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OVERVIEW OF MYCOSIS FUNGOIDES AND SÉZARY SYNDROME

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ABSTRACT

Mycosis fungoides and Sézary syndrome are common cutaneous T-cell lymphomas: a type of non-Hodgkin lymphoma. An overview of the literature and a range of computed tomography images of Sézary syndrome are presented.

Keywords: cutaneous, lymphoma, lymphadenopathy, subcutaneous oedema

LAY ABSTRACT

A skin type of cancer that spreads to lymph nodes and organs is described.

INTRODUCTION

A lymphocyte is a type of white blood cell in our immune system. A lymphoma occurs when lymphocytes grow and uncontrollably multiply. The two most common types of lymphoma are: Hodgkin's lymphoma and non-Hodgkin's lymphoma (NHL).[1] This overview covers different types of lymphocytes, mycosis fungoides (MF), and Sézary syndrome (SS).[2] Examples of SS at computed tomography are presented.

LYMPHOCYTES

Blood consists of red blood cells (erythrocytes), white blood cells (lymphocytes, for example), platelets (thrombocytes), and plasma. A lymphocyte is a type of white blood cell in our immune system: B-lymphocytes (B-cells), T-lymphocytes (T-cells), and natural killer cells (NK cells). B-cells fight infections, T-cells attack foreign cells (e.g., cancer cells), and NK cells contain substances which, for example, can kill tumour cells.[3]

LYMPHOMA CELLS

T-cell and B-cell are the first and second lineage of lymphoma cells; and natural killer (NK) cells the third lineage.[3] T-lymphocytes account for 15% of all NHLs.

Extranodal NK/T-cell lymphoma is an aggressive malignancy. It occurs in two forms: the nasal type, and leukaemia. It develops almost exclusively in non-nodal sites: 80% occur in the nose, nasopharynx, oropharynx, and Waldeyer's ring. NK/T-cell lymphoma may rarely infiltrate the liver, spleen, skin, lymph nodes as well as bone marrow. Peripheral blood may also be involved. This condition is referred to as aggressive NK/T-cell leukaemia/lymphoma.[3]

MYCOSIS FUNGOIDES (MF)

MF (aka Albert-Bazin syndrome) is the most common form of cutaneous T-cell lymphoma (CTCL). In MF T-cell lymphocytes become cancerous. MF is a type of NHL. MF usually affects the skin, but may progress internally over time. T-cells may be slow or fast (aggressive) growing.[2] Symptoms may include dry skin, itching which may be severe, and erythroderma. MF occurs more often in men usually between 50-60 years. Non-specific skin lesions may be the onset of disease causing delay in diagnosis. MF may become aggressive over time. Sèzary cells are formed when T-cell lymphocytes occur in the blood. In 20% of patients CTCL may become highly malignant with dissemination into various organs. Systemic infection, usually from *Staphylococcal* or *Pseudomonas* organisms, may lead to death.[1,2]

Signs and symptoms of MF vary from patient to patient. Cutaneous manifestations may present as patches, plaques, which are typically thicker raised lesions, or tumours which are raised lesions and may ulcerate. Patients may have all three presentations or only one. MF usually presents with flat, red scaly patches hence may be misdiagnosed as psoriasis, eczema or dermatitis.

Treatment of MF can be divided into skin-directed or systemic therapies. MF is sensitive to radiation. The treatment varies from topical corticosteroids to topical chemotherapies (e.g., nitrogen mustard). Other treatment options are electron beam radiation and phototherapy. The skin and affected internal organs may be treated by systemic therapies. Use may be made of chemotherapeutic drugs and biologic modifiers.[1,4] MF accounted for 72% of CTCL cases reported for the period 1973-2002 in the United States of America.[5]

SÉZARY SYNDROME (SS)

It is not known whether SS is an advanced form of MF or a separate disease. SS is a T-cell lymphoma of the skin and peripheral blood and was recognised in 1938.[2] It is a type of NHL.

SS accounted for 2.5% of CTCL cases reported for the period 1973-2002 in the United States of America.[5] Staging of patients is useful for guiding and treatment and in assessing progress.[6] There are four treatment groups.

- T1: platelets and plaques affecting <10% of body surface area
- T2: patches and plaques affecting >10% of body surface area
- T3: presence of tumours (i.e., raised dome-shaped lesions >1cm in diameter)
- T4: erythroderma affecting >80% of body surface area.[7]

It is not invariable for patients to progress from one T stage to another. Patients with T1 disease have an excellent prognosis and healthy life expectancy. Those with T2 disease have a median survival of 10-12 years with a 25% risk of disease progression to a more advanced level. T3 means that the disease is usually in an advanced stage; erythroderma affects more than 80% of the body surface in T4 disease. Oedema may occur.

Those with visceral involvement are high risk cases with a median survival of 1-2 years. Those who only have tumours or erythroderma have a median survival of 4-5 years. Those who only have erythrodermic CTCL have a worse prognosis than those with patch or plaque disease. Advanced age (>65 years), number of previous treatments, enlargement of peripheral lymph nodes and leukaemia cells in the blood worsen the prognosis.[7]

ROLE OF IMAGING IN MF AND SS

CT should be performed as part of the initial staging, and as a follow-up baseline in patients with advanced MF, SS, and variant CTCL. Upstaging of the disease may also occur. Inguinal and axillary nodes occur frequently. Visceral involvement is infrequent.[8]

According to Raanani[9] CT and PET/CT are mainly used for diagnosis of MF. Since internal organ involvement without lymphadenopathy is rare in patients with T1 and T2 disease means that CT has limited value. It is routinely used in those with T3-4 disease to assess degree of involvement of viscera and lymph nodes. CT assessment of lymph node enlargement is however limited to size and shape of the node, and is also poor in detecting bone marrow involvement.[9]

PET/CT can increase the sensitivity of detection of affected lymph nodes and to judge the response to treatment with those in the advanced disease stage. The superior sensitivity of fludeoxyglucose (FDG) has resulted in an increased use for staging and response to treatment of aggressive lymphomas. 18FDG PET/CT plays an important role in (i) the detection and staging of lymphoma, (ii) restaging after treatment, (iii) monitoring treatment response, and (iv) radiotherapy planning.[10] PET/CT improves detection of extranodal involvement and accuracy of baseline staging compared to CT.[11] MRI is used in the diagnosis and follow-up of extranodal T-cell lymphoma.[12]

Pulmonary involvement may occur in SS.[13] Figures 1a and b depict pulmonary involvement in SS. The tumour-node-metastasis-blood (TNMB) classification of SS is used to assess lymph node involvement.[14] Figure 1a and Figures 2 to 4 show lymph node involvement. Oedema may occur in SS. Figures 5a and b show prominent subcutaneous oedema.

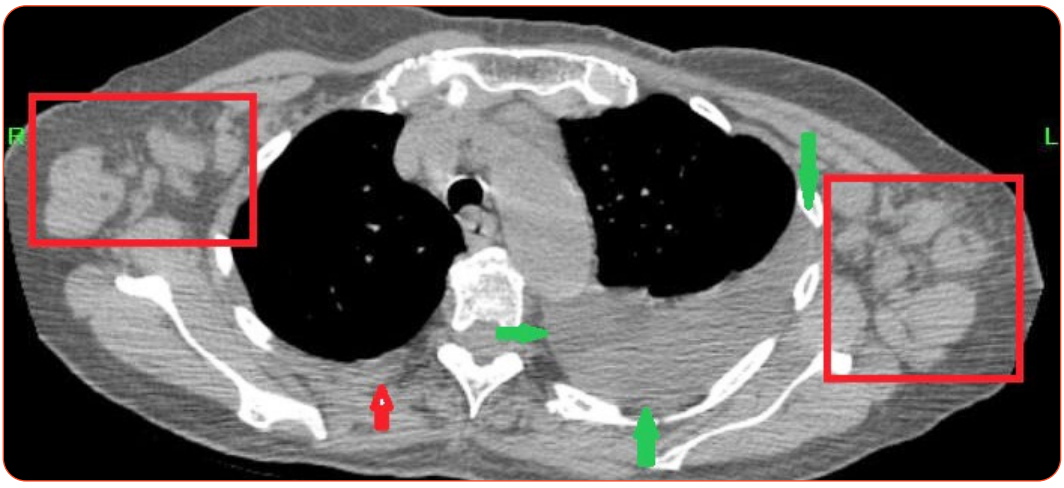

Figure 1a. Axial view showing bilateral pleural effusion. Green arrows (L). Red arrow (R). L>R. Large bilateral axillary lymphadenopathy (red squares).

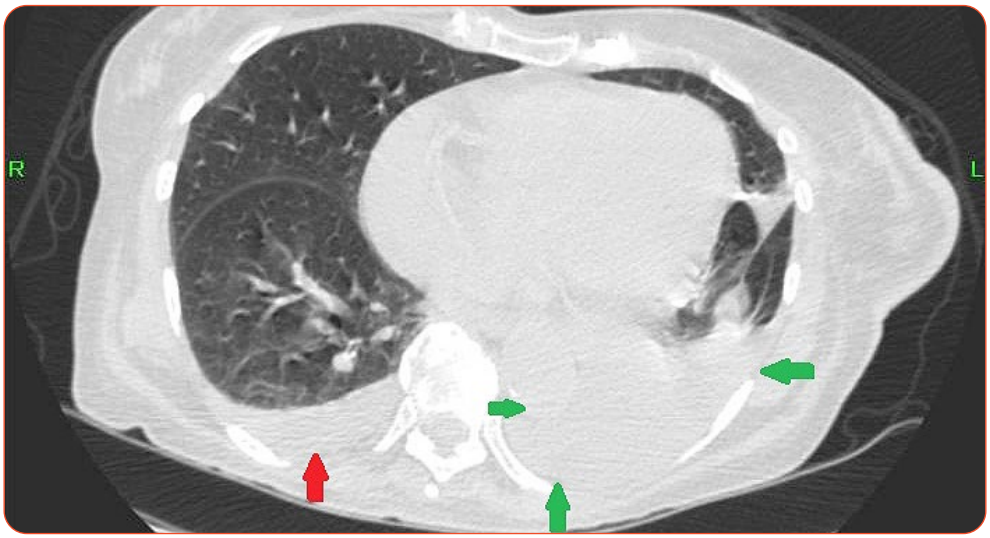

Figure 1b. Axial view showing bilateral pleural effusion. Red arrow (R). Green arrows (L). No parenchymal masses.

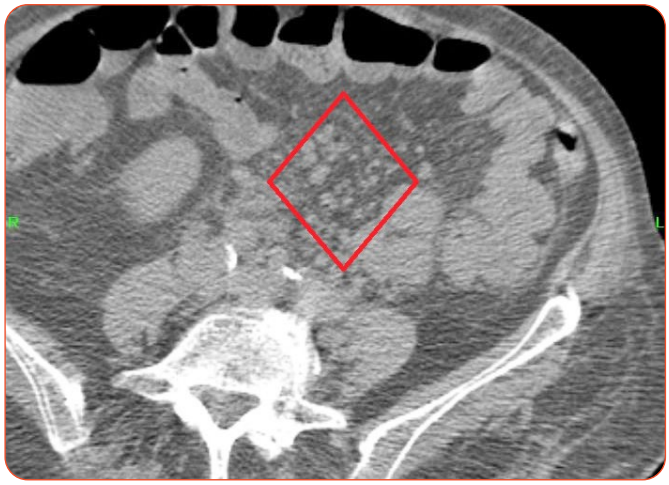

Figure 2. Axial view showing multiple enlarged mesenteric nodes (red diamond).

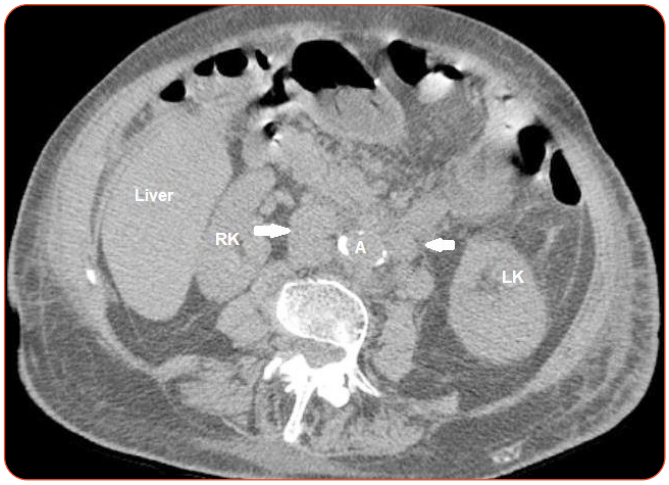

Figure 3. Axial view showing para-aortic lymphadenopathy (white arrows). A = aorta. RK = right kidney. LK = left kidney.

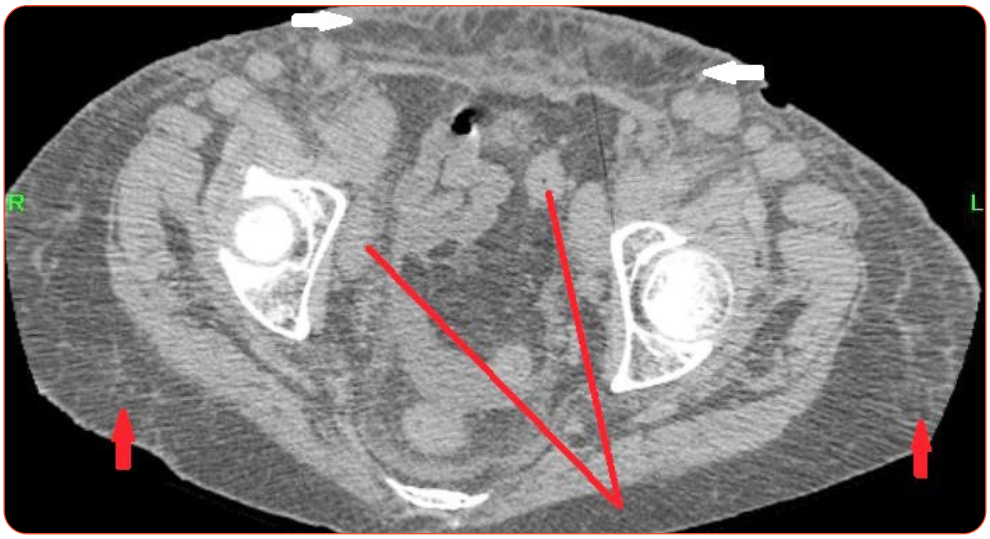

Figure 4. Axial view showing prominent subcutaneous oedema anteriorly in the lower abdominal wall (white arrows) and posteriorly involving the lower back and buttocks (red arrows). Red lines = pelvic lymphadenopathy.

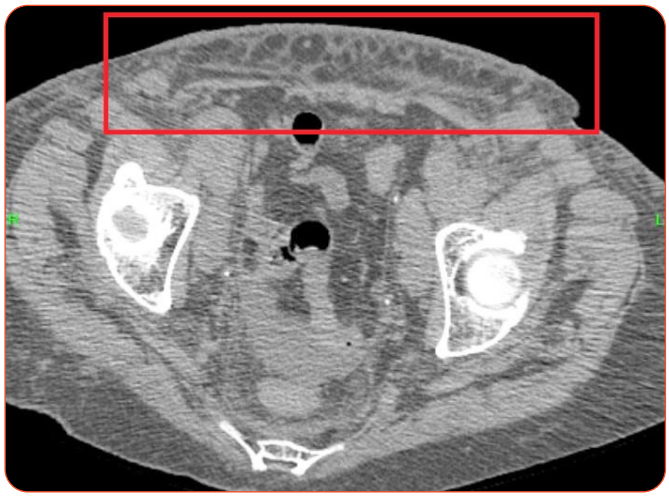

Figure 5a. Axial view showing prominent subcutaneous oedema anteriorly in the lower abdominal wall (red rectangle).

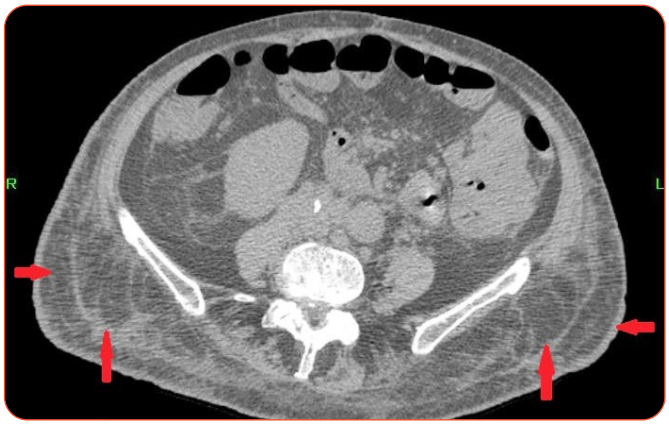

Figure 5b. Axial view showing prominent subcutaneous oedema posteriorly (red arrows).

CONCLUSION

Mycosis fungoides and Sézary syndrome are cutaneous T-cell lymphomas. They are a type of non-Hodgkin lymphoma. Imaging is useful in staging and follow-up of extranodal T-cell lymphomas.

CONFLICT OF INTEREST

Nil to declare.

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