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Case Report

Mycosis fungoides: a rare case report

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ABSTRACT

Lymphoproliferative disease has many sub-types but the most-commonest subtype of cutaneous T-cell lymphoma is mycosis fungoides (MF). When it presents as solitary or oligo-lesional lesion, radiation therapy is considered potentially curative. This paper presents a case of 17-year-old female who developed two skin lesions in her left knee. She was diagnosed as a case of MF in patch stage. Localised radiation therapy was given to the lesion. MF prognosis depends overall on its stage and is usually better in the early stages. MF is rare and often requires a clinical-pathological correlation for its diagnosis, which is often delayed, especially in the early forms.

Keywords: MF, Patch stage, Oligolesional lesion

INTRODUCTION

Mycosis fungoides (MF) is a rare type of primary cutaneous lymphomas which comprises of a heterogeneous group of non-Hodgkin lymphomas that originates from skin-tropic memory T lymphocytes.[1] It generally presents as patches and plaques characterized by an indolent clinical course, with slow or no disease progression. Only rarely does extracutaneous involvement occur with a correspondingly poor prognosis.[2] MF is known to affect people between the age of 55 and 60 years, with a male predominance (male: females, 2:1). While its occurrence is described in children and adolescents, MF is rare in this age group.[3-4]

Management of MF is based on staging. Early-stage MF (IA-IIA) are treated by skin directed therapies (SDTs) including topical corticosteroids, retinoids, phototherapy and radiotherapy (localised or total beam electron therapy). Refractory MF or advanced stage (IIB-IVB) often warrant systemic treatments in combination with SDTs.[5] In clinical settings, especially in the initial stages, MF can be difficult to differentiate from inflammatory skin diseases such as eczema and psoriasis. Because of the small number of neoplastic T-lymphocytes in the infiltrate, histopathological examination is often diagnostically not reliable in early stages, which is why clinico-pathological correlation is crucial for making a correct diagnosis.[2]

CASE REPORT

A 17-year-old female attended in our OPD in the department of radiation oncology, regional institute of medical sciences, Imphal on August 2023, with complaints of two skin lesions over the left knee for the last one week. On examination, the lesions were mildly erythematous, papulo-nodular, 2 in number measuring 1×1 cm^2 and 1×0.5 cm^2 each on the medial aspect of left knee. The lesion was firm, non-tender, non- ulcerative and there was no local rise of temperature over the lesion and the surrounding region. On taking further history, it was found that she was referred to us from the department of dermatology for the two new skin lesions.

The patient gave history of itchy scaly hypopigmented macules of varying sizes over the bilateral buttock, knee and forearm for the past 2 years. There was gradual increase in the size of the hypopigmented macules with decreased intensity of hot and cold sensation over the

lesion. A punch biopsy of the lesion from the buttock region was done, which yielded an ortho-keratotic epidermis with mild lymphocytic infiltration in a pericapillary and perineural distribution in the dermis. No granulomas were identified and Wade Fite Faraco Stain for Lepra Bacilli was negative with features suggestive of but inconclusive for indeterminate Hansen's disease. She received treatment with PB-MDT (Pauci-bacillary multi-drug treatment) for 6 months. On April of 2023, she complained of increasing size of the old lesions post RFT (Released from treatment), with no new lesions, no tenderness or altered sensations over the lesion and neighbouring region.

A punch biopsy was taken again from the lesions in the buttock and left knee. Histopathological examination (HPE) from the specimen show skin with orthokeratotic epidermis. The dermis showed moderate lymphocytic infiltration in a pericapillary, peri-adnexal and perineural distribution. A few scattered neutrophils and occasional eosinophils were identified and with scattered atypical mononuclear cells. No granulomas or evidence of epidermotropism were seen. ZN (Ziehl-Neelsen) stain revealed no acid-fast bacilli. Mantoux test came negative. On Immunohistochemistry (IHC) study, cells were positive for CD2, CD3, CD4, CD20 and negative for CD5, CD7, CD8, ALK-1, TdT, EBERish, PAX-5 and Granzyme-B. Ki-67 was 60%. These features were suggestive of lymphoproliferative disease and treated in that line from May 2023. Clinically, the lesions correlated with that of MF (patch stage) with multiple well to ill-defined hypopigmented macules and patch of different sizes showing mild atrophy and was staged at $T_1N_0M_0B_0$ (Stage IA) with <10% BSA.

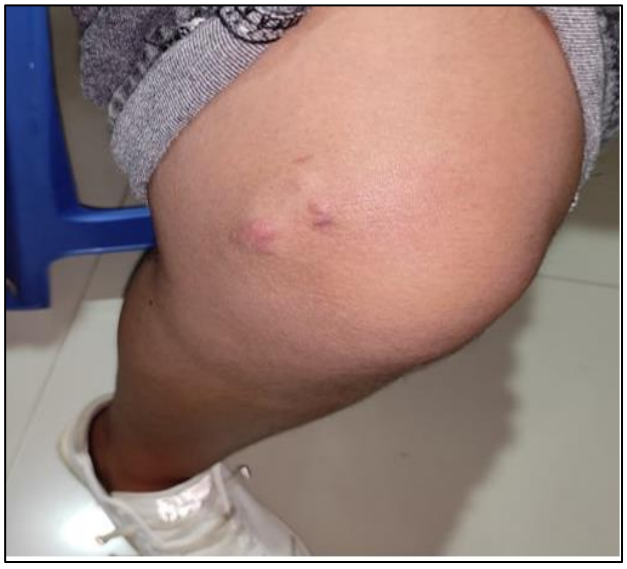

Figure 1: Skin lesion over the left knee before treatment.

Her treatment started with PUVA (psoralen plus ultraviolet-A radiation), topical steroids, topical retinoids. On August 2023, she complained of 2 new lesions over her left knee and she was referred to our department. The old lesion showed decreased erythema with normal pigmentation over the bilateral buttock, knee and complete re-pigmentation over the bilateral forearm. Baseline investigations (CBC, LFT, KFT, SE, RBS, chest X-ray, USG-W/A) was done and were found to be within normal limits. A localised external beam radiation therapy (EBRT) was given to the left knee lesion at a surface dose with Cobalt-60 teletherapy machine, to a total dose of 30Gy in 15#, 5 days a week over 3 weeks by enface technique. Margins of 2 cm was taken around the lesion and the area of visible disease was bolus with 0.5 cm tissue-equivalent material. During the course of EBRT, the patient also continued her topical retinoids and topical steroids. The present case had a very good response to localised superficial radiation therapy. During her monthly follow up, the lesions were noted to have decreased in size with a mild hypopigmented patch over the region.

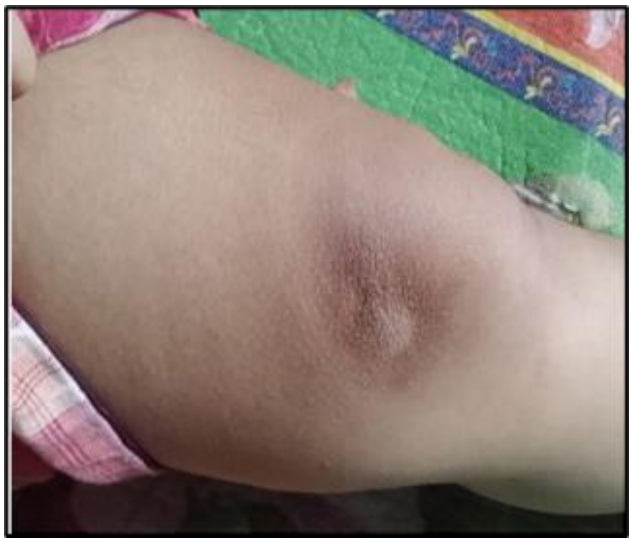

Figure 2: Skin lesion over the left knee after treatment.

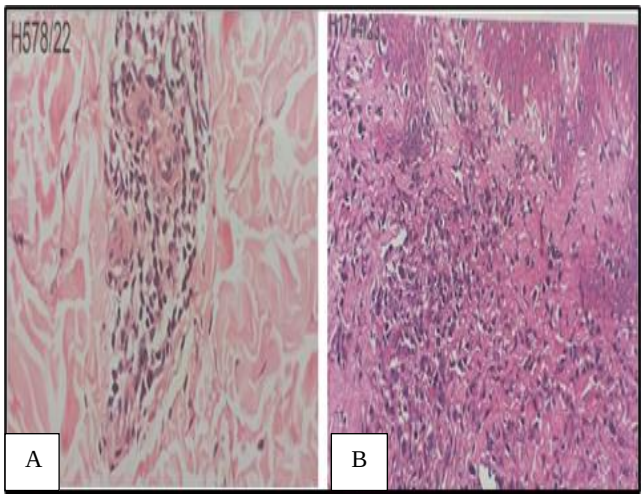

Figure 3 (A and B): Dermis of mild lymphocytic infiltration.

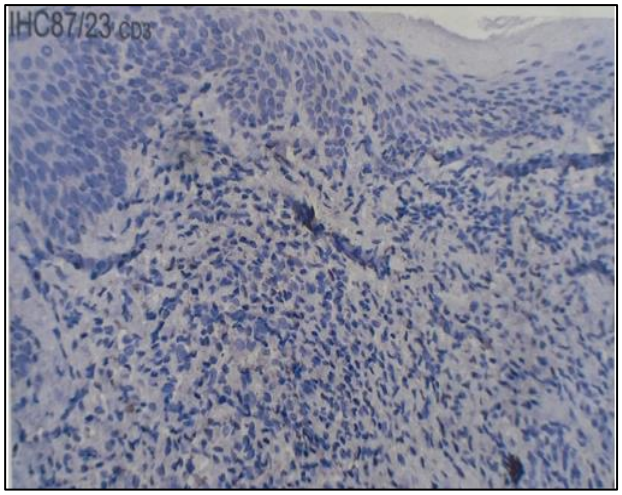

Figure 4: IHC of CD3.

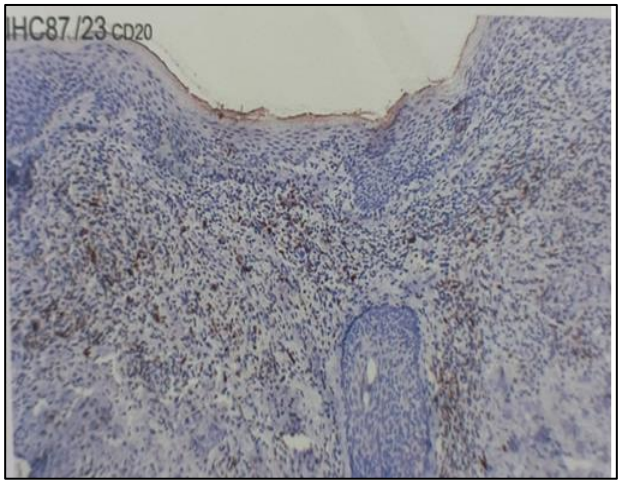

Figure 5: IHC of CD20.

DISCUSSION

Primary cutaneous lymphomas are a heterogenous group of extra nodal non-Hodgkin lymphoma which, by definition, are confined to the skin at the time of diagnosis. Primary cutaneous lymphomas are classified by the world health organisation-European organisation for cancer research and treatment (WHO-EORTC) into cutaneous T-cell lymphomas (CTCL) and cutaneous B-cell lymphomas (CBCL).[3] Contrary to nodal non-Hodgkin lymphoma, most of which are B-cell derived, approximately 75% of primary cutaneous lymphomas are T-cell derived, of which 2/3rd may be classified as MF or Sezary syndrome, which is a rare leukemic variant of CTCL.[6]

Indolent lymphomas usually have a chronic course with frequent recurrences with the disease generally considering as incurable despite treatment, with a mean survival of 5 to 10 years.[3] MF often shows diagnostic difficulties due to its absence of specific features and lesional polymorphism, may require multiple biopsies and always require clinic-pathologic correlation and relatively non-specific in early stages. The WHO-EORTC recognises three distinct variants of MF namely folliculotropic MF, pagetoid reticulosis and granulomatous slack skin.[7] Patients affected by MF have long pathological stories with possible phases of remission and progression,[7] similar to our present case, and so treatment require multidisciplinary team approach to ensure both correct diagnosis and the best treatment choices.

Therapy can vary from skin-directed therapy (SDTs), localised radiation therapy (RT), total skin RT, systemic therapies, or chemotherapy, and in some cases associated with poor prognosis, allogenic stem cell transplantation may be recommended. Localised, superficial radiation therapy is a highly effective treatment approach for eradication of unilesional disease. Palliation of multisite disease by total skin electron beam therapy (TSEBT) is also recommended which often leads to long-term disease-free intervals.[8] Photons as well as electron beam have been used and doses have range from 0.7 to 35 Gy and may be fractionated.[9-11] Field and dose guidelines from the international lymphoma radiation oncology group recommended a dose range of 24-30 Gy for uni-lesional MF and pagetoid reticulosis. As majority of patients have good respond to lower radiation doses, 20-24 Gy is encouraged.[8] Given the "curative" nature of this treatment, a margin of ≥2 cm is generally taken.[12,13]

Early identification of favourable patients is needed to give timely treatment with RT and sparing the patients from exposing to toxic medications. At present RT response to the goal of curing without overtreating the minimal stage MF, affording durable results with less side effects and maintaining good quality of life. The patient is currently under monthly follow up. Till date, no new lesions were reported in other parts of the body.

CONCLUSION

Radiation therapy, for primary cutaneous lymphomas especially MF, when presented as a solitary or oligolesional disease, is considered potentially curative. Electron therapy, lower energy X-rays, TSEBT etc, are indicated depending on the type and extend of the skin lesions. Histopathologic classification and clinical course of primary cutaneous lymphomas must be taken into account when determining the overall treatment strategy. It can be noted that radiation therapy to skin lesions plays a major role in the treatment of these lymphomas pertaining to achievement of excellent local control.

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