Crusted Norwegian Scabies in an Adult With Langerhans Cell Histiocytosis

Mishaps Leading to Systemic Chemotherapy

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Background: Crusted Norwegian scabies is a rare hyperkeratotic variant of scabies infestation. We report herein a case of crusted scabies in a woman with underlying Langerhans cell histiocytosis (LCH).

Observations: A 49-year-old woman with LCH was hospitalized owing to marked thrombocytopenia. Her hyperkeratotic skin eruption was thought to be secondary to LCH because several years earlier, she had positive biopsy findings and had been diagnosed as having LCH. After a 1-month hospital stay, her laboratory values improved despite worsening of her skin lesions.

Conclusion: Analysis of skin scrapings confirmed the presence of scabies, and resolution was achieved with a 1-dose ivermectin treatment.

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Crusted Norwegian scabies is a rare, massive form of infestation by Sarcoptes scabiei var hominis characterized by markedly hyperkeratotic skin. We report herein a case of crusted scabies in a 49-year-old woman with underlying Langerhans cell histiocytosis (LCH). To our knowledge, this is the first case of crusted scabies reported in an adult patient with LCH.

REPORT OF A CASE

A 49-year-old woman with LCH and diabetes mellitus was hospitalized owing to marked thrombocytopenia. The patient was diagnosed with LCH 3 years earlier by skin biopsy specimen evaluation and had been undergoing treatment with prednisone and thalidomide for recurrent symptoms. At admission, the patient was lethargic and bedbound with generalized, crusted, erosive skin lesions, presumptively due to the LCH. Prednisone treatment was initiated to stabilize her platelet count; however, she subsequently developed bacteremia with methicillin-resistant Staphylococcus aureus, Staphylococcus epidermidis, and Pseudomonas aeruginosa.

During her 1-month hospital stay, her laboratory values improved despite worsening of her skin lesions. The patient remained lethargic, with crusted erosive plaques with necrotic areas on the scalp, face, trunk, groin area, and diffusely on her upper and lower extremities. Skin scrapings were taken from several sites, and analysis confirmed the presence of scabies. After 2 applications of 5% permethrin cream without visible response, 12 mg of ivermectin (Mectizan; Merck & Co Inc, Whitehouse Station, NJ) was tried and completely resolved the skin lesions within 1 week. The patient's functional status markedly improved as her skin cleared, and she was discharged 1 week after the ivermectin treatment. No recurrence or adverse effects occurred during the 6 months of follow-up.

COMMENT

Crusted Norwegian scabies is an uncommon, exceptionally contagious variant of scabies that results from failure of the host immune system to control the cutaneous proliferation of the scabies mite, with ensuing hyperinfestation and an accompa-
nying inflammatory and hyperkeratotic reaction. Thick crusts in this variant often obscure burrows, and nails may appear dystrophic. Pruritus is uncommon owing to an impaired host response; this impairment may be caused by neurological factors or by other factors accompanying a debilitated condition. The lack of an impulse to scratch contributes to the massive infestation and mite count.

Predisposing conditions have been associated with diseases that alter T-cell function such as human immunodeficiency virus, human T-cell lymphotrophic virus 1, T-cell lymphoma, and leukemia. Transplant recipients are also predisposed. Systemic lupus erythematosus, rheumatoid arthritis, malnutrition, diabetes mellitus, and various neuropathies have also been associated.

The skin biopsy findings in crusted scabies can be unreliable. The inflammatory response to scabies can appear as lymphohistiocytic infiltration on analysis of skin biopsy specimens. When immunostained, the histiocytic component may have the phenotype of a Langerhans cell (CD1a+/S100+). This can lead to confusion when the diagnosis of LCH is considered in a patient. There have been reports of misdiagnosis of scabies infestations as LCH in pediatric patients, and as a result these patients underwent unnecessary chemotherapeutic treatment (Table). The atypical presentation of crusted sca-
abies can result in delayed diagnosis or misdiagnosis with serious consequences, especially in debilitated or immunodeficient patients prone to life-threatening complications. Septicemia is a common complication and is frequently polymicrobial requiring treatment with broad spectrum systemic antibiotics.

Safe and effective use of a single dose of oral ivermectin (200 µg/kg) to resolve crusted scabies lesions has been reported. However, it is recommended to use the mainstay of topical scabicidal agents as a first line. The use for which ivermectin was approved by the US Food and Drug Administration is for treatment of strongyloidosis and onchocerciasis; caution should be exercised when using it as a scabicidal agent. A single report described an association between ivermectin and an increased risk of death.

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REFERENCES