

### SUMMARY

The terminology "lymphocytoma cutis" is confusing. It has many synonyms e.g. Spiegler-Fendt sarcoid, pseudo-lymphoma of Spiegler-Fendt, miliary lymphocytoma, lymphadenosis benigna cutis and benign lymphadenomatous granuloma.

Although the first case was described by Jadassohn in 1906, it was Kauffman-Wolf in 1921 who gave it the name lymphocytoma cutis.

Lymphocytoma cutis can occur at any age but most commonly it presents in late teenage and early adult life. Both the circumscribed and the disseminated forms occur two to three times more frequently in women than in men.

In the majority of cases the provoking factors are not known. Light sensitivity, insect bites, trauma, virus infection, co-existing malignant tumours (i.e. dermatofibrosarcoma protuberans), other dermatoses (e.g. erythema chronicum migrans and acrodermatitis atrophicans) and autoimmunity have been proposed. Most writers suggest that pre-existing lymphoreticular tissue undergoes hyperplasia.

Clinical diagnosis of lymphocytoma cutis is seldom conclusive without the aid of histologic examination. Lymphocytoma cutis is divided into localized and disseminated forms. Localized lymphocytoma cutis, the more common form has a short course. There may be isolated lesions or groups of lesions in localized areas. The areas of predilection

are the face, earlobes, nipples, vulva and scrotum. Typically the lesions are small, superficial, translucent, firm, non tender, pinkish-red to violet papules or nodules. The lesions of this variety may disappear spontaneously and usually without recurrences. The disseminated form is rare and tends to have a protracted course. Miliarial lesions or tumours may be dispersed over several areas of the body. There are no areas of predilection, but most commonly the face, trunk , and extremities are involved. The shape of these lesions may be similar to the smaller localized variety or may consist of erythematous plaques or tumours. New lesions may be developing while older ones disappear without treatment. Recurrences are common. In both types general health is never affected. There is no enlargement of the spleen or lymph nodes and the peripheral blood is normal.

Clinical differential diagnosis should include trichoepithelioma, syringoma, adenoma sebaceum, lichen nitidus, Boeck's sarcoid, lupus miliaris disseminatus faciei, histiocytoma, granuloma faciale, lymphocytic infiltration of the skin, chronic discoid lupus erythematosus, polymorphous light eruption and malignant lymphoma. All of these conditions may be differentiated histologically.

The histopathologic descriptions of lymphocytoma cutis in many available publications show several variants. A heavy polymorphous infiltrate is present in the dermis,

usually separated from the epidermis by a narrow zone of normal collagen. The infiltrate consists of two types of cells namely histiocytes and lymphocytes. The two types of cells lie either intermingled with one another or in a follicular arrangement. In the latter type of arrangement one sees as a rule lymphocytes surrounding islands of histiocytes resulting in structures that resemble the follicles of lymph nodes. Not infrequently nuclear dust can be seen in the follicular centres as a result of disintegration of nuclei. The epidermis is essentially normal or atrophied. It may show occasionally foci of spongiosis and parakeratosis. Skin appendages may or may not be affected. The infiltrate may surround the hair follicles and actual infiltration of the sweat glands may be seen rarely. Using direct and indirect immunofluorescent techniques, the lymphocytes in lymphocytoma cutis were found to be of the T type. This raised the possibility that there is a local immune reaction of the cell mediated type which may play a role in its aetiology.

The histological difficulties arise mainly with malignant lymphomas, lymphocytic infiltrations of the skin of Jessner, the plaque type of polymorphous light eruption, chronic discoid lupus erythematosus and insect bite granuloma. Special care have been taken for the differentiation from malignant lymphomas due to the great resemblance exist.

However, in favor of a benign diagnosis is the presence of epidermal changes, patchy polymorphous cellular infiltrate with germinal centers and polychrome bodies. In addition, the vascular changes are usually prominent. Lesions of malignant lymphoma, on the other hand, have relatively little epidermal changes and have patchy or diffuse infiltrates with a monomorphous picture. Mitotic figures, poorly differentiated cells, and atypical cells may be common. The vessels show relatively few abnormalities.

Basically, lymphocytoma cutis is a benign process although it may occasionally occur in association with malignant tumours. Rarely disseminated nodules may undergo malignancy.

As the exact aetiology is unknown, the treatment is largely non specific. The circumscribed form usually responds rapidly to radiotherapy although recurrence is possible. The effective dose varies from 500-1,500 rads; or the treatment may be given in fractional doses for example 75-100 rads weekly for 5-7 doses. The disseminated form does not respond so readily to X-ray therapy and shows a greater tendency to recur. Good results from the use of procaine penicillin (a total of 2,000,000 units for a child and 12,000,000 units for an adult) have been reported. It is well known now that lymphocytoma cutis may regress or even completely resolves under intralesional, topical

and/or systemic corticosteroid therapy. However, recurrences of the lesions in the previously involved sites may occur after the treatment is stopped especially in the disseminated type.