Peripheral T Cell Lymphoma Presenting with Hypereosinophilia

V Mehta*, S Hameed⁺, C Balachandran[#], A Roy^{**}

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Introduction

Cutaneous lymphomas are a heterogeneous group of neoplastic illnesses that emerge through clonal proliferation of lymphocytes in the skin. They can be classified as cutaneous B cell lymphomas (CBCL) and the cutaneous T cell lymphomas (CTCL). CTCL are an uncommon type of Non Hodgkin's lymphoma and peripheral T cell lymphomas (PTL) are among the rare variants of CTCL. We report a case of peripheral T cell lymphoma in a 50 year old man.

Case Report

A 50 year old previously healthy male presented with generalised pruritus of six months duration not responding to emollients and antihistamines. One month later he noticed a single nodule on his right arm, which was followed by development of multiple nodules on his face, arms, back and chest over a period of next two months. This was associated with significant weight loss of 5-6 kgs and loss of appetite. He gave a short history of fever of about five to seven days and upper respiratory tract infection. There was no history suggestive of scaly erythematous patches or plaques in the past hypopigmented anaesthetic skin lesions, neuritis, urticarial lesions, haemoptysis, abdominal pain or any other systemic complaints.

On general examination mild pallor and generalized lymphadenopathy (cervical, axillary, inguinal) was present. Enlarged para aortic lymphnodes were discovered on an abdominal scan. There were no epitrochlear lymph nodes. Systemic examination revealed hepatosplenomegaly. Cutaneous examination showed multiple, firm, non tender, discrete subcutaneous nodules measuring 2-4 cms over the face, neck, trunk and extremities, associated with generalised xerosis (Figs. 1). Based on the above findings a differential diagnosis of lepromatous leprosy, lymphoma, and pseudolymphoma were considered. Preliminary investigations revealed raised total white blood cell (WBC) counts of 85700/cumm with a massive increase in the absolute

eosinophil count (64270 cells/ cu mm). Peripheral smear revealed marked increase in WBC's and eosinophils (75%) without any abnormal cells.

Slit skin smear for acid fast bacilli (AFB) was negative. Bone marrow aspiration and biopsy showed a reactive marrow with marked eosinophilia. Skin biopsy, both from nodules and normal skin showed a dense infiltrate composed of lymphocytes, histiocytes and eosinophils in the dermis. Epidermotrophism was absent (Figs. 2,3). Axillary lymph node biopsy revealed a polymorphous population of cells with sheets of pale histiocytes, large lymphoid cells with cleaved or jelly-fish nucleus, and immunoblasts along with eosinophils, plasma cells and mature lymphocytes (Fig.4). Mitotic figures and vascular proliferations were also noted. These findings were suggestive of Non Hodgkin's lymphoma i.e. peripheral T cell lymphoma. Immunohistochemistry confirmed it to be CD 3, CD 45 positive, and CD 30 negative (Figs. 5,6). Patient was treated symptomatically and referred to oncology for initiation of appropriate chemotherapy.

Discussion

Our patient was a classical case of disseminated nodal peripheral T cell lymphoma (PTCL) with cutaneous involvement. PTCL is an uncommon variant of CTCL and accounts for 10% of all cases of Non Hodgkin's lymphomas. It occurs in all age groups although peak occurrence is in the sixth and seventh decades. Most PTCLs are aggressive malignant conditions with only cutaneous anaplastic large cell lymphoma and mycosis fungoides displaying an indolent clinical course [1]. The World Health Organization (WHO) and the European Organization for the Research and Treatment of Cancer (EORTC) [2], have proposed the classification for cutaneous T cell lymphomas based on their clinical behaviour (Table 1).

Peripheral T cell lymphoma unspecified is a heterogeneous entity that manifests with sudden onset of

*Assistant Professor, ⁺Postgraduate Student, [#]Professor and Head (Department of Skin & STD), ^{**}Associate Professor (Department of Pathology), Kasturba Medical College, Manipal, Karnataka.

Fig. 1 : Clinical photograph depicting the nodular lesion on the neck

Fig. 2 : Low power photomicrograph (10x) showing a polymorphous cell population

Fig. 3 : High power photomicrograph (40x) showing atypical lymphocytes in the skin

localized or generalised plaques, nodules or tumours. The absence of prior or concurrent patches of mycosis fungoides differentiates this condition from classic mycosis fungoides transforming to diffuse large cell lymphoma. In the past this subgroup used to be called as tumeur d'embry type of mycosis fungoides. Patients are generally human T-cell lymphotropic virus (HTLV 1) negative and prognosis is poor [3]. PTLs

Fig. 4 : Axillary lymphnode biopsy showing large lymphoid cells with sheets of histiocytes, eosinophils and plasma cells

Fig. 5 : Immunohistochemistry showing CD3 positive

Fig. 6 : Immunohistochemistry showing LCA positive

have been further classified based on histology and immunophenotype into low grade, high grade and unclassifiable [4]. According to the EORTC classification our case can be appropriately classified as large cell CD30 negative peripheral T-cell lymphoma unclassified based on the short history of multiple subcutaneous nodules and no concurrent patches of mycosis fungoides. Histopathologically it is characterized

Table 1**EORTC classification for cutaneous T cell lymphoma**

	Frequency (%)	5 year survival (%)
Indolent clinical behaviour		
Mycosis fungoides	44	88
MF variants and subtypes		
Folliculotropic MF	4	80
Pagetoid reticulosis	< 1	100
Granulomatous slack skin	< 1	100
Primary cutaneous CD 30+ lympho-proliferative disorder		
Primary cutaneous anaplastic large cell lymphoma	8	95
Lymphomatoid papulosis	12	100
Subcutaneous panniculitis like T cell lymphoma	1	82
Primary cutaneous CD4 + small/medium size pleomorphic T cell lymphoma	2	75
Aggressive clinical behaviour		
Sezary syndrome	3	24
Adult T cell leukemia/lymphoma	NR	NR
Extranodal NK/T cell lymphoma (nasal)	NR	NR
Primary cutaneous peripheral T cell lymphoma unspecified	2	16
Primary cutaneous aggressive epidermotrophic CD8 + T cell lymphoma	1	18
Cutaneous gamma/delta T cell lymphoma	1	NR

NR - not reported

by nodular or diffuse infiltrate of medium to large pleomorphic T cells and immunoblasts with absence of epidermotrophism. This variant carries a poor prognosis with a five year survival rate of 15%. It must be differentiated from the pleomorphic small to medium CD 30 negative PTL unclassified, which carries a good prognosis with an estimated five year survival rate of

62- 80% [5]. Nonspecific lesions such as generalised pruritus, alopecia, hyperpigmentation, prurigo may be observed in NHL. Hypereosinophilia is also one of the paraneoplastic markers of lymphoma and it is seen in upto 29% of the patients with PTCL [6, 7]. In our patient no specific treatment was directed towards eosinophilia as it was considered as an association with lymphoma. We report this case due to its rare occurrence and its unusual association with such high peripheral eosinophilia.

Conflicts of Interest

None identified

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