Austin Journal of Dermatology

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Special Article - Dermatology Clinical Cases and Images

Primary Cutaneous Diffuse Large B Cell Lymphoma-Leg Type Mimicking Cellulitis in a Patient with Acquired Immune Deficiency Syndrome (AIDS)

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Received: August 30, 2017; **Accepted:** February 13, 2018; **Published:** February 21, 2018

Abstract

The incidence of lymphoma in individuals with Human Immune Deficiency Virus (HIV) infection exceeds that of the general population. The increased risk for lymphoma has been attributed to multiple factors, including the transforming properties of the retrovirus, the immunosuppression and immune dysregulation, opportunistic infections with other lymphotrophic viruses such as Epstein-Barr virus and human herpesvirus 8. So here we report a case of swelling of right thigh in a 40 year old female patient with a retroviral disease which later turned out to be a primary cutaneous diffuse large B cell lymphoma-leg type.

Keywords: Cutaneous B cell lymphoma; Retropositive; Cellulitis

Introduction

HIV-1-associated lymphoma was first incorporated into the U.S. Center for Disease Control and Prevention (CDC) case definition of AIDS in 1985 [1]. Prior to the use of effective Antiretroviral Therapy (ART), HIV-1-associated lymphomas constituted 3 to 4% of all AIDS-defining illnesses reported to the CDC. Following the advent of effective prophylactic regimens for common HIV-1-associated opportunistic infections (ex, *Pneumocystis jiroveci*, *Toxoplasma gondii*, Cytomegalovirus) and the development of antiretroviral monotherapies, the incidence of HIV-1-associated lymphomas remained constant at 1.6% per year [2]. Without effective antiretroviral therapy, it is estimated that 5-10% of all HIV-infected individuals will have lymphomas as either an initial or subsequent AIDS-defining condition [3].

Case Presentation

A 40 year old female who was seropositive for HIV presented with history of swelling of the right thigh on and off since 5 months following which she developed red raised lesions over the right thigh. Initially localized, later gradually spread to involve the whole of the right thigh. There was history of ever, on and off with pain at the site of the lesion for 5 months. Patient was diagnosed to be retrovirus positive 2 years ago and started on Antiretroviral Therapy (ART) two months ago. The patient was not on ART initially because of her negligence. On examination, Multiple discrete to coalesced erythematous nodules and plaques with central area of oozing and crusting was present over the right thigh (Figure 1,2). On palpation, the lesions were firm, indurated and tender. Right inguinal lymphadenopathy was present. Systemic examination was normal. Blood investigations revealed anemia with haemoglobin 10g/dl and the CD4 count of 50 cells/mm^3. Histopathology, Skin with dermal discrete monomorphic tumor cells showing vesicular nucleus and prominent nucleoli. Altered dense collagen seen. Other cells like eosinophils and plasma cells were also seen (Figure 3,4). Histopathology favoured cutaneous B cell lymphoma, IHC markers was positive for CD20 (Figure 5). The other IHC markers were not advised as the patient was not affordable for investigations. Positron emitted tomography did not show any foci of malignancy anywhere else.

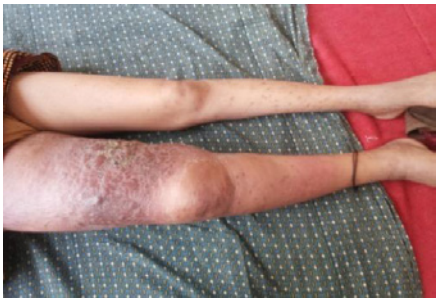

Figure 1

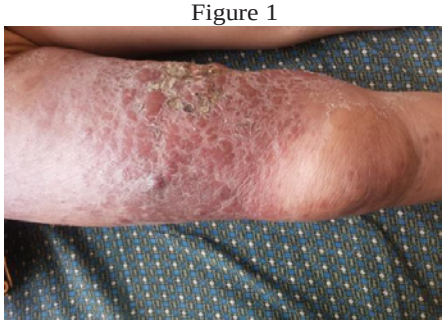

Figure 2

Figure 1,2: Multiple discrete to coalesced erythematous nodules and plaques with central area of oozing and crusting over right thigh.

The patient was started on ART, later referred to a regional cancer institute for further management of the tumor. Unfortunately the patient succumbed to death within 3 days of discharge from our hospital.

Discussion

India is one of the countries with the highest HIV burden. In the developed countries, 34% of AIDS patients suffer from cancer,

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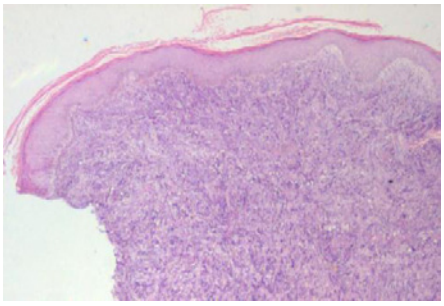

Figure 3: Shows skin with near normal epidermis. Dermis shows dense, diffuse, discrete tumour cells.

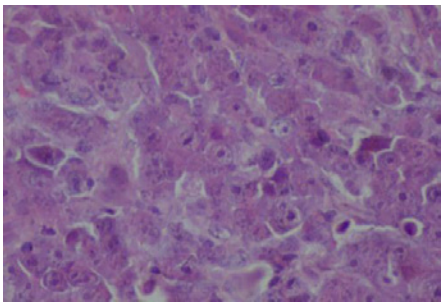

Figure 4: High power magnification - tumor cells with vesicular nuclei, prominent nucleoli with increased mitosis.

while this incidence is only 3%-4% in the Indian population [4]. While incidence rates of NHL (non-Hodgkin lymphoma) in India are lower than Australia and Western countries such as Canada and United States (10-17 per 100,000 populations), it is one of the common cancers in urban populations in India. Among the urban Population-Based Cancer Registries (PBCRs) in India, Delhi has the highest rates of 5.1 per 100,000 persons per year. This is followed by Mumbai, Chennai, Bhopal and Bangalore, in the decreasing order [5]. They have been less likely to respond to chemotherapy, with shorter overall survival. Reported predictors of poor survival include low CD4 count at diagnosis, having a prior ADI (AIDS defining illness), Central nervous system presentation, other extra nodal presentations, advanced stage of NHL, high Lactate Dehydrogenase (LDH) levels and older age [6,7].

Among the cutaneous lymphomas 85% cases comprise of cutaneous T-cell lymphomas and the remaining 15% cases are of B-cell origin [8]. Cutaneous B-cell Lymphomas (CBCL) are a specific group of NHL that has been considered as an advanced stage of the neoplasm by both the European Organization for Research and Treatment of Cancer (EORTC) and the World Health Organization (WHO) classifications [9]. CBCL is uncommon and includes a wide range of clinical manifestations, histopathology, immuno phenotype and prognosis. Skin involvement includes single or multiple infiltrative macules, papules, subcutaneous nodules, ulcerative lesions and infiltrative dermo hypodermic plaques. The number of lesions, single versus multiple, is a prognostic factor in primary CBCL [10]. Patients with NHL showed cutaneous compromise in 15-20% of cases, and in 5% of these, skin lesions are the first manifestation of the neoplasm, as in our case [11].

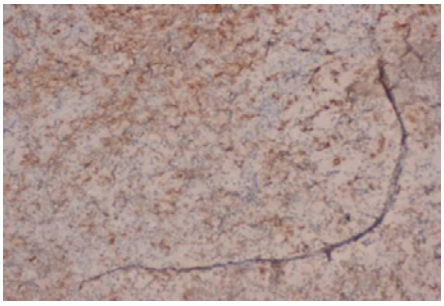

Figure 5: IHC-CD20: Most of the tumor cells show membrane positivity with CD-20.

AIDS associated B-cell lymphomas are commonly described to have atypical morphology, extra nodal involvement as primary manifestation and an aggressive clinical course. The majority of HIV-related lymphomas are Diffuse Large B-Cell Lymphomas (DLBCL). DLBCL are a heterogeneous group of NHL with distinct clinico pathological entities, including B-cell lymphomas with plasmablastic differentiation. The differential diagnosis between these histopathological subtypes is made by the immuno histochemical examination with monoclonal antibodies directed against CD20, CD45, CD10, CD138, plasma cell clone, VS38c, BCL6, mib 1, kappa and lambda light chains and cyclin D1 [12].

The pathogenesis of HIV associated lymphoma is the presence of clonal macrophages harboring HIV integrated in a single site in polyclonal B-cell lymphoproliferative states or lymphomas, that these clonal macrophages play a central initiating role in early lymphoma genesis wherein other immunologic factors play secondary and tertiary roles leading ultimately to lymphoma. Many molecular and virologic events occur that appear to be critical for lymphoma genesis; however, HIV might contribute directly to this process by driving early stages of lymphoma genesis through expansion of clonal macrophages and antigen-driven B-cell proliferation. Loss of B-cell maturational control has been demonstrated by studies of immunoglobulins associated with AIDS-related lymphoma. The randomly mutated nature of immunoglobulin variable region genes in lymphoma suggests that lymphomas may be outgrowths of B cells responding to any antigen. The overall immunodeficiency induced by HIV infection also plays a likely role, supported by recent and encouraging observations of decreased frequency of lymphomas in patients treated with ART. Most AIDS-related lymphomas appear to be outgrowths of antigen-driven B cells with growth control influenced by abnormal T-cell and antigen-presenting cell processes [13].

In addition to reducing the overall risk of lymphoma, ART has had other effects on the epidemiologic characteristics of HIV-related lymphoma. A study linking the San Diego County Cancer Registry data with the San Diego County AIDS registry showed that the incidence of highly aggressive B-cell lymphomas such as immunoblastic Diffuse Large B-Cell Lymphoma (DLBCL) was reduced from 38% of HIV-associated non-Hodgkin lymphomas cases in the pre-ART era to 19% in the post-HAART era [14].

This case has been reported for the rarity of its occurrence, the mode of presentation mimicking cellulitis and to emphasizes the need for early initiation of ART.

References

1. Harnly ME, Swan SH, Holly EA, Kelter A, Padian N. Temporal trends in the incidence of non-Hodgkin's lymphoma and selected malignancies in a population with a high incidence of Acquired Immunodeficiency Syndrome (AIDS). Am J Epidemiol. 1988; 128: 261-267.
2. Moore RD, Kessler H, Richman DD, Flexner C, Chaisson RE. Non-Hodgkin's lymphoma in patients with advanced HIV infection treated with zidovudine. JAMA. 1991; 265: 2208-2211.
3. Hamilton-Dutoit SJ, Pallesen G, Franzmann MB, Karkov J, Black F, Skinhoj P, et al. AIDS-related lymphoma. Histopathology, immunophenotype, and association with Epstein-Barr virus as demonstrated by in situ nucleic acid hybridization. Am J Pathol. 1991; 138: 149-163.
4. Chitale AR. Cancer and AIDS. Indian J Pathol Microbiol. 2005; 48: 151-160.
5. A Project of the National Cancer Registry Programme (Indian Council of Medical Research). 2006.
6. Little RF, Gutierrez M, Jaffe ES, Pau A, Horne M, Wilson W. HIV-associated non-Hodgkin lymphoma: Incidence, presentation and prognosis. JAMA. 2001; 285: 1880-1885.
7. Tirelli U, Errante D, Spina M, Vaccher E, Serraino D, Boiocchi M, et al. Long-term survival of patients with HIV-related systemic non-Hodgkin lymphomas. Hematol Oncol. 1996; 14: 7-15.
8. Salama S. Primary cutaneous B. cell lymphoma and lymphoproliferative disorder of skin: Current status of pathology and classification. Am J Clin Pathol. 2000; 114: 104-128.
9. Swerdlow SH, Campo E, Pileri SA, Harris NL, Stein H, Siebert R, et al. The 2016 revision of the World Health Organization classification of lymphoid neoplasms. Blood. 2016; 127: 2375-2390.
10. Grange F, Bekkenk MW, Weschler J, Meijer CJ, Cerroni L, Bernengo M, et al. Prognostic factors in primary cutaneous large B-cell lymphomas: A European multicenter study. J Clin Oncol. 2001; 19: 3602-3610.
11. Villafañe MF, Marcelo C. Primary cutaneous B-cell lymphoma successfully treated with highly active antiretroviral therapy alone: a case report and review of literature. Indian J Dermatol. 2011; 56: 418-420.
12. De Paepe P, De Wolf-Peeters C. Diffuse large B-cell lymphoma: A heterogeneous group of non-Hodgkin lymphomas comprising several distinct clinicopathological entities. Leukemia. 2007; 21: 37-43.
13. Valerie L, Michael S. Pathogenesis of HIV associated lymphoma. Comprehensive, up-to-date on HIV/AIDS treatment, prevention, and policy from the University of California San Francisco. November 2002.
14. Diamond C, Taylor T, Aboumrad T, Anton-Culver H. Changes in acquired immunodeficiency syndrome-related non-Hodgkin lymphoma in the era of highly active antiretroviral therapy. Cancer. 2006; 106: 128-135.

Austin J Dermatolog - Volume 5 Issue 1 - 2018
ISSN : 2381-9197 | www.austinpublishinggroup.com

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