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ACRODERMATITIS CHRONICA ATROPHICANS AND MALIGNANT LYMPHOMA

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Abstract. The case is presented of a 66-year-old female patient with the constellation of acrodermatitis chronica atrophicans, a malignant lymphoma, in the previously unaffected skin, and also fibroid nodules. The necessity of checking patients with acrodermatitis for the development of malignancies is pointed out.

Recently Lagerholm et al. (9) reported on the first case of a basal cell carcinoma which had developed in the atrophic skin of a patient with acrodermatitis chronica atrophicans (a.c.a.). Case reports on tumour formation in this skin disease are rather numerous. They are mostly related to tumours on the basis of the altered skin. Even reticulum-cell sarcomas have been described (8, 11). Outside the acrodermatitic area, in clinically unaffected skin, only lymphocytomas (lymphadenosis cutis benigna Bäfverstedt) have been observed which are thought to have an etiologic connection with a.c.a. (1). Some short reports are concerned with the blastomatous degeneration of lymphocytomas (2, 7, 10). A primary lymphoma in the normal skin of a.c.a. has never been described.

The present paper reports on the very rare constellation of a.c.a., malignant lymphoma and fibroid nodules.

CASE REPORT

A 66-year-old female patient with the appearance on both forearms and legs of livid-reddish skin lesions which in some areas become thin, wrinkled, and atrophic. The skin disease began over 2 years ago, and about 1 year later some indolent, dull-red, solid nodules developed on the elbows (Fig. 1). One year subsequently, on the left acromion, a reddish and painless plaque developed which rapidly increased by marginal extension and the primary plaque became tumorous (Fig. 2). Only one enlarged lymph node was palpable in the left axilla. The inflammatory changes of a.c.a. and the fibroid nodules rapidly responded to treatment with penicillin, but the lymphoma did not recede until after X-ray therapy. Four weeks after irradiation several tumours of the same appearance as those first visible on the left acromion developed in the inflammatory skin on both sides of the right foot.

Laboratory investigations. E.S.R. 12/23 mm (Westergren method). Hemoglobin was 12.8 gm per 100 ml, erythrocytes 5.0 million per mm^3, reticulocytes 1.9% of erythrocytes. White cell count was normal. A direct Coombs test and tests for agglutinins were negative. Serum total protein was 6.9 gm per 100 ml with 1.2 gm per 100 ml gamma globulin. Slight cryoglobulinemia was present. The immunoelectrophoretic analysis revealed a marked increase in IgM (860 mg per 100 ml) without monoclonal character, IgA and IgG being within normal range. In a sternal marrow aspirate, plasma cells were numerous.

Histology. Biopsies were taken from the right upper thigh (*a*), and from the tumour on the left shoulder (*b*). One of the periarticular nodules was totally excised (*c*).

(*a*) The histological examination disclosed marked edema and infiltration of lymphocytes and histiocytes in the upper and middle layers of the dermis, compatible with an early stage of a.c.a.

(*b*) The tissue specimen excised from the tumour on the left shoulder is invaded throughout the entire corium and down to the upper layer of the subcutaneous tissue by a proliferation of medium-sized lymphoid cells and large reticulum-cell elements the nuclei of which are sharply demarcated, with light cytoplasma and clearly visible nucleoli. At certain points there are numerous atypical mitoses (Fig. 3) and in the area of dense infiltration, argyrophile fibres are demonstrable.

(*c*) The small nodule from the elbow reveals characteristic subcutaneous fibroid alterations but lacks any elastic fibres in the nodule which is surrounded by a lymphocytic infiltration.

DISCUSSION

Recently we reported on a patient with the association of a.c.a., fibroid nodules and a mono-

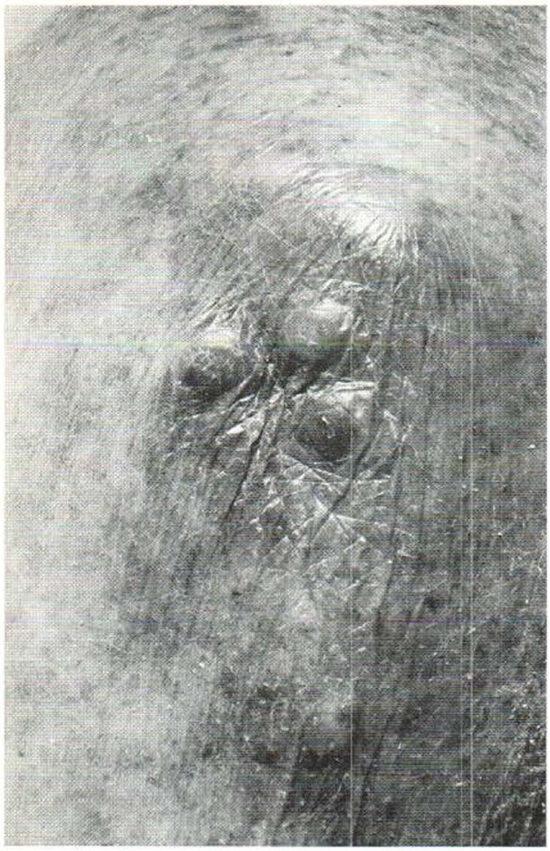

Fig. 1. Characteristic fibroid nodules in the atrophic and still partly inflammatory skin near the elbow.

clonal gammopathy (4). The coexistence of this skin disease and a lymphoproliferative disorder was considered as contributing to a further understanding of the potential importance of pathogenic

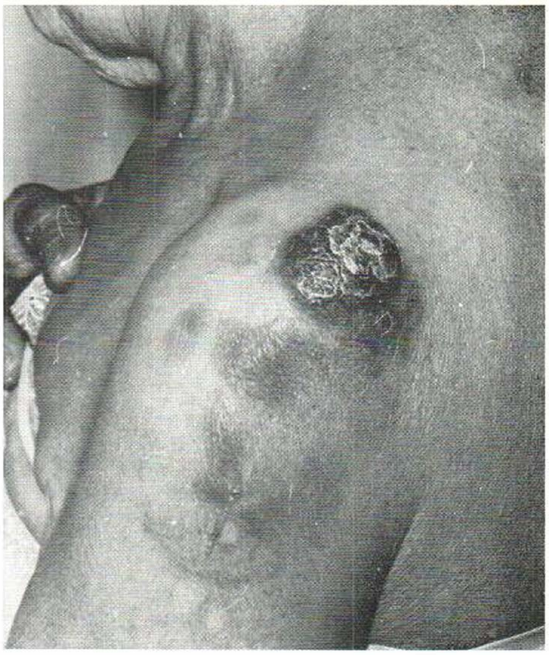

Fig. 2. Malignant lymphoma on the left shoulder with scaly and crusty appearance and peripheral extension.

relationships among these disorders. The case described here once more draws attention to this problem. It is in keeping with the statement of Hauser who first noted that a.c.a. is a general affection of the organism (5, 6). The malignant lymphoma of our patient had developed primarily in clinically unaffected skin. The connection with the skin disease is supported by the occurrence of new lymphomas in the acrodermatitic area of the right foot.

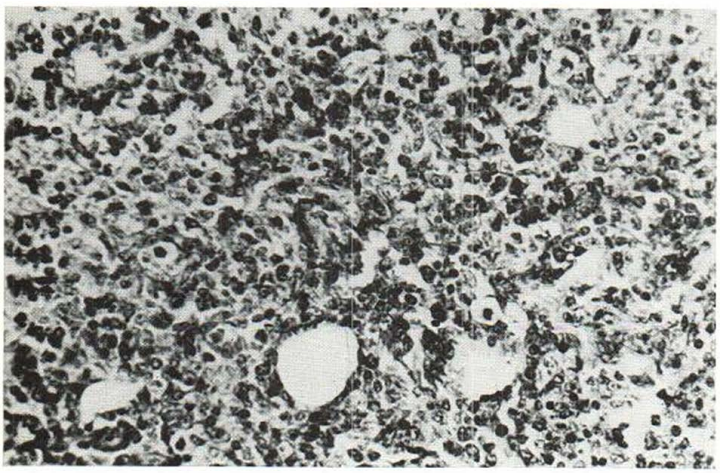

Fig. 3. Dense infiltration of the upper subcutaneous tissue by medium-sized lymphoid cells and large reticulum cells with numerous mitoses. Giemsa, × 130.

Tumorigenesis in a.c.a. obviously depends on the stage of the skin lesions. Whereas lymphoreticular tumours develop in the course of protracted infiltration, such tumours as fibromas, neuromas and myomas are to be found in the atrophic final stage (14). Above all, this is true of the squamous cell and basal cell carcinomas. The development of the skin carcinomas is probably due to the same mechanism as in carcinogenesis of the aged skin. The lymphoreticular disorders may have an immunological basis. Indications in favour of this hypothesis are the plasma cell infiltrations in the skin, lymph nodes and bone marrow (6), the high incidence of cryoglobulinemias, and the associations of a.c.a. with monoclonal gammopathy (4, 12, 13). The concurrent development of lymphoproliferative disorders may arise secondary to chronic antigenic stimulation, for example, by autoimmunization or chronic infection. Further investigation of the unknown triggering factor that leads to both the a.c.a. and the lymphoproliferative disorders are proceeding (3).

The present case with a.c.a., malignant lymphoma, fibroid nodules and a marked elevation of IgM is of theoretical as well as practical importance. Patients suffering from a.c.a. must be examined at regular intervals for the development of malignancies and immunological derangements.

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