Erythema Annulare Centrifugum

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Abstract

Erythema annulare centrifugum (EAC) is classified as a gyrate erythema among other erythemas. It is rare and of unknown etiology. Some authors believe it may be a hypersensitivity reaction to infections whether that be viral, bacterial, fungal or parasitic. Occasionally, it may be drug induced. The eruption has also been reported with chronic systemic disease and rarely, with internal malignancy. In most instances however, no cause can be found hence it may be idiopathic. Clinically it presents with annular or arcuate lesions with a "trailing scale" behind an advancing erythematous border in the superficial type, which is usually pruritic. The deep type presents with lesions of similar configurations but is usually nonscaly and nonpruritic. A case of EAC is presented with a short review of the literature.

Case Report:

A 61 year old Sudanese gentleman, presented with a six months history of multiple annular lesions on the left leg, both thighs and the left upper arm, all lesions showed a trailing scale, and a hyperpigmented or erythematous border (Fig.1), the largest lesions on the leg was pruritic while the others were associated with a burning sensation. The patient gave history of right vocal cord carcinoma about three years prior to his cutaneous eruption, which was treated surgically followed by radiotherapy. He has no other medical problems, is not known to be allergic to any food or medication, and has no previous history of a similar eruption or other skin disease.

A battery of investigations were done including a complete blood count, renal function test liver function test, and erythrocyte sedimentation rate which were all within normal limits. His chest x-ray showed no abnormal findings, and stool for occult blood was negative. Skin scrapings examined by direct microscopy and culture were negative for fungi. A biopsy obtained from a left thigh lesion showed epidermal changes of parakeratosis and mild spongiosis, while the dermal changes included a few extravasated red blood cells and a perivascular lymphocytic infiltrate (Fig.2). Special stains for fungi yielded negative results. These features were consistent with the clinical diagnosis of EAC. The rash responded well to applications of clobetasol propionate ointment, all lesions cleared within two weeks.

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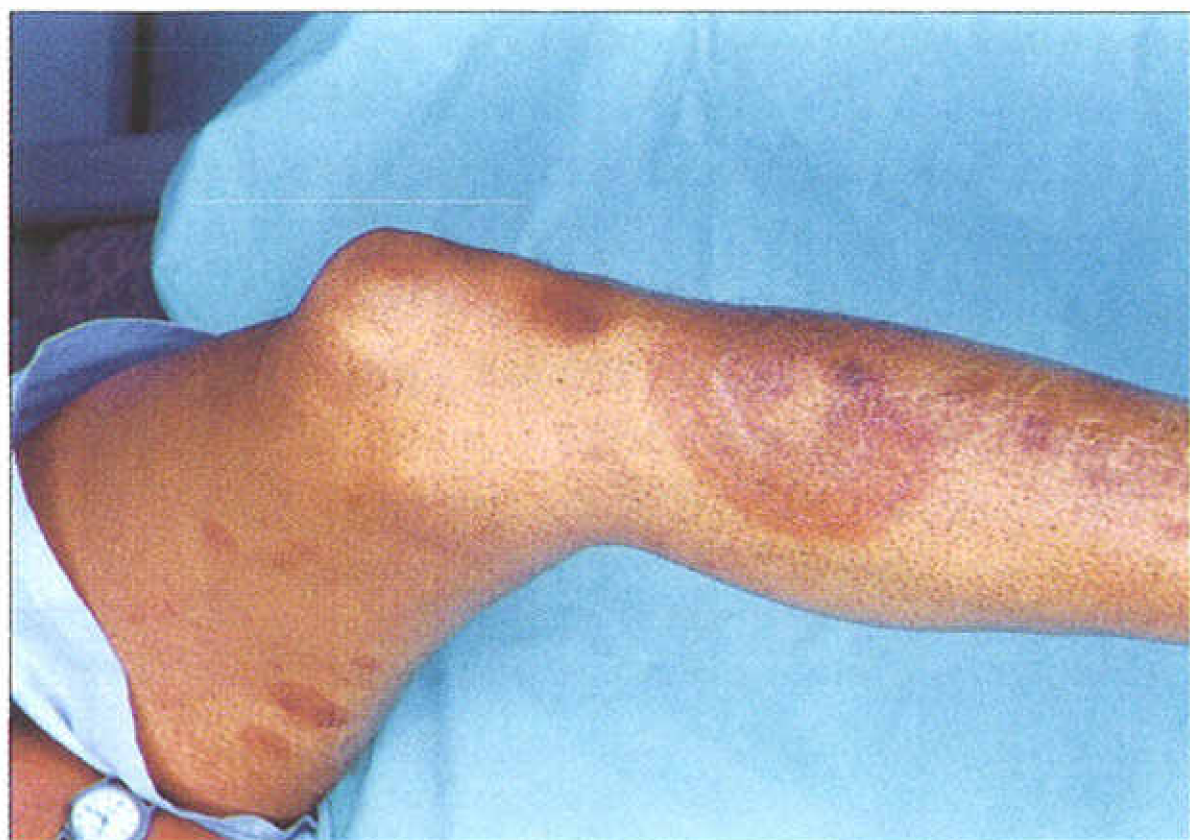
Discussion:

Erythema annulare centrifugum, described by Darier[1] in 1916, is regarded s one of the gyrate or figurate erythemas. It is an uncommon disorder but the incidence is difficult to determine because most reports involve single cases or small series of patients. In one review the incidence was estimated to be one case per 100,000 population per year.[2] The eruption is characterized by a slowly migrating configurate lesions, which have a predilection to the lower extremities and the trunk. The lesions gradually expand with central clearing and eventually resolve spontaneously within a few weeks or follow a chronic recurrent course, which may continue for many years. Erythema annulare centrifugum can occur in all age groups and there is no predilection for either sex.[2, 3]

The classification of gyrates erythemas has always been associated with controversy. Since Darier's original description of this disorder, different types of Erythema that have an annular configuration have been included as cases of EAC. In addition, new terms have been created to classify different histologic and clinical variants, which led to considerable confusion. In an attempt to clarify this confusion, Ackerman [4] suggested that there are two types of gyrate Erythema. A superficial scaly type with histologic features that include epidermal changes of parakeratosis and spongiosis with a superficial perivascular infiltrate and a deep non-scaly type, as described by Darier, which is characterized histologically by superficial and deep perivascular lymphohistiocytic "cuffing" with normal epidermis. Bressler and Jones [5] studied patients with gyrate erythemas and favored using the terms deep and superficial gyrate erythemas. The deep variety presents with an indurated non-scaly border and is generally not pruritic, whereas lesions of the superficial form tend to be more often pruritic and has a characteristic "trailing scale" behind an advancing erythematous edge. The



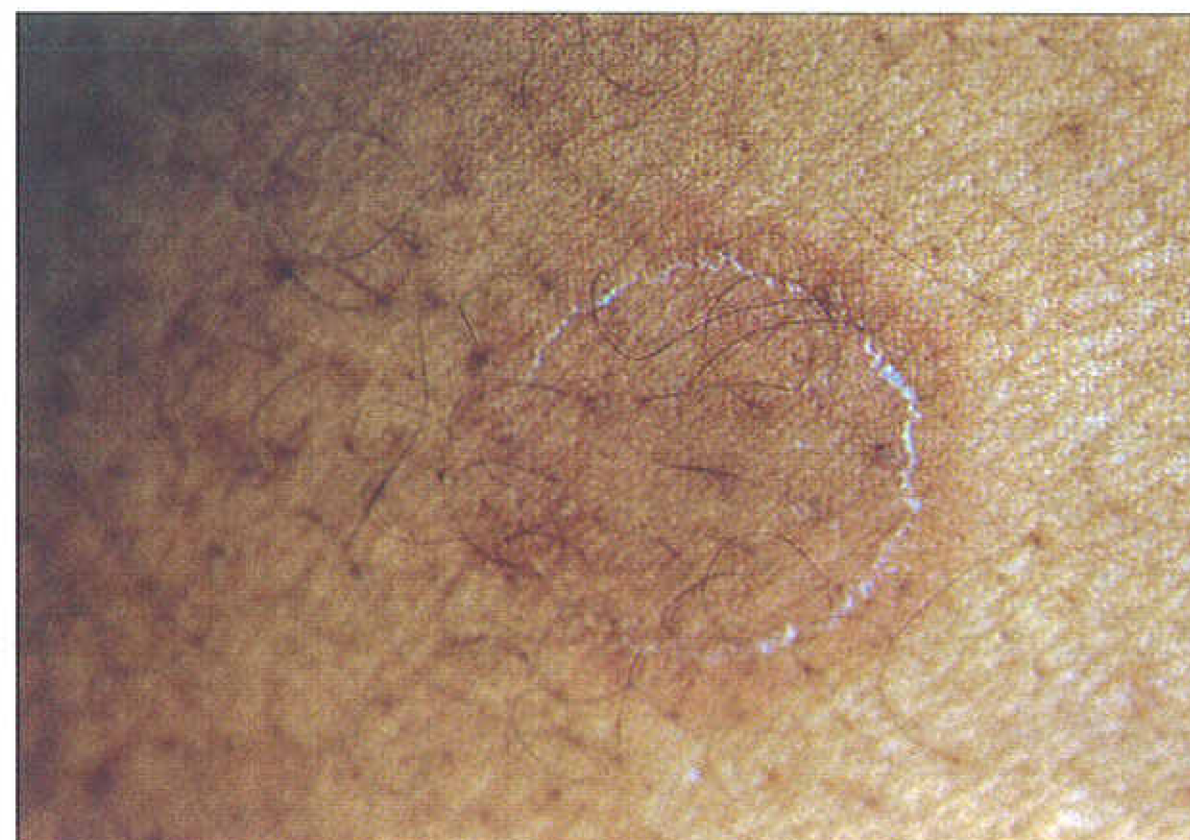
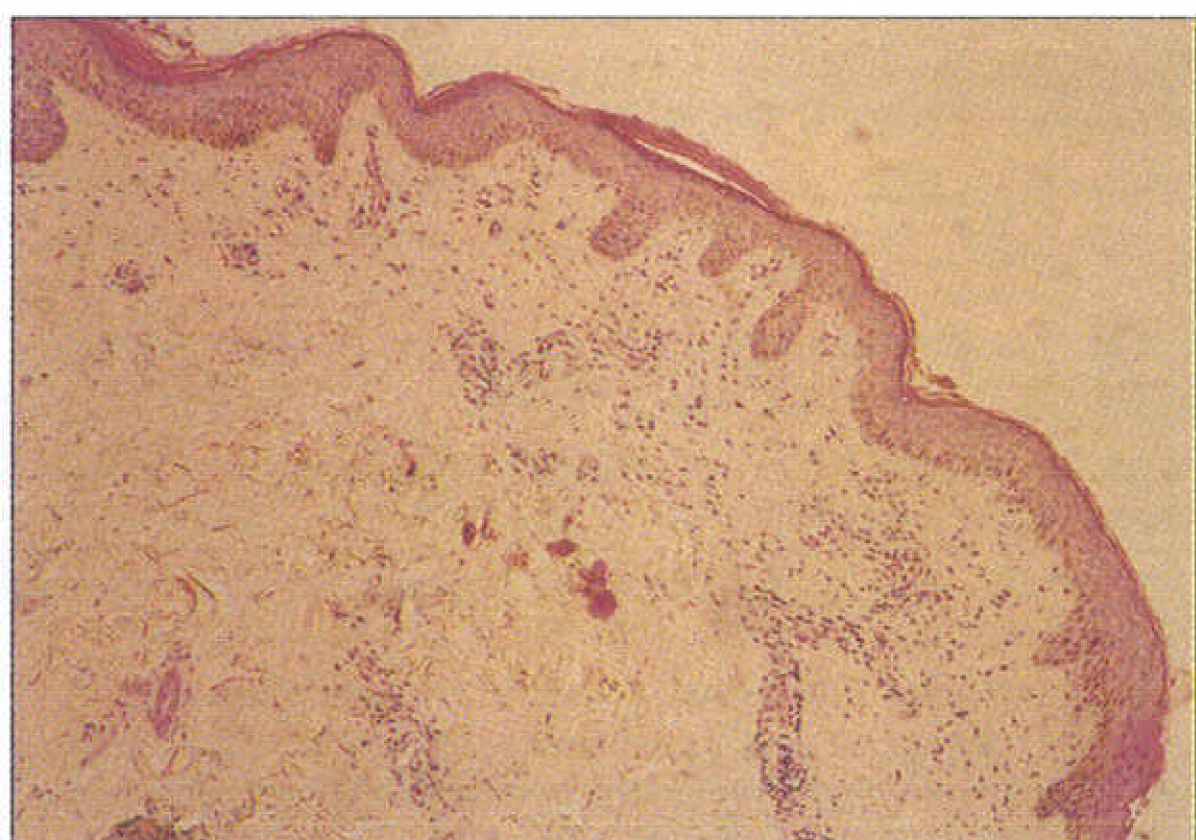


Figure 1: (A and B) Close-up of an annular lesion with a trailing scale behind the advancing erythematous border.



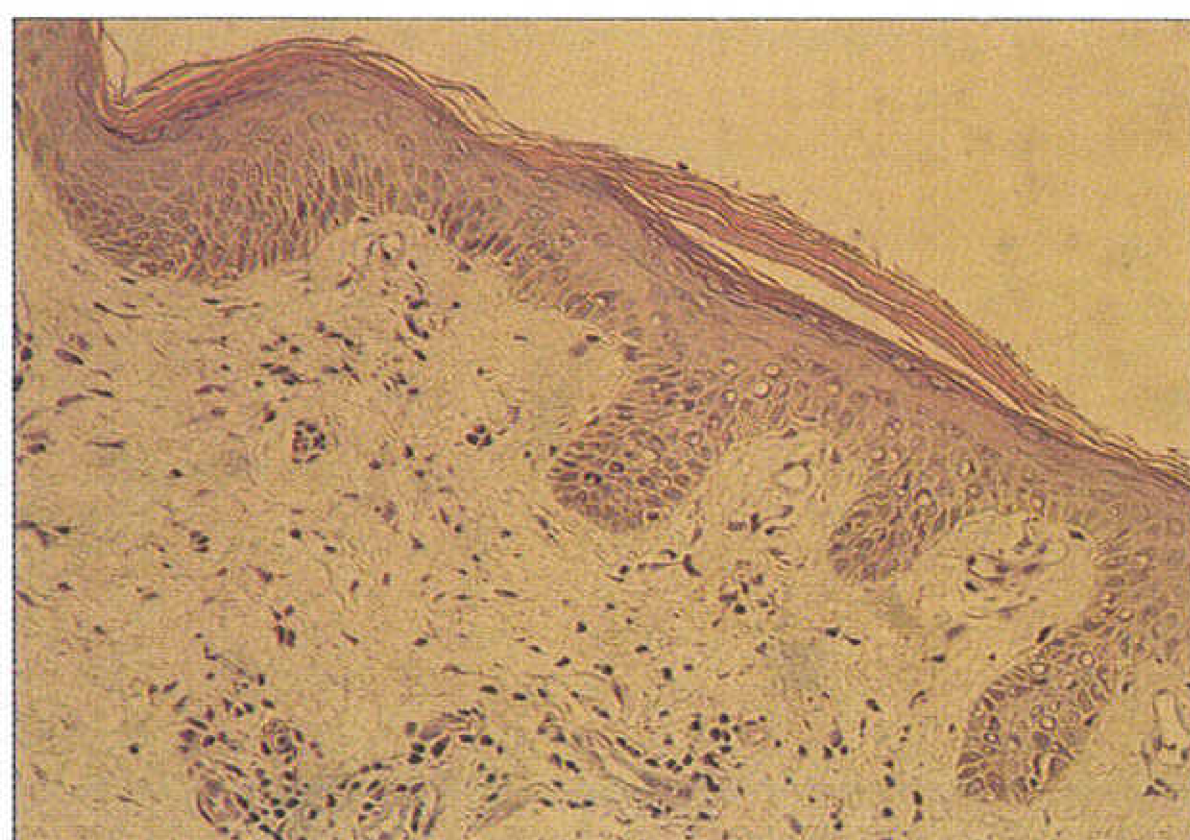


Figure 2: (A and B) Perivascular lymphocytic infiltrate in the upper and mid dermis.

eruption in our patient showed clinical and histological features consistent with the superficial form.

The pathogenesis of EAC is still unknown. In some patients the eruption has occurred in association with a variety of disorders, however, a causal relationship could not always be proved. [2, 3] Erythema annulare centrifugum has been reported in association with different infections such as tinea, candida albicans, E. coli, tuberculosis, ascariasis and viral infections such as molluscum contagiosum and the Epstein-Barr virus. [6-12] In addition, it has occurred with various systemic diseases including thyroid and liver disease, hypereosinophilic syndrome and recurrent appendicitis.[13-16] It has also been described as a hypersensitivity reaction to blue cheese penicillium of following the ingestion of some drugs. [17-20] It may also occur in pregnancy or follow the menstrual cycle. [21, 22]

Although early reports [23, 24] of EAC associated with malignant processes has led to the suggestion that it may be a marker of underlying malignancy, some authors have questioned the significance of these associations. Mahood [2] studied a series of 24 cases and concluded that the incidence of malignancy in patients with EAC is not increase. It must be noted however, that there are documented cases

of close association between EAC and various malignances including Hodgkin's disease, multiple myeloma, prostatic carcinoma, carcinoid of the bronchus, nasopharyngeal carcinoma and malignant histiocytosis. (25-29) Interestingly, the course of the eruption in these patients tended to parallel that of the underlying malignancy.

Even though our patient had a history of vocal malignancy it did not correlate temporally with the EAC lesions since it had occurred about 3 years prior to the onset of the eruption. The possibility that EAC may be heralding a new malignancy or a recurrence of the old one was ruled out by thorough clinical and laboratory evaluation. However, we shall continue to monitor our patient for any evidence of malignancy that may appear in the future as EAC has been reported to precede a malignancy by two years and there fore, not necessarily always occur concurrently with the tumor. (29)

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