LYMPHOMATOID PAPULOSIS

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Lymphomatoid papulosis is a rare disease affecting young adults Characterized by recurrent papules on the trunks and limbs which heal spontaneously without leaving any effects. Sometimes, the lesion may be larger and undergo necrosis and ulceration. The present case illustrates one such presentation of the disease including diseases competing for differential diagnosis.

CASE REPORT

A 39 years old man presented to the dermatologist at the local Military hospital in March, 1981, with an ulcer on the right leg. For the last five years he had recurrent lesions which appeared as papules on the legs and trunk, on an average of about three lesions per year. These lesions caused itching, became necrotic and ulcerated but healed spontaneously in a couple of months, leaving behind depigmented scars. In January, 1981, he developed afresh lesion on the right leg, which became indurated and healed partially resulting in ulcer formation. The ulcer which was rounded measured 2.5 cms across. Its margins were relatively sharp and the base was covered by necrotic matter. The lesion did not respond to usual treatment and was consequently biopsied for histological examination, carried out at the pathology department, Army Medical College, Rawalpindi in March, 1981. Further clinical examination did not reveal any lymphnode enlargement in the groins or elsewhere in the body.

Histological findings

The biopsy of the ulcer revealed that the epidermis was ulcerated for the most part. There was dense pervascular inflammatory cell infiltrate in the upper dermis, consisting mostly of the lymphocytes, macrophages and plasma cells. There were cells with large irregularly shaped hyperchromatic nuclei. A few mitotic figures were also seen. The final histological diagnosis was arrived at after consultation with professional colleagues at St. Thomas's Hospital, London.

Investigations

The peripheral blood counts, ESR (12 mm/hour wintrobe), blood urea, electrolytes and liver function tests were within normal limits. No hilar or mediastinal lymphnode enlargement was detectable on chest radiography and the lung fields were clear.

DISCUSSION

The term "Lymphomatoid Papulosis" describes a clinically benign but histologically malignant condition[1]. It has been accepted as a clinicopathological entity[2] and over 80 cases had been recorded in the world literature upto 1977. The largest numbers of reported cases[3] is 110. The lesions vaguely resemble clinically those of pityriasis lichenoides at varioliformis. These occur in crops over a period of years, causing itching, necrosis, ulceration, but heal spontaneously in a couple of months leaving behind depigmented varioliform scars. Microscopy may reveal mild invasion of epidermis by lymphoid cells, a picture resembling pityriasis lichenoides. However, most of the lesion in LP are sharply circumscribed with a heavy lymhocytic infiltrate in the upper dermis including a few large cells having hyperchromatic nuclei with irregular shape. Some cells in mitoses are a common feature of the dermal infiltrate. The mitoses may be numerous in the earlier lesion but later a typical cell tends to disappear. A

definite histological diagnosis of lymphomatoid papulosis may not be possible in older lesions especially when superficial necrosis has developed in the overlying epidermis. It is essential to exclude lesions, which may simulate the picture of lymphomatoid papulosis such as Actinic Reticuloid, Arthropod bites and Mycosis fungoides. The actinic reticuloid is observed only in elderly men on the sun exposed areas of the body. The inflammatory infiltrate is deeper, often pleomorphic which may invade the epidermis. The diagnosis is established by hypersensitivity to ultraviolet and visible light. The arthropod bites are usually solitary lesions. On histology, these may resemble lymphoma, mycosis fun goides and lymphomatoid papulosis; the lesion may persist for many months. The biopsy shows massive mixed infiltrate usually containing eosinophils and some atypical mononuclear cells. The infiltrate may be monomorphic. Scabies account for quite a few cases. Mycosis fungoides (M.F.) is a lymphoreticular neoplasm arising in the skin often remaining confined to the skin for many years even throughout life. The epidermis in this condition is acanthotic with long and rounded rete ridges. The papillae are correspondingly prominent and often bulbous. There is usually some intra-cellular and inter-cellular oedema in the epidermis which is often invaded by mononuclear cells which may be single or in groups within small cavities called Pautrier's Microabcesses highly pathognomic of this lesion. Some of these cells have hyperchromatic irregularly shaped nuclei in its classical form, the infiltrate in the dermis spares the dermal papillae. In the early pre-tumour stage cellular infiltrate may form a relatively dense and broad zone in the subpapillary layer and may extend deep in the dermis. The infiltrate consists of a mixture of cells in which the mononuclear cells predominate. However plasma cells, eosinophils and neutrophils may also be present in that order of frequency. Recent work has shown the presence of TCR gene re-arrangement in pityriasis lichenoides varioliformis et acuta (PLVEA) [4]. Thus this disease might represent either an inflammatory dermatosis with clonal T cell expansion or alternatively a benign T cell neoplasm. Similar arguments can be used for other cutaneous diseases such as pagetoid reticulosis and granulomatous slack disease. Genotypic analysis shows that these diseases are also characterized by clonal T cell proliferation[5] though they behave in a benign way clinically. TCR gene re-arrangement findings in lymphomatoid papulosis[6] regressing a typical histocytosis[7] and lymphomatoid granulomatosis[8] pose equally perplexing problems, because T cell monoclonality is not observed in all cases and its presence is not always associated with poor prognosis. In the light of these new insights we might wonder whether or not there are clonal and non-clonal inflammatory dermatoses. We know in the large intestine both benign adenomas and malignant tumours in familial polyposis coli can show clonal 5 q 21 deletion and the spectrum of ulcerative colitis, dysplasia and malignancy is well recognised. It can thus be hypothesized that some skin diseases may evolve through a similar spectrum which is presumably a step-wise sequence of specific molecular/genetic events awaiting elucidation.

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