

A Lady with Facial Indolent Erythematous Infiltrations

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CASE SUMMARY

History

A 54-year-old Chinese housewife complained of two-year history of mildly itchy erythematous patches over the forehead and bilateral temporal areas. The lesions were localized and had increased in size very slowly for two years. Besides mild itchiness, the lesions were asymptomatic and there was no associated sensory disturbance. She had no systemic upset, and denied previous use of cosmetics or medication over the face. There were no remarkable travel history and she recovered uneventfully from hepatitis 10 years ago.

Physical examination

Well-defined erythematous dermal indurations with different sizes were identified over the forehead, bilateral temporal areas, left upper cheek and left infra-auricular region (Figure 1). Apart from a small excoriation over the left temporal lesion, no epidermal changes were found. The lesions were firm in consistence, non-tender and had no sensory loss. Neither regional lymphadenopathy nor hepatosplenomegaly was found on physical examination.

Differential diagnosis

Clinically the diagnosis was most compatible with lymphocytoma cutis, but cutaneous lymphoma, which might sometimes present similarly, should be ruled out. Lupus timidus can be differentiated from lymphocytoma cutis by histology and immunofluorescence. Jessner's disease usually waxes and wanes; and in lymphomatoid contact dermatitis, more epidermal change will be expected. Moreover, the dermal infiltrate in the last two



Figure 1: Multiple infiltrated facial erythematous plaques

conditions mainly consists of T-cells with no lymphoid follicle. Angiolymphoid hyperplasia with eosinophilia is a disease of endothelial cell proliferation. Follicular mucinosis is usually accompanied by alopecia and dermal mucin. In this locality, lepromatous leprosy must be excluded.

Investigations

Complete blood picture and blood biochemistry were normal, except a raised gamma GT level (65U/L, reference range: <35U/L). Anti-nuclear factor was normal and hepatitis B surface antigen and antibody were negative. A 4-mm punch skin biopsy was performed on the left temporal lesion, which showed moderately dense lymphocytic infiltrate centered around blood vessels and skin appendages. Lymphoid follicles with germinal centres were present. There was no destruction of skin appendages. A scattering of plasma cells, histiocytes and occasional giant cells were seen. A grenz zone was detected, and the epidermis was unremarkable.

Diagnosis

The diagnosis was lymphocytoma cutis.

Treatment and progress

She was treated with potent topical steroid. In a latter visit, the lesions were found to be less erythematous and slightly decreased in size. The itchy sensation had subsided.

REVIEW ON LYMPHOCYTOMA CUTIS

Definition

It is a benign infiltrative lymphocytic condition of the skin; some authors regard it as cutaneous B-cell pseudolymphoma (CBPL).¹

Clinical features

The disease can occur at any ages but most commonly below the age of forty and has a female predilection. There are two main types of presentations. The most common type presents as localized, asymptomatic solitary or few erythematous nodules or plaques favoring the face and extremities. Disseminated

involvement was usually found in older patients. The lesions enlarge very slowly. The patients are generally in good health without evidence of systemic involvement. Malignant transformation has been rarely reported.²⁻⁴

Aetiology

The cause of lymphocytoma cutis is unknown in most instances. It has been reported to be associated with infections such as herpes zoster, Lyme disease, gold earrings,⁵ trauma, acupuncture, tattoo, insect bite, medication and vaccination. It is believed to be a reactive hyperplasia of foci of pre-existing lymphoid elements to antigens. However, it has been suggested that chronic antigenic exposure may lead to abnormal immune response and result in malignant transformation.⁶

Differentiation from cutaneous lymphoma

The utmost importance before diagnosing lymphocytoma cutis is to rule out cutaneous lymphoma. Clinically, both conditions can be quite similar, but malignant lymphoma tends to be larger and more widely distributed. In addition, extracutaneous sites involvement is more common in malignant lymphoma. In essence, lymphocytoma cutis is a benign and reactive process, thus polymorphous lymphocytes infiltration, normal looking lymphocytes and formation of germinal centres are useful supporting features. However, at times, differentiation from malignant lymphoma still cannot be readily made from ordinary H&E staining. In this situation, immunophenotypic assay and Southern blot analysis of immunoglobulin are needed (Table 1). When malignant lymphoma is suspected, CT scan and bone marrow aspiration/biopsy should be carried out to look for visceral involvement. Any suspicious lymph node should be biopsied.

Treatment

There is no treatment of proven value for lymphocytoma cutis. Glucocorticoids (topical, intralesional & systemic),⁷ penicillin, antimalarials,⁸ minocycline, cryotherapy⁹ and radiotherapy¹⁰ had been used.

Prognosis

Lymphocytoma cutis usually runs a protracted benign course. Local or distant recurrence can occur following any form of treatment. Since malignant B-

Table 1. Histological and immunophenotypic differentiation between lymphocytoma cutis and cutaneous B-cell lymphoma

	Lymphocytoma cutis	Cutaneous B-Cell Lymphoma (CBCL)
Histological features	<ul style="list-style-type: none"> • "Top heavy" in the dermis • Spares appendageal structures • Polymorphous infiltrate of small lymphocytes • Lymphocytes benign appearance • Mitosis is rare 	<ul style="list-style-type: none"> • "Bottom heavy" extending into the fat • Often involve adnexal structures • Monomorphous infiltrate of large lymphocytes • Abnormal looking lymphocytes • Mitosis is common
Immunophenotypical features	<ul style="list-style-type: none"> • No t(14,18) translocation, bcl-2 proto-oncoprotein¹² • Polyclonal • Polytypic light chain expression (λ / κ) • Compartmentalization: The periphery of lymphoid follicles is composed of T-cells. The germinal centres are populated by polyclonal B-cells. Langerhans' cells are present at the external boundary of the follicles 	<ul style="list-style-type: none"> • t(14,18) translocation and bcl-2 oncoprotein present • Monoclonal • Monotypic light chain expression • Abnormal expression or loss of surface antigens • MT2/CD45RA expression is exclusively restricted to follicular CBCL, whereas germinal centres of CBPL are negative.

cell lymphoma transformation occurred very rarely, no prognostic factors have been concluded so far.¹¹ However, clonal B-cells were more commonly found in the lesions from those patients who had progressed to malignant lymphoma.

Learning points:

Lymphocytoma cutis is diagnosed by its clinicopathological features. Cutaneous lymphoma sometimes causes diagnostic difficulties. Long-term follow-up for these patients is needed.

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