Our Dermatology Online

Case Report

Disseminated eczema or CTCL: Usefulness of high-frequency ultrasonography in the assessment of skin lesions of cutaneous T-cell lymphoma

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ABSTRACT

Cutaneous T-cell lymphoma (CTCL) presents a diagnostic and prognostic challenge in evaluating disease progression. High-frequency ultrasound (HFUS) is a non-invasive imaging technique that has emerged as a valuable tool for assessing dermal alterations, including infiltrative intensity in CTCL. Herein, we report the case of a 74-year-old male patient with a long-standing history of neurofibromatosis type 1 (NF1) since childhood who was also diagnosed with Sézary syndrome (SS) and whose disease remission was confirmed through HFUS examination during PUVA and methotrexate therapy. HFUS facilitated the assessment of infiltrative intensity, visualization of tumor characteristics, and monitoring of disease progression and treatment response. The high-resolution imaging provided by HFUS gave detailed and precise information about the affected skin layers, contributing to the diagnostic process and aiding in the management of CTCL.

Key words: CTCL, Ultrasonography, Diagnostic imaging, Skin diseases, Sézary syndrome

INTRODUCTION

Cutaneous T-cell lymphomas (CTCLs) encompass a heterogeneous group of non-Hodgkin lymphomas characterized by the clonal expansion and infiltration of malignant T lymphocytes within the skin. CTCLs predominantly manifest as primary cutaneous neoplasms without evidence of extracutaneous involvement at the time of diagnosis [1]. The incidence rate of CTCL is reported to be 10.2 cases per million individuals [2], and the conclusive diagnosis of CTCL presents a diagnostic challenge due to the absence of pathognomonic clinical features especially in the early stage [1]. High-frequency ultrasound (HFUS) is an emerging diagnostic modality that holds promise for the evaluation and monitoring of CTCL. Despite being a safe, cost-effective, and non-invasive technique, its application in the assessment of skin neoplasms remains limited. However, preliminary evidence from the literature suggests that HFUS exhibits significant diagnostic value and potential in the management of CTCL, warranting further exploration and validation in clinical settings [3].

CASE REPORT

Skin sonographic examinations were conducted at the Department of Dermatology in Wrocław, utilizing a taberna pro medicum (tpm) GmbH device manufactured in Lüneburg, Germany. The device employed advanced high-frequency ultrasound (HFUS) technology operating at a frequency of 22.5 MHz. The transducer enabled deep tissue penetration up to a maximum depth of 8 mm, providing remarkable vertical (axial) resolution of 80 µm and impressive horizontal (lateral) resolution of 200 µm. To acquire and store data, the state-of-the-art DUBmicro®tpm software was effectively employed. This cutting-edge

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software facilitated the seamless recording and systematic storage of ultrasound data obtained during the meticulously performed examinations. For the evaluation of echogenicity measurements, both A-mode and B-mode imaging techniques were employed.

A 74-year-old male patient diagnosed with neurofibromatosis type 1 was urgently admitted to the Department of Dermatology in April 2023 due to the presence of erythroderma (redness and scaling of the skin) accompanied by generalized desquamation (skin peeling) suggestive of generalized eczema. The patient reported a history of occupational exposure to various irritants, including oils and coolant fluids related to car manufacturing, and intermittent mild skin changes. Additionally, the patient had had skin lesions associated with neurofibromatosis type 1 since childhood and eczema since 2018. The patient provided informed written consent to participate in our study and undergo photographic documentation.

In November 2018, the patient was initially diagnosed with palmoplantar eczema, which responded well to topical glucocorticosteroid treatment. However, the condition later recurred and extended to involve the entire body, prompting a skin biopsy in May 2020 that confirmed generalized eczema. Intravenous antihistamines led to rapid skin improvement. Abnormalities in peripheral blood morphology warranted a hematological consultation and bone marrow examination, which revealed lymphoid infiltration with polymorphic morphology.

Further investigations included erythropoietin assessment, trephine bone marrow biopsy, tryptase concentration measurement, and screening for specific genetic mutations, all yielding negative results. Abdominal and peripheral lymph node ultrasounds in October 2020 were unremarkable, except for a reactive lymph node in the left axillary region.

Dermatological treatment involved systemic glucocorticosteroids and antihistamines, initially resulting in local improvement. Despite continued systemic therapy and medication adjustments, the skin condition gradually worsened. A skin biopsy in March 2021 did not indicate lymphoma yet showed minimal lymphoid infiltration. Peripheral blood immunophenotyping was unremarkable. Treatment approaches included methotrexate, methylprednisolone, and gabapentin.

In May 2021, higher doses of methylprednisolone and gabapentin were administered, alongside methotrexate and topical therapy. Despite initial improvement, the patient discontinued medication, leading to symptom worsening. Methotrexate was reintroduced in August 2021, combined with systemic steroids, antihistamines, gabapentin, and topical treatments. Eventually, methotrexate was discontinued in July 2022 due to a significant improvement in skin lesions and itchiness.

During hospitalization in April 2023, a comprehensive physical examination revealed generalized cutaneous alterations characterized by erythroderma, hyperkeratosis, desquamation, fissuring of the hands and soles of the feet, and numerous neurofibromatosis tumors (Figs. 1a and 1b).

Pertinent laboratory investigations demonstrated leukopenia, elevated erythrocyte sedimentation rate (ESR), increased C-reactive protein (CRP) levels, fasting hyperglycemia (113.0 mg/dL), total bilirubinemia (2.0 mg/dL), elevated lactate dehydrogenase (LDH) activity (286 mg/dL), hyperuricemia (10.6 mg/dL), and raised B-microglobulin levels. Manual peripheral blood smear examination did not reveal the presence of Sézary cells. Furthermore, a histopathological evaluation of skin specimens obtained in February 2023, exhibited an epidermis that was slightly acanthotic with mild hyperkeratosis and focal parakeratosis. Epidermotropic proliferation of small to medium-sized pleomorphic (*cerebriform*) lymphocytes formed intraepidermal Pautrier's microabscesses (Fig. 2). In

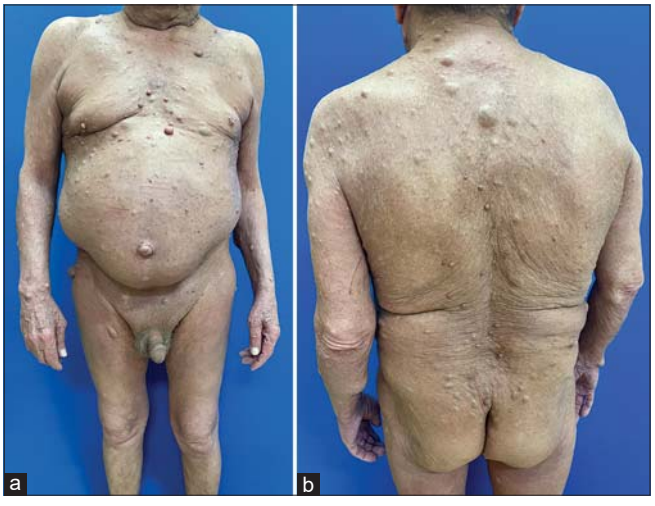

Figure 1: (a) Diffuse erythematous and infiltrated lesions on the trunk and extremities, a feature of CTCL and numerous neurofibromatosis tumors (front).(b) Diffuse erythematous and infiltrated lesions on the trunk and extremities, a feature of CTCL and numerous neurofibromatosis tumors (back).

immunohistochemistry, the neoplastic lymphocytes showed immunophenotype CD3+, CD4+, CD5+, CD8-, and CD7-/+ (partial loss). Cytotoxic markers such as TIA-1 and granzyme B were negative. In the course of employing HFUS to examine skin affected by Sézary syndrome, we ascertained pronounced infiltration within the dermal layer, coupled with notable augmentation of its thickness (Fig. 3a). Additionally, we also conducted an examination of neurofibromatosis lesions (Fig. 3b). During hospitalization, PUVA-therapy and topical therapies such as Encortolon ointment and urea ointment were administered, resulting in alleviation of pruritus. Nevertheless, due to an inadequate therapeutic response following PUVA treatment (total dose was 37.08 mJ/m^2 during ten sessions), methotrexate was reintroduced. Also, mSWAT was completed with a score of 51.

DISCUSSION

Despite the growing interest in HFUS as a diagnostic tool for patients with CTCL, there remains a limited number of studies in the literature focused on this particular area [4-7]. In a study conducted by Yazdanparast et al. [4] comprising a cohort of 21 CTCL patients, HFUS examination revealed significant differences in hydration, pH, melanin content, and erythema index between the lesional areas and unaffected skin. Moreover, the dermal echo density in the CTCL lesions was found to be significantly reduced compared to the adjacent normal skin. These findings underscore the potential utility of HFUS as a non-invasive and cost-effective modality for the diagnostic evaluation and longitudinal monitoring of CTCL. In a study conducted by Zi Han Niu et al. [5], involving a cohort of 62 patients presenting with erythema and scales, the investigation of epidermal thickness and subepidermal low-echo band (SLEB) thickness demonstrated a notable diagnostic significance in the differential diagnosis of early-stage mycosis fungoides from psoriasis vulgaris and eczema. A study conducted by Polańska et al. [6] demonstrated the significance of the subepidermal low-echo band (SLEB) parameter measured using high-frequency ultrasound (HFUS) in patients with CTCL. The investigation observed a notable decrease in SLEB thickness among patients undergoing phototherapy, implying that monitoring this parameter in CTCL patients could serve as a valuable tool for assessing the efficacy of treatment interventions.

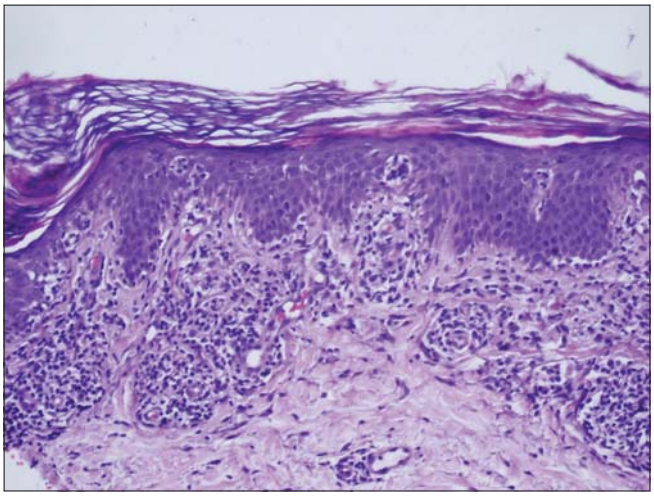

Figure 2: Dense upper dermal band-like epidermotropic infiltrate by atypical lymphocytes forming Pautrier's microabscesses (H&E).

In a study conducted by Iris Wohlmuth-Wieser et al. [7] comprising a cohort of thirteen patients, HFUS examination demonstrated an augmented epidermal thickness in individuals diagnosed with CTCL and psoriasis when compared to those with atopic dermatitis. Our observations indicated conspicuous dermal infiltration and heightened dermal thickness in regions affected by Sézary syndrome, thereby accentuating the informative capacity of HFUS in delineating disease-associated alterations.

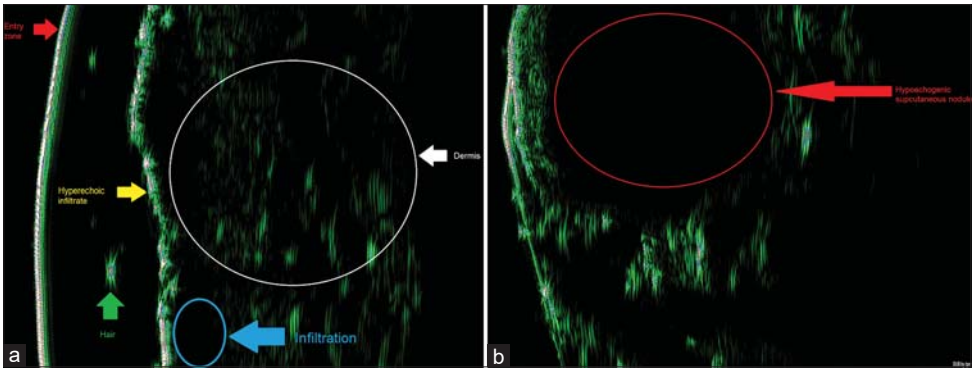

Figure 3: (a) Massive infiltration in the dermis (sonographic picture). (b) Sonographic feature of the neurofibroma in the skin.

CONCLUSION

The use of HFUS for the evaluation of CTCL furnishes valuable insights into both the underlying pathophysiology of the disease and its diagnostic prospects. While HFUS exhibits promising diagnostic utility in CTCL, the existing literature offers a relatively circumscribed exploration of its comprehensive capabilities. Noteworthy investigations conducted by disparate researchers have underscored HFUS's aptitude in discerning CTCL from other dermatological conditions based on parameters such as hydration, pH, melanin content, and echo density. Moreover, inquiries into the thickness of the subepidermal low-echo band (SLEB) have unveiled its potential as an indicator of treatment responsiveness and as a discriminator between CTCL and alternative skin disorders.

REFERENCES

1. Hristov AC, Tejasvi T, Wilcox RA. Mycosis fungoides and Sézary syndrome: 2019 update on diagnosis, risk-stratification, and management. Am J Hematol. 2019;94:1027-41.
2. Korgavkar K, Xiong M, Weinstock M. Changing incidence trends of cutaneous T-cell lymphoma. JAMA Dermatol. 2013;149:1295-9.
3. Wang Y, Niu Z, Liu J, Zhu Q, Liu Z, Liu Y, et al. Value of high-frequency ultrasound in accurate staging of mycosis fungoides/Sézary syndrome. J Ultrasound Med. 2020;39:1927-37.
4. Yazdanparast T, Yazdani K, Ahmad Nasrollahi S, Izadi Firouzabadi L, Humbert P, Khatami A, et al. Biophysical and ultrasonographic changes in early patch/plaque stage of mycosis fungoides, compared with uninvolved skin. Skin Res Technol. 2021;27:1029-34.
5. Niu Z, Wang Y, Zhu Q, Liu J, Liu Y, Jin H. The value of high-frequency ultrasonography in the differential diagnosis of early mycosis fungoides and inflammatory skin diseases: A case-control study. Skin Res Technol. 2021;27:453-60.
6. Polańska A, Dańczak-Pazdrowska A, Olek-Hrab K, Osmola-Mańkowska A, Bowszyc-Dmochowska M, Żaba R, et al. High-frequency ultrasonography: New non-invasive method in assessment of skin lymphomas. Skin Res Technol. 2018;24:517-21.
7. Wohlmuth-Wieser I, Ramjist JM, Shear N, Alhusayen R. Morphologic features of cutaneous T-cell lymphomas using dermoscopy and high-frequency ultrasound. J Clin Med. 2020;10:17.

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