Mycosis Fungoides: Tumor D'emblee – A Rare Presentation: A Case Report

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ABSTRACT

Mycosis Fungoides (MF) is a condition characterized by infiltration of skin by neoplastic T lymphocytes. The disease has various stages viz. premycotic, patch, plaques, nodules, tumors and erythroderma. 'Tumor d'emblee' is a variant of tumor stage that develops from an apparently normal skin without prior patch or plaque stage. We present Case of a 70 year old lady with a reddish nodule over her right leg without prior plaques or itching. Fine Needle Aspiration Cytology, Histopathology and Immunohistochemistry studies were done to diagnose it as mycosis fungoides. Mycosis fungoides is the most common subtype of cutaneous T cell lymphoma. Tumors are the initial manifestation in 10% of patients.

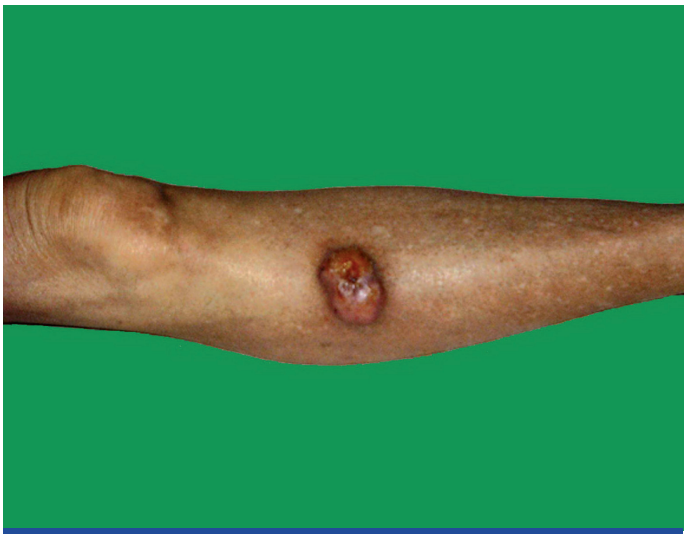
Key Words: Mycosis Fungoides, Tumor d'emblee

INTRODUCTION

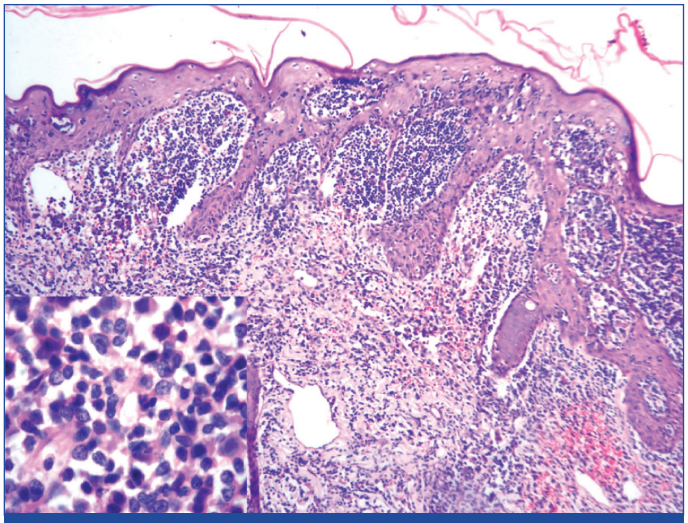
Mycosis Fungoides (MF) is a condition which is characterized by the infiltration of the skin with plaques and nodules which are composed of T- lymphocytes. The disease is extremely variable in its clinical course and presentation. The disease has various stages, viz premycotic, patch, plaques, nodules, tumours and erythroderma. "Tumour d'emblee" is a variant of the tumour stage that develops from apparently normal skin without the prior patch or the plaque stage [1,2,3]. Tumours are the initial manifestation in approximately 10% of the patients [4].

CASE REPORT

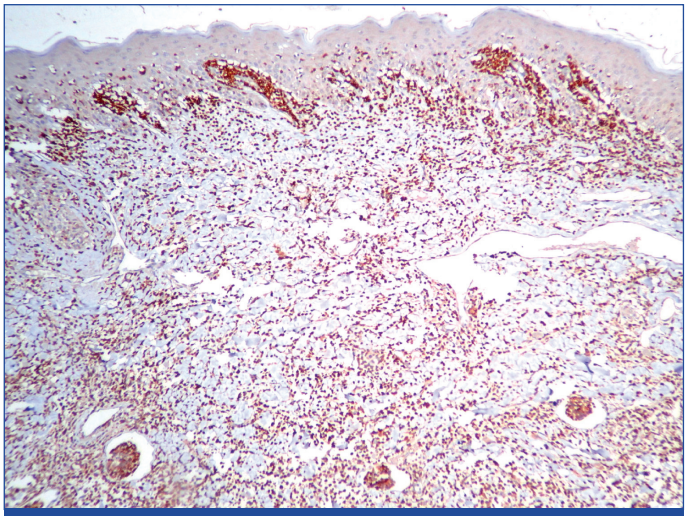
A 70 year old lady presented with a nodule over the extensor surface of the right leg, which was gradually increasing in size since last three months. There was no history which was suggestive of any preceding skin changes or dermatitis. Her past medical history was unremarkable. There was no history of anorexia, weight loss, fever or night sweats. On examination, the nodule measured 3 centimetres in diameter and it was erythematous with ulceration [Table/Fig-1]. There was neither induration nor discolouration in the surrounding skin [Table/Fig-2] . There was no significant lymphadenopathy. Her systemic examination was unremarkable and no organomegaly was noted. Fine needle aspiration which was done from the nodule, revealed singly dispersed, round cells with scanty cytoplasm and an irregular nucleus, which was suggestive of cutaneous lymphoma. Her chest X-ray, ultrasound of the abdomen, peripheral smears and bone marrow studies were within normal limits. Her histological examination showed a dense infiltrate throughout the dermis and in the subcutis forming nodules [Table/Fig-3]. The epidermis showed focal epidermotrophism. The infiltrate showed small to medium sized cells with cerebriform nuclei. Based on the clinical and histological findings, a diagnosis of Cutaneus T Cell Lymphoma (mycosis fungoides – d'emblee type) was made. Immunohistochemistry revealed tumour cells which were positive for the leucocyte common antigen and CD3 [Table/Fig-3] and negative for CD20 and CD30, thus confirming the diagnosis of Cutaneus T Cell Lymphoma. As Merkel cell carcinoma was a histological differential



[Table/Fig-1]: Erythematous ulcerated nodule on the extensor aspect of the leg



[Table/Fig-2]: Microphotograph showing dense infiltrate throughout the dermis (H&E x100),Inset shows cells with cerebriform nuclei (H&Ex1000)



[Table/Fig-3]: Microphotograph showing CD3 positivity in tumor cells (IHCx100)

diagnosis, the markers for synaptophysin and chromogranin were also done and both were negative. The patient refused chemotherapy and was lost to follow up. However, an enquiry revealed that the patient did not have any systemic symptoms and she died after 4 months due to an unrelated illness. During these four months, the patient was neither hospitalized nor investigated.

DISCUSSION

MF is relatively rare, accounting for 1% of all the Non-Hodgkin's lymphomas [4]. However, it represents the most common type of cutaneous lymphoma. The natural history of MF is one of slow progression, with patches and plaques giving rise over time to nodules that may eventually ulcerate. Histology reveals the predominance of small pleomorphic (cerebriform) cells. MF is often indistinguishable from pseudolymphoma, in that, cerebriform nuclei in the dermal infiltrate and pautrier microabscesses in the epidermis are frequently seen. However, a history of drug intake and the disappearance of the infiltrate on the withdrawal of the drug will help to differentiate it from MF. Malignant T cells of the Sezary syndrome have phenotypic and antigenic overlaps with those of MF. If the peripheral blood does not reveal abnormal cells, buffy coat preparation and flow cytometry are helpful to establish the diagnosis of Sezary syndrome. In 1885, Vidal and Brocq described the "tumor d'emblee" variant in which nodules are seen from the outset[4] .This presentation is not well characterized and some of these patients may have other forms of T cell malignancies. [1,5] Nodular MF that has undergone large cell transformation, has to be differentiated from unclassified peripheral T cell lymphoma. . These tumours also present as nodules without the pre-existing plaques or patches of MF. On histology, prominent, nodular or diffuse infiltrates of medium to large pleomorphic T cells and immunoblasts are seen. But epidermotropism is usually absent. Primary cutaneous anaplastic CD 30+ large cell lymphomas clinically present with ulcerated nodules, mostly on the trunk and can resemble nodular MF, but on histology, these does not show epidermotropism and there is a dense infiltrate of large anaplastic T cells with the $CD30^+$ antigen and frequent mitosis. These patients generally have systemic involvement at the time of the diagnosis. As this patient did not have systemic or extracutaneous spread and the absence of large cell transformation at the time of the initial presentation, unspecified, primary, cutaneous T cell lymphomas and primary, cutaneous anaplastic CD30+ large cell lymphomas were not considered as the diagnosis.

The standard treatment for the early lesions includes psoralensin association with UV-A (PUVA), irradiation and interferons or a combination of these three modalities. In the late stages, in addition to PUVA, retinoids, interferons, conventional chemotherapy, extracorporeal photopheresis and radiotherapy have been used [2,4].

Classical cutaneous phases of mycosis fungoides may frequently overlap or occur simultaneously. The prognosis is directly related to the extent of the disease progression, with more advanced lesions like cutaneous nodules and extracutaneous dissemination, thus showing an aggressive course. Since there was no hospitalization at the time of death, the course of the disease in this patient was not documented. If only tumours are present without prior or co-existing patches or plaques, the diagnosis of MF should be questioned. A detailed systemic examination and immunohistochemistry work up to rule out high grade cutaneous pleomorphic lymphoma or other variants of T cell lymphoma has to be carried out.

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