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Primary Cutaneous Anaplastic Large Cell Lymphoma

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Abstract: Primary cutaneous Anaplastic large cell lymphoma (ALCL) is a rare type of cutaneous T-cell lymphoma. Here we are reporting a case of a 39 year old lady presented with an ulcerated nodular lesion in the right anterior axillary fold and another nodular lesion at the back.
Keywords: Anaplastic large cell lymphoma (ALCL), T-cell

INTRODUCTION

Primary cutaneous Anaplastic large cell lymphoma (ALCL) is a rare type of cutaneous T-cell lymphoma which is characterized by solitary or localized nodules or plaques. This entity should also be considered in the differential diagnosis of erythematous nodular/ ulcerative skin lesions.

CASE REPORT

A 39 year old lady presented with an ulcerated nodular lesion in the right anterior axillary fold (Fig. 1). She had another nodular lesion at the back (Fig. 2). There was no itching and no history of fever or weight loss. Her serum LDH was 163 U/L. A biopsy from the lesion showed dense infiltrate of immature large lymphoid cells which were positive for CD3 and CD30 and negative for ALK and was diagnosed as primary cutaneous anaplastic large cell lymphoma. She received chemotherapy with cyclophosphamide, vincristine, doxorubicin and prednisolone followed by local irradiation and achieved complete remission. However she relapsed 6 months later by similar nodular lesions and progressed thereafter.

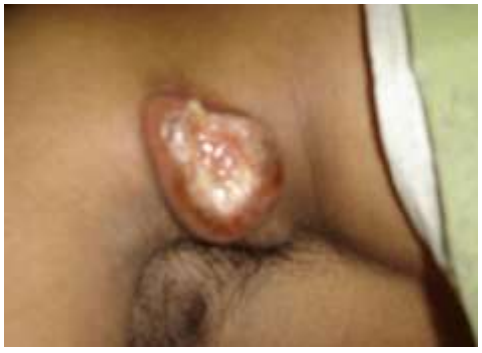

Fig. 1: Nodular lesion in the right anterior axillary fold

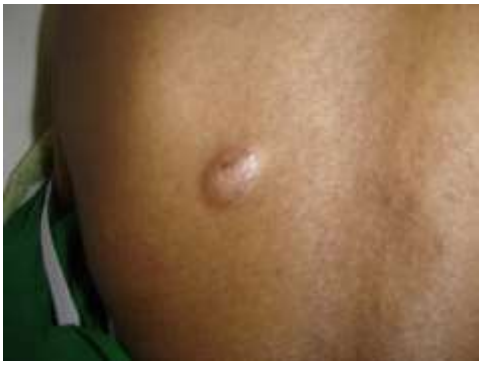

Fig. 2: Nodular lesion at the back

DISCUSSION

Anaplastic large cell lymphoma (ALCL) first recognized in 1985, is a biologic and clinically heterogenous subtype of T-cell lymphoma characterized by expression of the CD30 antigen [1]. Clinically ALCL may present as 3 distinct entities - primary cutaneous ALCL which is localized cutaneous disease or as widespread systemic disease- primary systemic anaplastic lymphoma kinase (ALK)-positive ALCL, primary systemic ALK-negative ALCL [2].

Primary cutaneous ALCL is an indolent cutaneous CD30-positive T-cell lymphoproliferative disorder, affecting mainly adults with a male preponderance. It usually presents as solitary or localized tumor nodules, often with ulceration. About 20% of cases are multifocal. Regional lymph nodes may be secondarily involved in 5-10% cases.

Histopathological findings include a diffuse, non-epidermotropic infiltrate with cohesive sheets of large CD30+ tumor cells. The differential diagnosis in skin biopsies includes lymphomatoid papulosis , transformed mycosis fungoides, and secondary skin involvement by systemic ALCL. Immunophenotypic and immunohistochemical (IHC) studies are critical in the definitive diagnosis of ALCL. Major immunophenotypic features of ALCL include $CD30^{+}$, $CD15^{-}$, PAX-5^{-}, and $CD45^{+}$. ALK protein is detected in most cases of systemic ALCL by IHC. Primary cutaneous-ALCL is typically ALK negative [3].

Primary cutaneous ALCL has a favorable prognosis, with a five-year survival of 95 percent, compared with 79.8 percent for ALK+ systemic ALCL and 32.9 percent for ALK− systemic ALCL [4]. Patients with multifocal disease require systemic chemotherapy with either a single-agent methotrexate or a combination regimen, the most commonly used one being CHOP. Complete remission rates with combination chemotherapy are around 90%, but two thirds of these patients relapse in a few months.

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