

Case Blog

Title: Mycosis Fungoides Tumour

Mat Samuji M.S* and Gilmore J

Department of Radiation Oncology, Cork University Hospital (CUH), Cork, Ireland

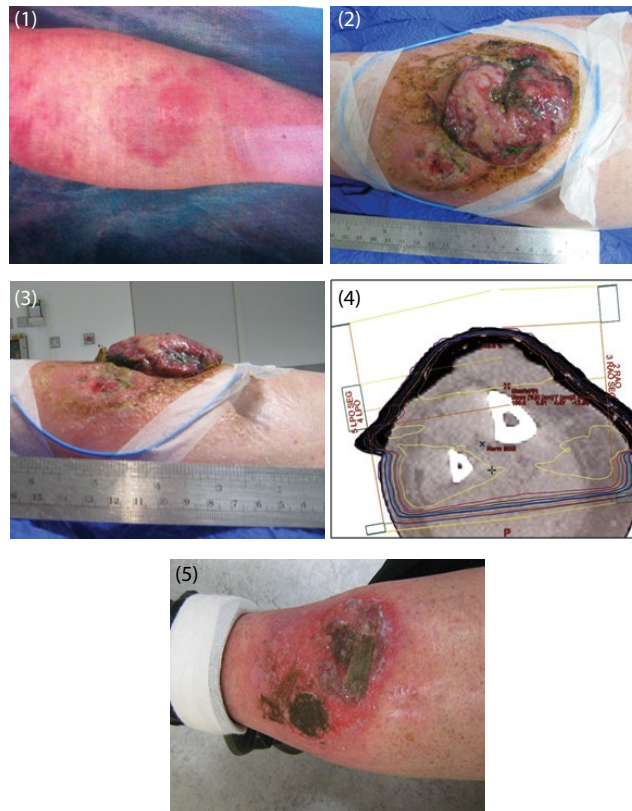


Figure 1: Initial plaque at left shin.
Figure 2: Tumour at left shin (anterior view).
Figure 3: Tumour at left shin (lateral view).
Figure 4: Radiation therapy fields.
Figure 5: Early result (2 weeks post-radiotherapy).

Introduction

Cutaneous lymphomas are a group of disorders characterised by localisation of malignant lymphocytes to the skin. The most common subtypes of primary cutaneous T-cell lymphomas are mycosis fungoides (MF) and Sezary syndrome (SS). We present a case of rapidly progressive mycosis fungoides who initially presented with erythematous plaques.

Keywords

Cutaneous T-cell lymphoma; Mycosis fungoides (MF); Sezary syndrome (SS)

Case Presentation

A 53 year old lady presented initially with 4 months history of polymorphic patch over her right flank. It became raised and tender lump developed within few weeks which were initially considered as an abscess. An excision biopsy was performed and diagnosis of primary cutaneous T-cell lymphoma or T-cell lymphoma of unspecified origin was made. Immunohistochemistry was positive for CLA and CD3 and negative for CD20, cytokeratins AE1/3 and CK 8/18. S100, HMB45 and CD30 were also negative. A staging CT scan was normal. No weight loss, night sweats or fever was noted in the history and past medical history

*Corresponding author: Mat Samuji M.S, Department of Radiation Oncology, Cork University Hospital (CUH), Cork, Ireland; E-mail: mohd.samuji@hse.ie

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was unremarkable. 2 months after the excision, two small erythematous patches have developed adjacent to the scar and it was treated with radiotherapy (40 Gy in 20 fractions). She remained well afterwards. On follow-up visit, a plaque was noted on her left shin and was kept under review (Figure 1). It progressed into a large ulcerating tumour within 4 months and a diagnostic biopsy confirmed a diagnosis of mycosis fungoides (stage IIB, T3N0M0) (Figures 2 and 3). Further examination revealed a small plaque at her left chest wall. A restaging CT scan was normal. Topical antibiotic was prescribed as it was malodorous. She was referred to radiation oncology and received 50 Gy in 25 fractions to tumour in left shin using 6 MV photon and 40 Gy in 20 fractions to left chest wall using 6 MeV electron (Figure 4). She is due further follow-up in medical oncology clinic for consideration of systemic therapy (Figure 5).

Discussion

Primary cutaneous lymphomas are composed of both T-cell (75%) and B-cell lymphomas and are rare conditions representing 2% of all lymphomas with an annual incidence of 0.3 to 1 per 100 000 [1,2]. MF was first described by Alibert in 1806 as a mushroom like neoplastic skin condition [3]. It most often presents in those aged 45 to 60 years but has been diagnosed in children and adolescents. It is 50% more common in black than in white patients and twice as frequent in men as in women [4]. The onset of MF is often insidious and initial cutaneous symptoms may be difficult to distinguish from other non-malignant pathologies of the skin [5]. The management of MF/SS is centered on a stage-base approach, and MF is classified into 4 clinical stages based on the TNM classification, which then is synthesized into a clinically based staging system broadly divided into early and advanced-stage disease [6]. The majority of patients have indolent disease and given the incurable nature of MF/SS, management should focus on improving symptoms and cosmesis while limiting toxicity [7]. Numerous studies have shown that prognosis is dependent on the magnitude of the cutaneous tumour burden. Increased skin surface area involvement is also associated with a poorer prognosis, as is lymph node involvement and the appearance of clonal T cells in the peripheral blood [8]. Treatment of early-stage disease (stage IA-IIA) typically involves skin directed therapies that include topical corticosteroids, phototherapy (psoralen plus UVA or UVB), topical chemotherapy, topical or systemic bexarotene, and radiotherapy. Systemic approaches are used for recalcitrant early-stage disease, advanced-stage disease (stage IIB-IV), and transformed disease and include retinoids, such as bexarotene, interferon, histone deacetylase inhibitors, the fusion toxin denileukin diftitox, systemic chemotherapy including transplantation, and extracorporeal photopheresis. Patients with MF/SS are increasingly managed by oncologists, and therefore how approaches for these lymphomas differ from those of other non-Hodgkin lymphomas is important to understand.

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