

Erythema annulare centrifugum: a rare case report

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Abstract

Erythema annulare centrifugum (EAC) is a rare skin disease that is thought to be caused by interactions between inflammatory cells, mediators, and foreign antigen substances. It typically starts as erythematous macules or urticarial papules that enlarge peripherally to form an arcuate or polycyclic plaque. Symptomatic relief is the main therapy for this disease because no effective treatment is yet available. We report the case of a 50-year-old male with reddish pruritic patches on both arms, around the thighs, on the abdomen, and on the buttocks. The complaint started 3 years prior to treatment and often recurred. Fungal examination was negative, and histopathological examination supported a diagnosis of EAC. The patient was prescribed a topical corticosteroid, topical antifungal, and antihistamine. Although the lesion only showed minimal improvement, the patient experienced significant reduction in pruritus with no occurrence of new lesions. Despite being a rare disease, EAC must be considered when encountering an annular erythematous plaque.

Keywords: annular, erythema annulare centrifugum, antifungal, corticosteroid

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Introduction

Erythema annulare centrifugum (EAC) is a chronic skin disorder that presents as a centrifugally-enlarging thin annular erythematous plaque. This unusual inflammatory condition is characterized by a polycyclic erythematous eruption that slowly enlarges at a rate of 1 to 3 mm/day. EAC was first described by Darier in 1916, and it was classified in 1978 by Ackerman into superficial and deep forms (1, 2).

The incidence of EAC is unknown. Adults are the population most often affected; however, this disease may also occur in children, and neonatal EAC cases have also been reported. Retrospective studies have shown that the age of onset ranges from 5 to 90 years, with the average age ranging from 40 to 50 years (3, 4).

Diagnosis is established on history, clinical features, and supporting examination. Diagnosis of EAC is generally carried out clinically by observing the presence of erythematous macular lesions or urticarial papules that enlarge peripherally to form an annular, arcuate, or polycyclic appearance. Histological examination typically shows characteristic changes in the form of epidermal parakeratosis and spongiosis with superficial perivascular infiltrates (5). No treatment seems to be effective for this disease and there is still a lack of consensus regarding the best approach. Current choices are focused on treating the underlying subjective complaints, which include topical use of steroids and antihistamine to reduce itching (3).

We report a 50-year-old man with a diagnosis of EAC. The patient underwent outpatient care and showed improvement after being prescribed a topical corticosteroid and oral antihistamine.

Case report

A 50-year-old man came to the dermatology and venereology outpatient clinic of Wahidin Sudirohusodo Hospital, Makassar, Indonesia with a chief complaint of pruritic, thick, red scaly patches on both arms, around the thighs, and on the abdomen and but-

tocks that had been present for the previous 3 years and had worsened 3 weeks before admission (Fig. 1).



Figure 1 | Initial presentation with reddish pruritic patches (a) on the buttocks and around the thighs; (b) on the abdomen, and (c, d) on legs.

Initially, the complaint appeared on the back of the leg in the form of pruritic reddish spots that extended to both thighs, the buttocks, and both arms. A history of a similar complaint was previously recognized but never completely resolved. He had previously been diagnosed with a fungal infection and psoriasis without any significant improvement. The patient also had diabetes

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mellitus. There was no history of hypertension, drug allergy, or similar complaint in the family.

From the physical examination, the patient's vital signs were within normal limits. His bodyweight and height were 65 kg and 167 cm, with a body mass index (BMI) of 23.31. The results of a general physical examination were within normal limits. Dermatological examination showed annular hyperpigmented plaques on both lower limbs and erythematous, scaly macules and papules on the abdomen, gluteus, and upper and lower extremities.

The patient was diagnosed with EAC with differential diagnosis of tinea corporis et cruris and psoriasis vulgaris. A potassium hydroxide microscopic examination was performed and was negative for a fungal infection. The patient was prescribed cetirizine 10 mg once daily, topical application of 3% salicylic acid and desoximethasone cream to be applied to the feet, topical application of 3% salicylic acid, 6% benzoic acid, petroleum jelly, miconazole 2%, and hydrocortisone 2.5% to be applied on erythematous plaques on the abdomen, buttocks, and hands, and fusidic acid cream for the excoriations. Histopathological examination of the lesions on the hand and foot showed epidermal hyperkeratosis, focal parakeratosis, mild spongiosis, mild papilla dermis edema, lymphocytes, and perivascular histiocytes (Fig. 2), which supported the diagnosis of EAC. After 2 weeks of treatment, although the improvement of the lesion seemed minimal, the patient reported marked improvement of subjective complaints with no occurrence of new lesions (Fig. 3).



Figure 3 | Significant clinical improvement of the lesions 2 weeks after therapy.

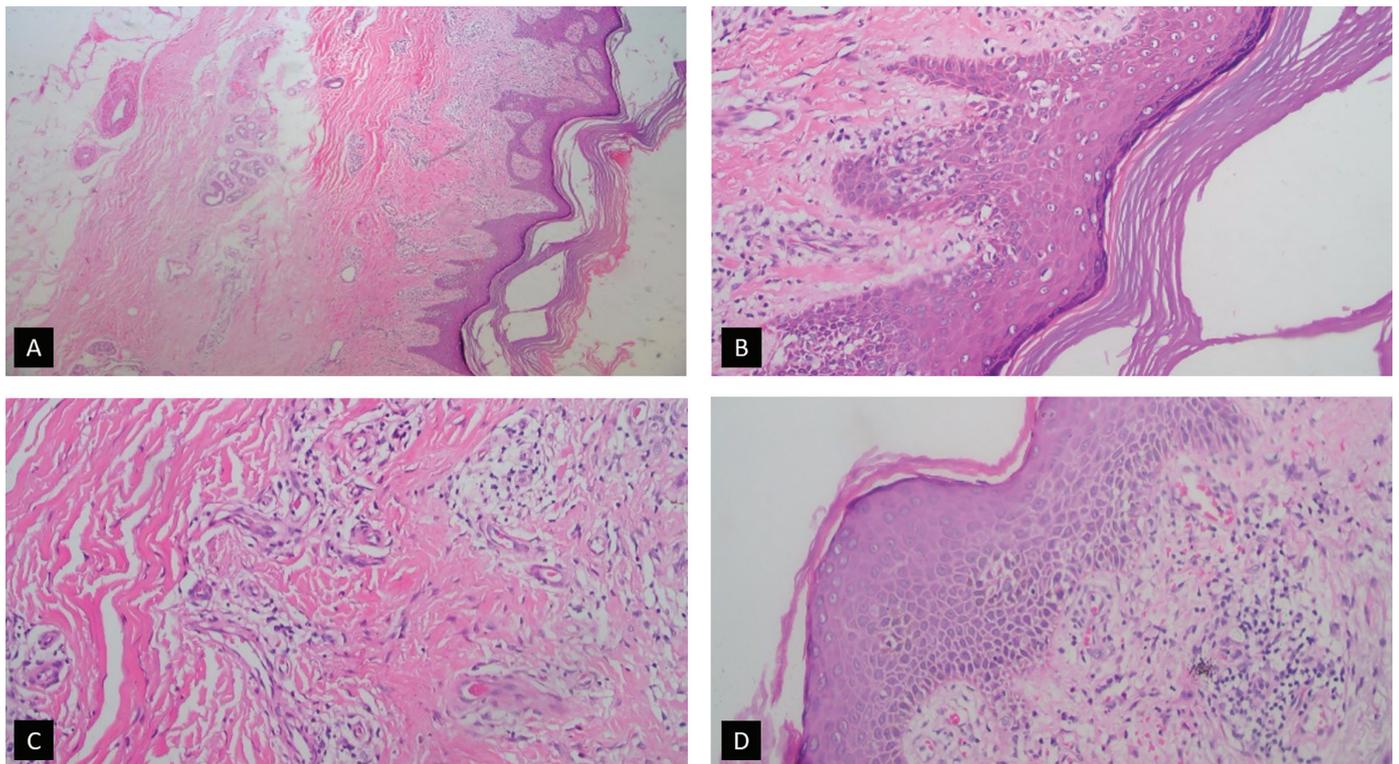


Figure 2 | Histopathological examination at (a) 4× and (b) 20× magnification showed hyperkeratosis, parakeratosis, and mild spongiosis; (c) lymphocytes and perivascular histiocytes (20×); (d) lymphocytes, perivascular histiocytes, hyperkeratosis, and mild spongiosis (20×).

Discussion

In this case, the patient complained of pruritic thick scaly patches on both arms, around the thigh, and on the abdomen and buttocks. EAC appears as one or more lesions that start as erythematous macules or urticarial papules and enlarge with peripheral extensions that form a ring, arcuate, or polycyclic pattern. The lesions appear as circular macules (discoloration of the skin) or

ovoid, or plaques (dense content lesions more than 1 cm in diameter that are predominantly surface rather than deep lesions) with a clear erythematous and central border (6, 7). EAC is divided into superficial and deep forms (8). The superficial form appears as fine scales or squares that are more prominent on the inner aspect than the edges. The lesions spread gradually to form a large annular plaque with central clearing, with the edge of the lesion often increasing by several millimeters a day. After a varied pe-

riod, the lesion can disappear or often change into a new lesion. In the deep form, there is no scale and the shape of the rings is infiltrated (6). Based on these descriptions, our case could be classified as superficial EAC.

A history of similar complaints that never completely resolved was recognized. The patient had been treated for fungal infection and psoriasis with insignificant improvement. Fungal infection was initially suspected due to the annular and central clearing nature of the lesion, which resulted in scaly erythematous annular lesions of different sizes, sometimes with papular, vesicular, or pustular edges (7). However, although central clearing is also present in dermatophytosis, the hyperkeratotic border observed in this case was not typical for tinea infections. In addition, microscope examination revealed no fungal elements. Psoriasis, the other differential diagnoses, also presents with thick-scaled erythematous plaques; however, the central clearing appearance in this case was not typical for psoriasis. This was corroborated by the histopathological examination, which did not reveal histopathological features of psoriasis such as elongated rete ridges and Monroe's abscess (5, 9).

The histopathological description of EAC is not pathognomonic; however, it is useful in eliminating other differential diagnoses. Histopathologically, EAC shows parakeratosis and spongiosis with superficial perivascular infiltrates (5). There is minimal papillary skin edema and no spongiosis (6, 10).

The etiology and pathogenesis of EAC are yet to be fully understood; it has been suggested that EAC may result from a delayed-type response to a variety of antigens. This phenomenon has been postulated to occur in relation to many conditions, such as infection, autoimmune diseases, malignancy, and exposure to drugs and food. However, in most cases, there is no detectable predisposing condition (it is idiopathic) (3).

Therapy for EAC is mainly directed at the subjective complaints (11). The patient was prescribed cetirizine 10 mg / 24 hours orally, 3% salicylic acid, and desoximetasone on the feet, and a mixture of 3% salicylic acid, 6% benzoic acid, miconazole, and hydrocortisone for lesions on the trunks and extremities. Topical steroids may reduce the erythema and decrease the superficial inflammation; potent topical corticosteroids may improve the appearance of the lesions, although they may not affect the overall clinical

course (11). Oral antihistamines may relieve pruritus, but they also do not alter the clinical appearance or course of the disease (12). For those with pruritus, corticosteroids and antihistamines may be of some benefit (11).

Although microscopic examination was negative for fungal infection, miconazole cream, a topical antifungal, was nonetheless given. One study showed that 40% of patients with EAC had a concomitant superficial dermatophyte infection (13). In addition, an antifungal has sometimes been useful and can be empirically beneficial (6). Although the clinical improvement seemed minimal, the patient showed marked improvement of subjective complaints. Due to limited clinical data, the main goal of current treatment options focuses on treating the underlying condition and alleviating subjective symptoms, including reducing itching (3, 7). Although oral corticosteroids have been shown to be beneficial, they were not administered in this case because the patient had diabetes mellitus. In addition, rebound phenomenon often occurs after corticosteroid withdrawal. Other agents such as tacrolimus, calcipotriene, oral metronidazole, etanercept subcutaneous, and subcutaneous α interferon have also been shown to be useful (1). Other therapies such as topical vitamin D analogs can be combined with ultraviolet irradiation as another therapeutic option (6).

EAC is a self-limiting disease that may last from only a few weeks to decades (6). EAC usually resolves after the underlying disease is treated (1, 14). The disease has a good prognosis, but lesions tend to recur over months to years, with most cases finally resolving spontaneously (15).

Conclusions

Although rare, EAC must be considered when encountering an annular erythematous plaque. To date, evidence for an effective treatment is still lacking, and therapy is mainly directed toward relieving symptoms.

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