Unusual presentation of mycosis fungoides as a lump in the scalp

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ABSTRACT

We report a case of mycosis fungoides in a 27-year-old woman who presented with a lump in the forehead. The condition was confirmed with tissue diagnosis. Gallium scintigraphy accurately delineated the complete extent of the disease, and served as a reference for objective assessment of response of the disease to treatment.

Keywords: gallium scintigraphy, lymphoma, mycosis fungoides

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INTRODUCTION

Mycosis fungoides (also known as Alibert-Bazin syndrome) is an indolent lymphoma. The term mycosis fungoides, which is in fact a misnomer, gives an impression of referring to some fungal disease. Literally, it means mushroom-like fungal disease, and was first described by Jean-Louis-Marc Alibert, the founder of French dermatology.[1] Mycosis fungoides is a subtype of non-Hodgkin cutaneous T-cell lymphoma, accounting for almost 50% of all primary cutaneous lymphomas. Its variants are primary cutaneous anaplastic large cell lymphoma, lymphomatoid papulosis, subcutaneous panniculitis-like T-cell lymphoma and primary cutaneous CD4+ small/medium pleomorphic T-cell lymphoma. The aggressive subtypes include Sézary syndrome and primary cutaneous aggressive CD8+ T-cell lymphoma.[2] Mycosis fungoides predominantly manifests in the male gender and shows a predilection for patients in the sixth and seventh decades of life. Children and adolescents are rarely affected. It is rare in Asians and Hispanics but relatively common among Caucasians. Generally, the disease progresses slowly with an unpredictable course.[3]

CASE REPORT

A 27-year-old Asian woman presented with a painless soft tissue swelling in the forehead for the past two months. Detailed history revealed a thickening of skin in her left lateral chest wall with an ulcerating lesion.

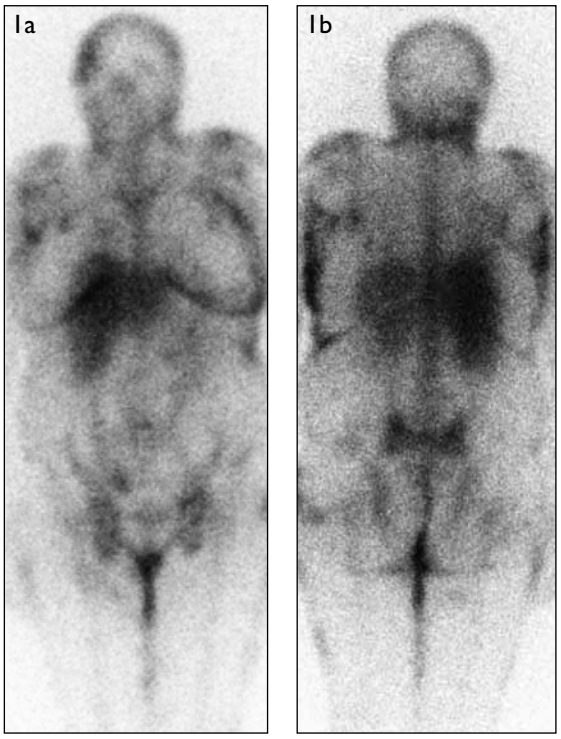

Fig. 1 Gallium whole body images in (a) anterior and (b) posterior views acquired 48 hours post injection show abnormally increased uptake of radiotracer, indicating the presence of disease in various regions of the body.

General physical examination showed that the patient was afebrile and all her vital signs were normal. A thorough systemic examination showed hepatomegaly and bilateral non-tender lymphadenopathy in the axillary and inguinal regions. A total of 90% of the patient's body skin was involved, with diffuse multiple erythematous patches/lesions. Her haematological and biochemical investigations were unremarkable, except for an erythrocyte sedimentation rate (ESR) of 34 mm in the first hour and lactate dehydrogenase (LDH) at 415 (normal range 100–190) U/L. Culture of skin scrapping was negative for pathogenic bacteria. The patient was referred to the Nuclear Medicine Department for further investigation by gallium-67 (Ga-67) whole body scintigraphy. A dose of 185 MBq Ga-67 gallium citrate was injected, and the imaging was performed 48 hours post injection.

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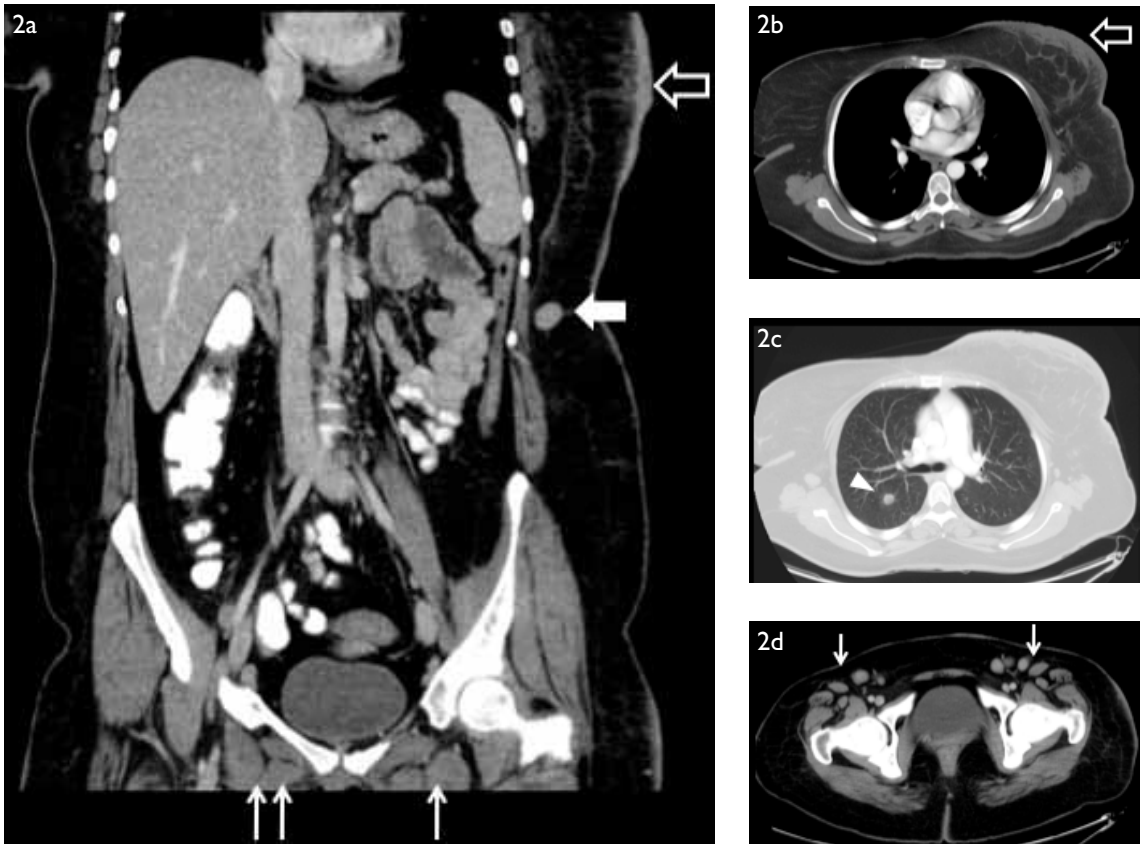

Fig. 2 CT images of the (a) chest; (b,c) abdomen; and (d) pelvis. Significant skin thickening is seen in the chest wall (a & b, hollow arrows) and an enhancing nodule is seen in the left lateral abdominal wall (a, thick arrow). A nodule in the right lung (c, arrowhead) and bilaterally enlarged inguinal lymph nodes are also seen (a & d, thin arrows).

The gallium image (Fig. 1) showed an abnormally increased uptake of tracer in the right half of the skull/scalp region (site of tumour/lesion), and right parotid, left supraorbital, bilateral cervical (more marked on the right side) and bilateral axillary (more marked on the right side) regions, as well as in both halves of the chest, both breasts, left lateral chest wall (ulcerating lesion site) and both the inguinal regions. Liver shadow was apparently enlarged. An important but unusual finding was the abnormally high (Ga-67) counts from the body surface, indicating skin uptake of Ga-67 citrate tracer (Fig. 1).

In order to clarify the findings of the gallium imaging, supplementary investigations, including computed tomography (CT) was performed (Fig. 2). CT showed widespread multiple thickened skin patches (Figs. 2a & 2b), multiple lung nodules (Fig. 2c) and generalised lymphadenopathy (Figs. 2a & 2d). Histopathological examination of the skin biopsy revealed acanthosis, epidermotropic lymphocytes with atypia and abnormal arrangement of collagen fibres in the dermis. Biopsy tissue indicated positivity for cluster of differentiation CD30+ and transformation into large cells. Atypical CD4+ mononuclear cell in addition to CD4+ and CD8+ large cells were also seen. Lymph node biopsy showed the presence of atypical large T-cell (CD 30+) and suspicion of blast transformation into cutaneous T-cell lymphoma (CTLC). Generalised increased uptake of Ga-67 in the skin, lungs and lymph nodes helped to correctly outline the extent of the disease (Fig. 1).

DISCUSSION

Mycosis fungoides is a slow, progressing lymphoma, which affects the skin in the form of patches. The three classical phases of mycosis fungoides consist of the patch, infiltrated plaque and tumour stages. The first phase is also referred to as the 'erythrodermic' phase, in which the patient's condition remains stable for many years and the skin lesions manifest superficially and can ulcerate before progressing into the next phase of the disease. With further progression, mycosis fungoides may involve the lymph nodes and other organs.[4] Atypical T- lymphocytes may be observed on histopathological examination. These phases may overlap or occur simultaneously. The diagnosis of mycosis fungoides is questionable if tumours are the only presentation, without the presence of patches or plaques.[2,5,6]

A detailed physical examination, including measurement of the total skin area involved and an

assessment of any involvement of internal organs, such as the liver, spleen and lymph nodes, is necessary in CTCL patients. The diagnosis of mycosis fungoides is frequently delayed for extended periods, as it can masquerade as other clinical entities such as eczema, dermatitis or fungal infection. Early skin lesions may mimic papulosquamous eruptions like tinea corporis, secondary syphilis or psoriasis.[7] Primary cutaneous lymphomas have various clinicopathological presentations; in order to classify patients correctly, it is crucial to correlate and integrate the clinical features with histopathological data. Investigations that are helpful in staging patients with clinically advanced mycosis fungoides include complete blood cell count, peripheral blood flow cytometry for T-cell subsets, biochemical analysis of blood (renal and liver functions, LDH), chest radiography, biopsy of palpable lymph nodes and bone marrow testing.

CT may be used for staging lymphoma. Magnetic resonance imaging is a competitive modality that provides better tissue discrimination. However, morphological imaging modalities depend on the size of the lymph nodes. Nuclear medicine techniques, on the other hand, depend on radiotracer accumulation mediated by factors associated with tumour composition and pathophysiology. Positron-emission tomography has greater sensitivity, but is more expensive and less widely available. Gallium scintigraphy remains a useful modality due to its widespread availability. Tumour-associated transferrin receptors, anaerobic tumour metabolism leading to low pH (gallium-transferrin dissociation) and increased permeability of tumour neovasculature play important roles in the positivity of gallium imaging.[8,9]

Gallium is a ferric ion analogue with a half-life of 78 hours. It decays with gamma emissions of 93 (37%), 185 (20%), 300 (17%) and 394 (5%) keV photopeaks. Reports regarding uptake of Ga-67 in lesions of mycosis fungoides are scarce. Although the negative predictive value of gallium imaging is low for initial diagnosis of the disease, it provides a powerful tool for evaluation of response to therapy, with reasonably high sensitivity and specificity (approximately 95%).[10,11] Ga-67 citrate is cost-effective and has a longer half-life of 3.26 days, which makes it suitable for delivery worldwide. An additional important clinical aspect of gallium imaging in indolent lymphoma is its ability to check for transformation of low-grade to high-grade lymphoma.

REFERENCES

1. Rapini RP, Bolognia JL, Jorizzo JL, eds. Dermatology. St. Louis, Mosby: Elsevier, 2007.
2. Willemze R, Jaffe ES, Burg G, et al. WHO-EORTC classification for cutaneous lymphomas. Blood 2005; 105:3768-85.
3. Koh HK, Charif M, Weinstock MA. Epidemiology and clinical manifestations of cutaneous T-cell lymphoma. Hematol Oncol Clin North Am 1995; 9:943-60.
4. Lippert BM, Höft S, Teymoortash A, et al. Mycosis fungoides of the larynx: case report and review of the literature. Otolaryngol Pol 2002; 56:661-7.
5. Willemze R, Meijer CJ. Classification of cutaneous T-cell lymphoma: from Alibert to WHO-EORTC. J Cutan Pathol 2006; 33 Suppl 1:18-26.
6. Whittaker S. Biologic insights into the pathogenesis of cutaneous T-cell lymphoma (CTCL). Semin Oncol 2006; 33 (Suppl 3):S3-6.
7. Ackerman AB. If small plaque (digitate) parapsoriasis is a cutaneous T-cell lymphoma, even an 'abortive' one, it must be mycosis fungoides! Arch Dermatol 1996; 132:562-6.
8. Larson SM, Rasey JS, Allen DR, et al. Common pathway for tumor cell uptake of gallium-67 and iron-59 via a transferrin receptor. J Natl Cancer Inst 1980; 64:41-53.
9. Nejmeddine F, Caillat-Vigneron N, Escaig F, et al. Mechanism involved in gallium-67 (Ga-67) uptake by human lymphoid cell lines. Cell Mol Biol (Noisy-le-grand) 1998; 44:1215-20.
10. Bar-Shalom R, Mor M, Yefremov N, Goldsmith SJ. The value of Ga-67 scintigraphy and F-18 fluorodeoxyglucose positron emission tomography in staging and monitoring the response of lymphoma to treatment. Semin Nucl Med 2001; 31:177-90.
11. Kubota K, Ishiwata K, Kubota R, et al. Tracer feasibility for monitoring tumor radiotherapy: a quadruple tracer study with fluorine-18-fluorodeoxyglucose or fluorine-18-fluorodeoxyuridine, L-[methyl-14C]methionine, [6-3H] thymidine, and gallium-67. J Nucl Med 1991; 32:2118-23.