Case Report

EXFOLIATIVE PSORIATIC ERYTHRODERMA: A CALL FOR HELP AND ATTENTION

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ABSTRACT

Erythroderma is an erythema in more than 90% of the body surface and represents a dermatological emergency with extensive erythematous skin and scaling. First described by Von Hebra in 1868, erythroderma represents a rare cutaneous inflammatory state with associated dysfunction of both the skin barrier and metabolic processes. Most cases result from pre-existing and chronic dermatoses such as eczema and psoriasis. Untreated, the patient may succumb to multisystem organ failure and high-output heart failure secondary to cutaneous volume loss. We present a case of Exfoliative Psoriatic Erythroderma as a patient's first presentation of psoriasis in the eighth decade of life, with no dermatological history, who started with erythrodermic psoriasis, developing a systemic inflammatory response, which was treated with methotrexate with good response. Erythrodermic psoriasis is a severe and rare variant of psoriasis vulgaris, the condition of which can be fatal. The objective of presenting this case is to call attention to this rare entity.

Keywords: Psoriasis; Erythroderma; rare entity; dermatological emergency; treatment; prognosis.

ERITRODERMIA PSORIATIKE EKSFOLIATIVE; NJË THIRRJE PËR NDIHMË DHE VËMENDJE

ABSTRAKT

Erythrodermia është një eritemë në më shumë se 90% të sipërfaqes së trupit dhe përfaqëson një urgjencë dermatologjike. E përshkruar për herë të parë nga Von Hebra në vitin 1868, erythroderma përfaqëson një gjendje të rrallë inflamatore të lëkurës me disfunksion të barrierës kutane dhe proceseve metabolike. Shumica e rasteve rezultojnë nga dermatoza të meparshme dhe kronike si ekzemat dhe psoriaza. Nëse nuk trajtohet, pacienti mund të vdesë nga insuficenca multiorganore dhe insuficenca kardiake. Ne prezantojmë një rast klinik të Erythrodermës Psoriatike Exfoliative si paraqitja e parë e psoriasisit ne një pacient në

dekadën e tetë të jetës, pa histori dermatologjike, qe fillon me psoriasis erythrodermike, duke zhvilluar një përgjigje inflamatore sistemike, me një përgjigje relativisht të mire ndaj terapise sistemik me metotreksat. Erythrodermia Psoriatike është një variant i rëndë dhe i rrallë i psoriasis vulgaris, e cila mund të rezultoje dhe ne fatalitet. Qëllimi i prezantimit të këtij rasti është risjellja ne vëmendje e këtij entiteti të rrallë dermatologjik.

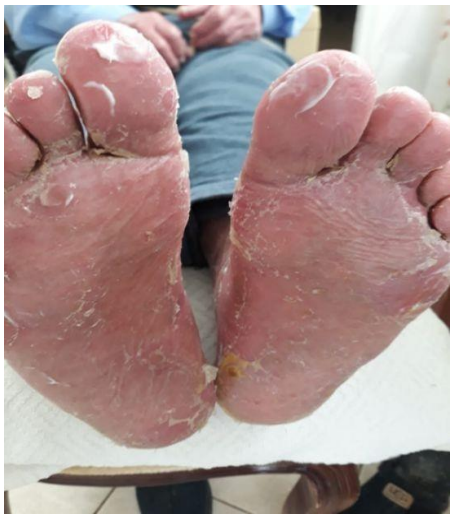
Fjale kyce: Psoriasis; Eritroderma; entitet i rralle; urgjence dermatologjike trajtimi; prognoza.

INTRODUCTION

The annual incidence of erythroderma is estimated to be 1–2 per 100,000 population in Europe, with a male preponderance (6). Erythroderma may be present at birth or may develop acutely or insidiously. Erythroderma is a manifestation of severe skin dysmetabolism, reported in severe forms of a range of diseases that can be broadly categorized into congenital, infective, inflammatory, immunobullous, neoplastic, iatrogenic, and idiopathic causes (7). Many diseases are associated with erythroderma, but most cases result from pre-existing and chronic dermatoses, such as psoriasis or eczema. The pathogenesis of erythroderma is poorly understood. Psoriasis is an autoimmune, inflammatory skin disease characterized by various variants, including plaque psoriasis, guttate psoriasis, erythrodermic psoriasis, and pustular psoriasis. These variants share three clinical features in common: erythema, thickening, and scaling (1,2,3). Erythrodermic psoriasis, in turn, has an incidence of approximately 2%, representing a severe variant that constitutes a dermatological emergency (5). Patients develop coalescent erythema, scaling, and exfoliation, affecting 90% of the body surface. The course of this dermatosis, as well as its prognosis, will depend on the severity, age group, presence of additional infections, and treatment adherence (4). The objective of presenting this case is to review the current and timely treatment of this rare entity and to bring attention to this “could be” fatal dermatosis.

CLINICAL CASE

An 82-year-old man, at least 10 years on treatment with antihypertensive medications, was diagnosed with benign prostatic hyperplasia 3 years ago on treatment with tamsulosin. No history of previous dermatosis. History of smoking with 20 cigarettes per day. He came with a condition of two months’ evolution, characterized by intermittent fever, chest pain, and dyspnea. He received non-dermatological consultation and treatment with intramuscular steroids and antibiotics for a presumable infection of the urinary tract, with no response. After two weeks, dermatosis was added to the trunk, which spread to the scalp and extremities, palms, and soles. The examination revealed a generalized dermatosis that spared only the mucosa; characterized by diffuse erythema, desquamation with whitish scaling, in a few areas with erythematous-scaly plaques of whitish scaling, in the rest of the skin and annexes, nails were involved too. (Fig.1,2,3,4,5,6)



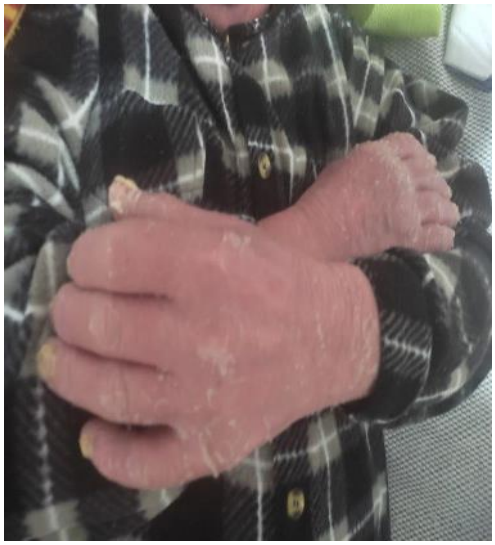
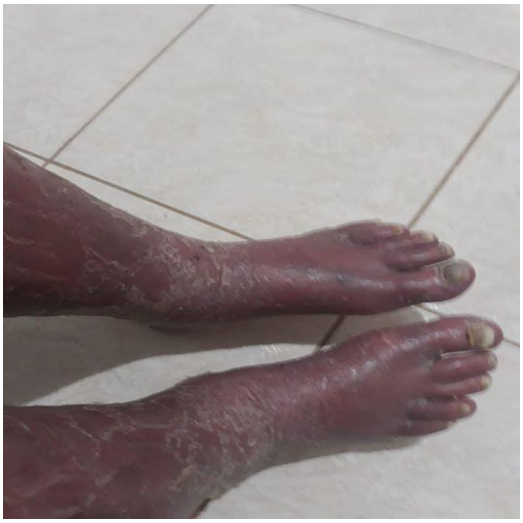


Figure1 et 2. Diffuse erythema, desquamation, and whitish scaling in both hands and feet.



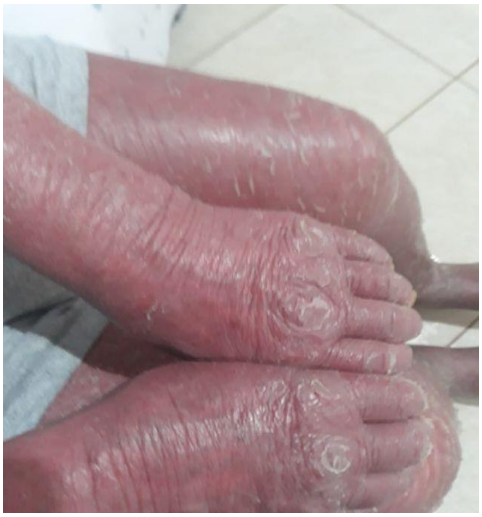
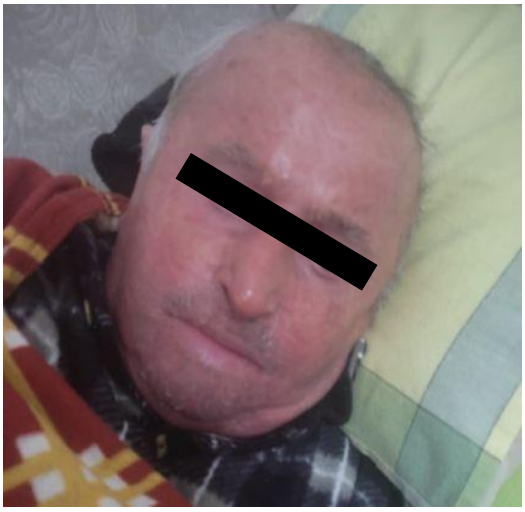
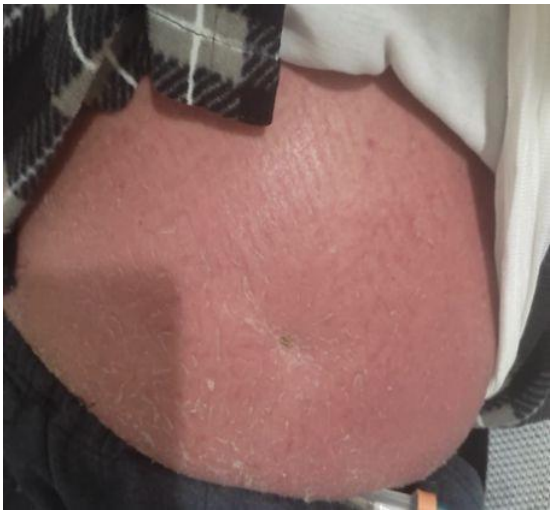


Figure 3 et 4. Whitish scaling, diffuse erythema of legs and arms, bilateral subungual hyperkeratosis.





Figures 5 et 6. Diffuse erythema, whitish scaling in the face, head, and trunk

The patient had a fever of 38 °C with leukocytosis of 15,700 mm^3, with neutrophils 76%, the blood chemistry showed no alterations. Systemic inflammatory response syndrome and erythroderma were integrated. The skin biopsy reported epidermis with psoriasiform hyperplasia, mild spongiosis, neutrophil infiltrate in the superficial layers, and parakeratosis, papillary dermis with dilated vessels and intense chronic inflammation. (Fig.7) With a broad differential diagnosis and no previous history of psoriasis, the diagnosis of EP in this patient was particularly challenging. In this case, a correlation of subtle pathology and clinical history led to the diagnosis of EP.

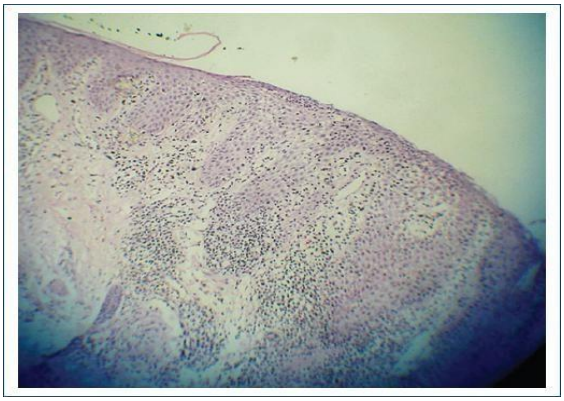


Figure 7. Epidermal psoriasiform hyperplasia with mild spongiosis, neutrophil infiltrate, parakeratosis; in papillary dermis with dilated vessels.

Treatment for EP includes adequate hydration, topical steroids, and vitamin D analogs, management of associated infections, and immunosuppression for uncontrolled symptoms. Methotrexate at a dose of 12.5 milligrams was started weekly for 3 months at a reduced dose, along with folic acid 5 mg once a week, emollients, and soap substitutes. The patient showed good skin improvement after one month. Liver function tests and blood counts remained normal. Follow-up was scheduled for 3 months, continuing topical treatment with emollients, 20% urea in thick skin areas, and topical corticosteroids. The patient is currently being followed up by the dermatology service.

DISCUSSION

Erythrodermic psoriasis is a rare and severe variant that affects 1-2% of patients with psoriasis, being the most common cause of erythroderma, responsible for approximately 25% of cases (5). Risk factors for the development of the disease include genetic factors, environmental factors, most involved are smoking, obesity, alcohol consumption, the intake of some drugs such as beta blockers, lithium, antimalarials, streptococcal infection, HIV. In the case here described, smoking and infection of the urinary tract were found to be the triggering factors (8,9). An important trigger for erythrodermic psoriasis is treatment with systemic glucocorticoids or abrupt discontinuation of systemic drugs such as methotrexate or cyclophosphamide (10). The pathogenesis of psoriasis is triggered by activated dendritic cells that produce IL-23 and TNF-α, stimulating the activation of CD4 Th17 lymphocytes and cytotoxic CD8 lymphocytes, which secrete IL-17 and IL-22 that sustain the inflammatory process and generate the epidermal hyperproliferation characteristic of the disease (11). The main clinical manifestation of erythrodermic psoriasis is the appearance of a generalized confluent erythema that affects between 75-90% of the total body surface. It has a rapid onset over several days or a gradual onset over several weeks, followed by exfoliation and desquamation of the skin that develops several days after the appearance of the erythema. The desquamation is usually accompanied by pain, itching, and photosensitivity in the areas with the greatest exfoliation activity. Most patients present nail disorders. The extra-cutaneous manifestations are fever, chills, malaise, tachycardia, arthralgia, and lymphadenopathy (12). The diagnosis should be made by investigating the personal or family history of psoriasis, history of possible triggers for the disease, and coexistence of clinical characteristics, emphasizing a physical explanation of the skin. No serological test confirms the diagnosis of erythrodermic psoriasis, so it is essential to perform a skin biopsy. The histological characteristics that most support the diagnosis of erythrodermic psoriasis were acanthosis, diffuse parakeratosis, diffuse hypogranulosis, and the presence of neutrophils in the epidermis and hypodermis. The histological characteristics in the biopsy were compatible, which confirmed the diagnosis in our case (13). Among the complications of erythrodermic psoriasis, hemodynamic alterations such as peripheral edema, high-output heart failure, shock, or acute renal failure have been described along with the thermoregulatory alterations and infections due to loss of the skin barrier. Being a systemic inflammatory disease, it is associated with several comorbidities, including psoriatic arthritis, uveitis, cardiovascular disease, metabolic and non-alcoholic syndrome (14,15). There is no consensus on the management of erythrodermic psoriasis. The most useful support recommendations are hemodynamic and body temperature monitoring, fluid and electrolyte replacement, nutritional support, treatment of associated infections, skin care with moist dressings, hydration, oatmeal baths, and topical corticosteroids to reduce pain and itching (16). Systemic therapy should be started immediately and should be chosen with respect to severity, patient comorbidities, and drug availability. The recommended first-line therapies are: cyclosporine (with an initial dose of 3 to 5mg/kg/day), which produces significant improvement in the first month and complete remission after one year. Infliximab (with an initial dose of 5mg/kg at week zero, two, and six, followed by 5 mg/kg every 8 weeks) (17,18). Alternative therapy is Methotrexate (with a weekly dose of 7.5mg to 25mg per day with folic acid supplementation 1mg/day), observing improvement after the first weeks of starting treatment (19). In countries with little access to drugs such as cyclosporine, infliximab, or new biologic therapies, like in Albania, either due to high cost or adverse effects, methotrexate and oral corticosteroids have been chosen as a second line. Our case had

a good response with methotrexate, but represents a call for help and attention to this rare entity.

CONCLUSIONS

This case represents an unusual presentation of EP in a patient without a prior history of psoriasis. When patients are critically ill with an erythrodermic rash, it is important to consider EP in the differential diagnosis. Skin biopsy and histological analysis aid in the diagnosis of EP, given the broad differential associated with erythrodermic skin reactions. Our case report may serve as a call for healthcare professionals to be aware of the symptoms and characteristics of exfoliative psoriatic erythroderma, including the need for immediate and appropriate treatment. With accurate treatment and supervision, patients can achieve significant improvement and avoid life-threatening complications.

Conflicts of interest: The authors declare that they have no conflicts of interest.

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