SkIndia Quiz 6
Multiple recurrent erythematous scaly annular plaques in a child

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A nine-year-old female child was referred for evaluation of relapsing, self-healing, slightly itchy, annular eruptions, mainly involving the face, trunk, and extremities, which had recurred yearly for the last three years [Figure 1]. The eruptions began as small erythematous papules [Figure 2] coalescing to form plaques with a central clearing and centrifugal spread. No fever, general symptoms or mucosal lesions were present. Her medical history was uneventful. A physical examination revealed an apparently healthy child with multiple 2- to 8-cm erythematous annular lesions on her chest, back, arms, and thighs, leaving occasional residual hyperpigmentation. The lesions used to regress after a few months of onset only to recur again. During the last three years, the disorder was partially controlled with moisturisers ,topical steroid and antihistamines as the main form of treatment. Neither inguinal nor axillary lymphadenopathy was present.

The patient's routine laboratory investigation, including hematological and biochemical analysis, and antinuclear antibody, disclosed no abnormalities. Direct potassium hydroxide examination and cultures for fungi and bacteria from the lesions failed to identify any microorganisms. The chest radiographs were consistently normal. The histopathology of the lesional biopsy is shown in Figures 3 and 4.

WHAT IS THE DIAGNOSIS?

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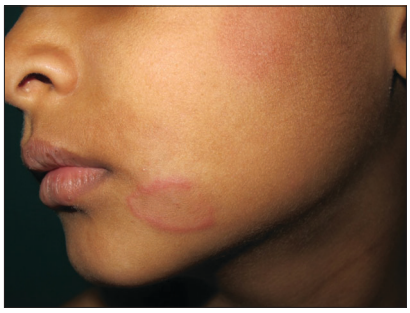

Figure 1: Erythematous annular mildly scaly plaque with central clearing over face

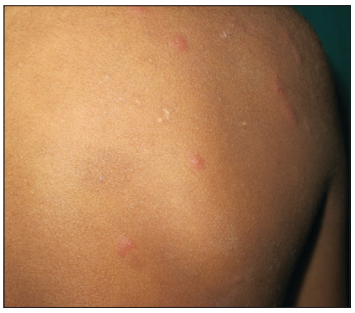

Figure 2: Early erythematous scaly plaques over the back

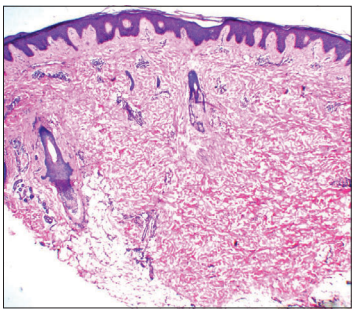

Figure 3: Skin biopsy from a lesion showing mild epidermal hyperplasia, with moderately dense superficial and dense perivascular, predominantly lymphocytic, infiltrate (H and E, ×50)

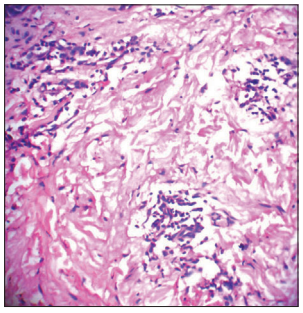

Figure 4: High-power view showing coat-sleeve-like lymphocytic infiltrate around the mid-dermal blood vessels (H and E, ×100)

ANSWER

Erythema annulare centrifugum (EAC)

Microscopic findings and clinical course

Lesional biopsy revealed slight psoriasiform epidermal hyperplasia, with a perivascular lymphohistiocytic infiltrate of variable density in the papillary and mid-dermis, with occasional eosinophils. Neither fibrinoid necrosis nor vascular damage was detected [Figures 3 and 4]. Periodic acid Schiff's stain was negative.

The patient was advised a mid-potentency topical steroid cream with moisturisers and oral antihistamines. The lesions improved over two weeks and patient is under follow-up.

DISCUSSION

Erythema annulare centrifugum (EAC), owing to its distinctive annular and polycyclic pattern, belongs to the family of 'figurate erythemas' which includes erythema gyratum perstans, erythema exudativum perstans, erythema marginatum perstans, erythema microgyratum perstans, erythema perstans, erythema figuratum perstans, erythema simplex gyratum, and eosinophilic annular erythema.[1]

Darier first described this condition, which does not affect a particular sex or age group. Two types of EAC are known, superficial and deep. In the superficial type there is desquamation following the advancing border and itching is more frequent. In the deep type, a dense perivascular infiltrate is seen, composed principally of lymphocytic cells distributed in 'coat-sleeve-like' manner in the middle and lower dermis. The endothelial lining of the affected vessels is intact. Occasionally, histiocytes and eosinophils are present. The epidermis is usually normal. The superficial variant histologically shows a superficial perivascular dermal lymphocytic infiltrate with papillary edema, spongiosis, and parakeratosis.[2] In the deep type of erythema annulare centrifugum there is an indurated and apparent border, without desquamation, and the lesion rarely itches. Histologically the infiltrate is deeper and involves the deep dermal vessels too.[2]

The pathogenesis of EAC is unknown. However, various associations of EAC, include *infections* like dermatophyte infections, intestinal candidiasis, *molluscum contagiosum*, Epstein-Bar virus, genital herpes, urinary tract infections, tuberculosis, and ascariasis; *neoplasm*s like bronchial, prostate, nasopharyngeal, ovarian, rectal, hepatic tumors, lymphoma, and leukemia; *drugs* like aldactone, amytriptilline, ampicillin, cimetidine, hydrochlorothiazide, penicillin, and piroxicam; *hematological disorders* like polycythemia vera, myelodysplastic syndrome, hypereosinophilic syndrome; and *autoimmune conditions* like hyperthyroidism and Hashimoto's thydroiditis.[1-3]

Diagnosis is mainly based on the characteristic clinicopathological picture. Laboratory test results are usually unremarkable, unless there is an associated disease.

The differential diagnosis includes tinea corporis, granuloma annulare, erythema multiforme, sarcoidosis, cutaneous lymphoma, benign lymphocytic infiltrate, lupus erythematosus, leprosy, and so on. These can largely be excluded by clinical examination, histopathology and bed side tests like the potassium hydroxide mount. Erythema annulare centrifugum, although a self-resolving disease, usually has a waxing and waning course of variable duration. It may last for months to years.[2]

No routinely used therapy is consistently effective unless a triggering factor can be identified and eliminated. However, no cause is found in most cases. Glucocorticoids are used therapeutically both in topical and systemic forms.[3] Oral metronidazole, topical Calcipotriol, and ultra-violet B phototherapy have been recently found to be effective in EAC, in a few cases.[3-5]

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