogenesis

The exact cause of pityriasis lichenoides remains poorly understood, but it is believed to involve an immune-mediated process. It is hypothesized that genetically predisposed individuals may mount an inappropriate immune response to an exogenous trigger, such as a viral infection, a medication, or environmental factors. Streptococcus, HIV, chickenpox, Epstein-Barr virus, cytomegalovirus, and hepatitis C have all been implicated as potential triggers for pityriasis lichenoides. Certain medications, such as antihistamines, estrogen-progesterone therapy, and the measles vaccine, have also been associated with disease onset in some cases.

Although a definitive causative agent is not always identified, there is a notable association between pityriasis lichenoides and infectious diseases, particularly streptococcal infections. This suggests that immune dysregulation, often triggered by an infection, could lead to the characteristic skin eruptions seen in pityriasis lichenoides.

Diagnosis

The diagnosis of pityriasis lichenoides is primarily clinical, supported by a skin biopsy to confirm the characteristic histopathological features. Blood tests may be conducted to exclude other causes of rash or to identify potential underlying infections or systemic involvement. However, PLEVA and PLC are not associated with abnormal blood counts or serological markers, while FUMHD may show elevated white blood cell counts and inflammatory markers, reflecting the systemic nature of the disease.

In clinical practice, it is important to differentiate PLEVA from conditions such as cutaneous T-cell lymphoma (CTCL), as the former can mimic the appearance of mycosis fungoides, a type of lymphoma. Exclusion of malignancy through careful histological analysis is essential in ensuring an accurate diagnosis.

Treatment Options

Treatment for PLEVA and PLC typically involves antibiotics and topical therapies to reduce inflammation and expedite lesion resolution.

- ***Antibiotics***: Erythromycin or tetracycline are commonly prescribed for PLEVA and PLC, as these agents help to control inflammation and shorten the disease duration. A prolonged course of oral antibiotics may be required for effective treatment, often lasting several weeks.
- ***Oral Steroids****:* In some cases, oral corticosteroids may be used adjunctively to accelerate the resolution of lesions. However, long-term steroid use is generally avoided due to the risk of systemic side effects.
- ***Phototherapy****:* Ultraviolet B (UVB) light therapy, particularly narrowband UVB, has been demonstrated to be effective in treating both PLEVA and PLC. This treatment works by suppressing T-cell activity and modulating the skin's immune response.

FUMHD, due to its severe nature, requires hospitalization and intensive therapy. The following medications are typically used:

- ***IV Immunoglobulin (IVIG)***: IVIG is administered to modulate the immune system and reduce inflammation.
- ***Dapsone***: Dapsone, an antibiotic with anti-inflammatory properties, is commonly used in severe cases to manage skin lesions and systemic symptoms.
- ***Cyclosporine and Methotrexate***: Immunosuppressive medications such as cyclosporine and methotrexate may be employed to control the immune response, particularly in cases refractory to other treatments.
- ***Supportive Care****:* Patients with FUMHD require supportive care to manage systemic symptoms, including fever, pain, and respiratory distress.

Prognosis

While PLEVA and PLC typically resolve spontaneously without long-term complications, FUMHD carries a significant risk of morbidity and mortality. Early diagnosis and aggressive treatment are critical to improving outcomes for patients with this severe form of the disease. Scarring and pigmentation changes may occur in patients with PLEVA or PLC, but these are usually not permanent.

Conclusion

Pityriasis lichenoides encompasses a spectrum of skin disorders ranging from mild to life-threatening conditions. Although the pathogenesis is not fully understood, immune dysregulation triggered by infections or medications is believed to play a central role. Treatment strategies include the use of antibiotics, topical corticosteroids, and phototherapy for milder forms, while more severe cases require immunosuppressive and immunomodulatory therapies. Prompt diagnosis and management are crucial, particularly in cases of FUMHD, to prevent serious complications and improve patient outcomes.

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