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Sézary Syndrome: A Documented Case at Yalgado Ouédraogo University Hospital in Ouagadougou, Burkina Faso

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Abstract

Sézary syndrome is characterized by erythroderma, diffuse adenopathy, atypical T lymphocytes in the blood (> 1000/µ Liter), and a dominant T clone in the blood, skin, and lymph nodes.

Sézary syndrome is rare (3% of cutaneous T-cell lymphomas). There have been hospital-based studies in Africa, but there is no estimate of its frequency.

It is an aggressive cutaneous T-cell lymphoma with a 5-year survival rate of 24%.

New hopes from targeted therapies should improve survival.

This article aims to review the clinico-biological and progressive picture under dermocorticoid, chlorambucil, and prednisone therapy in an 80-year-old man presenting with mycosis fungoides, adenopathies, hyperlymphocytosis, and Sézary cells.

Keywords: Sézary syndrome; cutaneous T lymphoma; flow cytometry; Burkina Faso

Introduction

According to the *European Organization for Research and Treatment of Cancer* (EORTC), Sézary syndrome (SS) is defined by the presence of erythroderma, diffuse adenopathies, atypical T lymphocytes in the blood (> 1000/mm3) and a dominant T-cell clone in the blood, skin and lymph nodes. In 2001, KIR3DL2 [KIR for killer immunoglobulin-like receptors] receptor (CD158k), a specific marker for atypical T lymphocytes in tumors called Sézary cells, was identified. It enables a more accurate diagnosis of Sézary syndrome, the level of which correlates well with the clinical course of the disease [1]. Sézary syndrome may arise de novo or follow mycosis fungoides. Mycosis fungoides is a dermatosis that evolves in 3 characteristic clinical stages. It is the typical form described by Albert and Bazin, with premycotic erythema, the infiltrated plaque stage, and the tumor stage [1].

Sézary syndrome is rare (3% of cutaneous T lymphomas): 30 to 40 new cases annually in the USA [2, 3].

It is more common in men, especially those over 60 [4].

Sézary syndrome ranks among the aggressive cutaneous T lymphomas. The prognosis is poor, with a five-year survival rate of around 25% and a median survival between 2 and 4 years [5].

The pathophysiology of SS is poorly known. This lymphoma is composed of mature memory T lymphocytes with a cutaneous tropism linked to the expression on their surface of specific markers such as CLA (cutaneous lymphocyte antigen), which interacts with the E-Selectin receptor present in the post-capillary venules of the dermis. These T lymphocytes are mainly recruited to the skin during cutaneous infections to prepare for an eventual specific immune response [6]. This oncogenicity could be due to immune dysregulation, especially an imbalance of cytokines: excess production of cytokines corresponding to the TH 2 profile (IL-4, IL-5, IL-10) would result in a marked reduction in apoptosis and the accumulation in the skin of these memory T lymphocytes, multiplying clonally [6].

In sub-Saharan Africa, the few cases diagnosed are not systematically published. Histology coupled with immunohistochemistry and immunophenotyping of blood lymphocytes are essential for diagnosis, but they are costly for patients in these countries with limited resources, thus limiting access to diagnosis. In these countries, geographical and financial access are also barriers to treatment. To our knowledge, there are no published cases of Sézary syndrome in Burkina Faso.

This study aimed to review the clinico-biological and progressive picture under dermocorticoid, chlorambucil, and prednisone therapy, with a literature review based on an observation of an 80-year-old man.

Observation

Mr. Z.A., an 80-year-old electrician, was referred for a hematology consultation at the Yalgado OUEDRAOGO University Hospital in Ouagadougou in January 2023 for anorexia, generalized body pruritus, diffuse erythematous infiltrated lesions with a scaly surface, and hyperleukocytosis with lymphocyte predominance on the automated blood count.

The illness reportedly began in August 2022 (six months ago) with a cracking of the fingers, hands, and feet and a feeling of stiffness in the feet. He consulted a peripheral health center, where an allergy to mosquito repellent cream was suspected. He then went for a dermatology consultation at Yalgado Ouédraogo University Hospital with a prescription for desloratadine tablet ("tablet") 5 mg: 1 tablet per day per os. In dermatology, he underwent a skin biopsy, a topical treatment with salicylate vaseline 5% applied twice a day, which resulted in incomplete remission; deworming with albendazole 400 mg/day for three days and intramuscular betamethasone 7 mg/ 3 weeks, which produced a transient complete remission but persistent hyperlymphocytosis, led to a referral to hematology.

He had a personal history of cerebrovascular accident in 2020 and a family history of a brother with diabetes. He was neither an alcoholic nor a smoker. He had anorexia and was up to date on vaccines. On clinical examination, the patient was apyretic at 37°3, had a WHO performance status of 0, and had a body mass index of 30.86 kg/m2. He had no splenomegaly, hepatomegaly (liver span at 12 cm in the midclavicular line), or conjunctival pallor. He presented with generalized, painless pruritic lesions associated with infiltrated, diffuse erythematous lesions with scaly surfaces (= erythroderma); palmoplantar keratoderma; onychodystrophy; alopecia; and loss of hair on the tails of the eyebrows (fig. 2, 3 and 4). The patient had a non-inflammatory left axillary adenopathy measuring 1.5 cm in diameter and a non-inflammatory right epitrochlear adenopathy measuring 1.5 cm. Table 1 shows the hemogram before treatment.

Table 1: Hemogram before treatment, and on D8 and D218 of treatment

Hemogram			
Hemogram components	Before treatment	On D8 of treatment with Prednisone, Chlorambucil, andCiprofloxacin	On D218 of treatment with Prednisone and Chlorambucil
White blood cells	77.46 Giga/L	69.30 Giga/L	34.43 Giga/L
Immature granulocytes	None	0.11 Giga/L	0.04 Giga/L
Neutrophils	5.65 Giga/L	8.76 Giga/L	3.76 Giga/L
Eosinophils	0.62 Giga/L	0.020 Giga/L	0.13 Giga/L
Basophils	0 Giga/L	0.28 Giga/L	0.08 Giga/L
Lymphocytes	69.87 Giga/L	58.28 Giga/L	29.41 Giga/L
Monocytes	1.32 Giga/L	1.96 Giga/L	1.05 Giga/L
Hemoglobin level	144.0 g/L	141.0 g/L	13.4 g/L
Mean corpuscular volume	89.7 fl	82.6 fl	83.41 fl
Mean corpuscular hemoglobin level	33.7 pG	29.9 pG	29.6 pG
Mean corpuscular hemoglobin concentration	37.5 %	36.2 %	35.5 %
RBC distribution index (RDW)	41.6 % AnisocytosisRDW≥ 16 %	43.2 % Anisocytosis RDW≥ 16 %	45.6 Anisocytosis RDW≥ 16 %%
Platelet	252.0 Giga/L	276.0 Giga/L	183.0 Giga/L

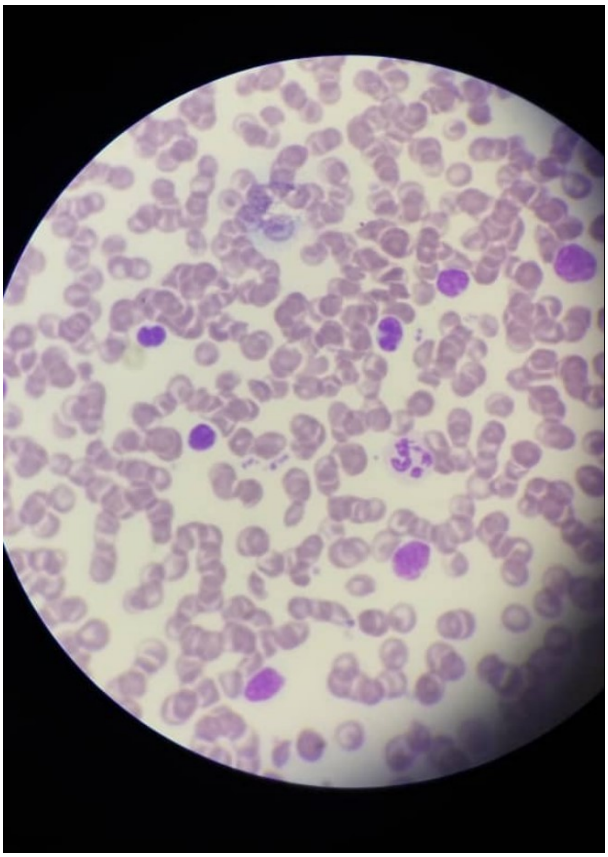

Figure 1: Blood smear

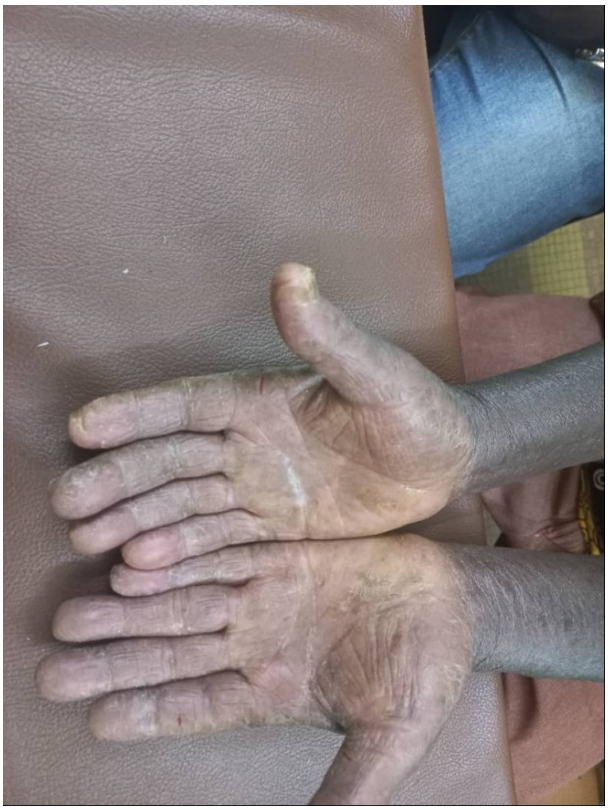

Figure 2: Anterior aspect of forearms and palmar aspect of hands before treatment.

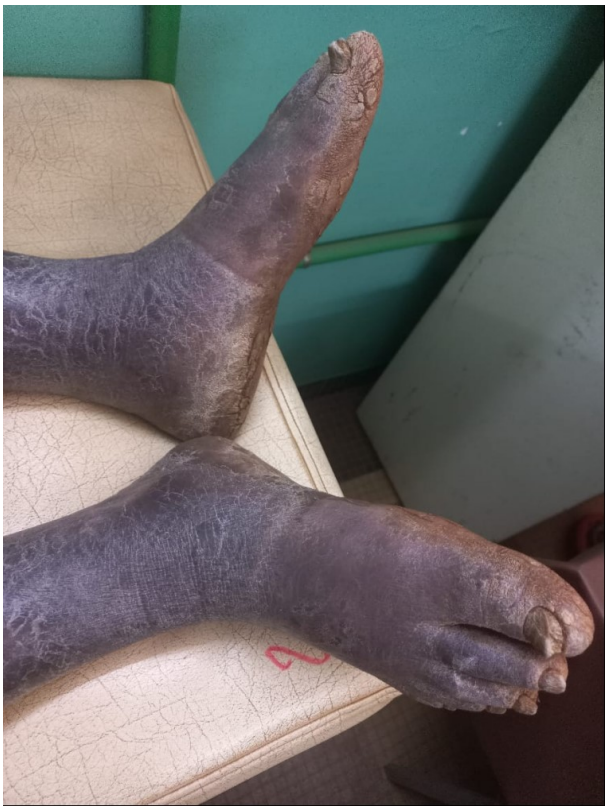

Figure 3: Dorsal aspect of the lower 1/3 of the legs and dorsal aspect of the feet before treatment.

Figure 4: Face before treatment

The blood smear (Figure 1) revealed 42% atypical lymphocytes, representing 30,742.8 atypical lymphocytes/cubic millimeter (large cells, high nucleo-cytoplasmic ratio, often irregular nucleus contour, fairly dense nucleus chromatin, "cerebriform" appearance of the nucleus), suggestive of Sézary cells.

Histology of the skin biopsy found a fragment of skin tissue with a thinned, hyperkeratotic epidermis and a mucous layer of irregular thickness. The basement membrane was regular. There was a moderate inflammatory infiltrate of the papillary and middle dermis, with cells with hyperchromatic nuclei and irregular outlines. Exocytosis was observed in the form of serosa-free cell clusters or isolated cells; the rest of the dermis appeared normal.

Immunophenotyping of blood lymphocytes highlighted a T hyperlymphocytosis (NK lymphocytes 0%, B lymphocytes 0%) expressing CD3, CD4, and CD5 markers, with loss of CD7 representing around 98% of lymphocytes, i.e. 60 Giga/L (= 60,000 Sézary cells/mm3); CD3+CD5+ were 99% and CD3+CD4+CD7- 100%. There was monotypy of the variable part of the T receptor: CD3+ TCR alpha-beta at 99%.

The pre-treatment workup was as follows: antiretroviral, anti-hepatitis B and C, Human T-Cell Lymphotropic Virus 1 (HTLV1) serologies were negative; fasting blood glucose 4.81 mmol/L; uricemia: 509.52 µmol/L; LDH:326 UI/L; calcemia: 2.25 mmol/L; natraemia: 145 mmol/L; kalemia: 4.10 mmol/L; magnesemia: 0.75mmol/L; phosphatemia 1;11 mmol/L; chloremia: 109.0 mmol/L; bicarbonate: 29.97 mmol/L; total proteinemia: 80.0 G/L, albuminemia 37.4 G/L (normal 40.2 to 47.6 G/L), discrete polyclonal gamma peak 20.7G/L.

Cervico-thoraco-abdominopelvic CT scan showed no splenomegaly, hepatomegaly, or deep adenopathy.

ECG: normal; cardiac echodoppler: dilated cardiomyopathy (dilatation of all 4 chambers), preserved systolic function (LVEF 63%), grade II to III MI, moderate PAH (42 X 10 mm Hg).

Our patient qualified as T4N3M0B2 according to the TNMB (tumor nodes metastasis blood) Revised classification [1, 7] and stage IVA2 of the European Organization for Research and Treatment of Cancer (EORTC) [1].

He had received chlorambucil 8 mg/day po from D1 to D3 combined with prednisone 120 mg/day po from D1 to D7, allopurinol 300 mg/7 days for D1 = D30. He underwent deworming before treatment with albendazole 400 mg/day p.o. as a single dose with a meal. Adjuvant treatment to the corticosteroid therapy included omeprazole, Diffu-K, a diet rich in calcium and vitamin D, and a low-salt diet. In addition to clinical monitoring, the work-up included blood count, complete blood ionogram, uremia, glycemia, serum urea creatinine, AST, and ALT.

The progression was notable on day 8 of treatment, with desquamation, melting of cutaneous edema (figure 5), and halving of adenopathy.

On day 9, fistulas appeared on the posterior and medial aspect of the buttocks (figure 6), along with a neutrophilic polynucleosis of 8.76 G/L. Ciprofloxacin 500 mg x 2/day for ten days cleared the pustules.

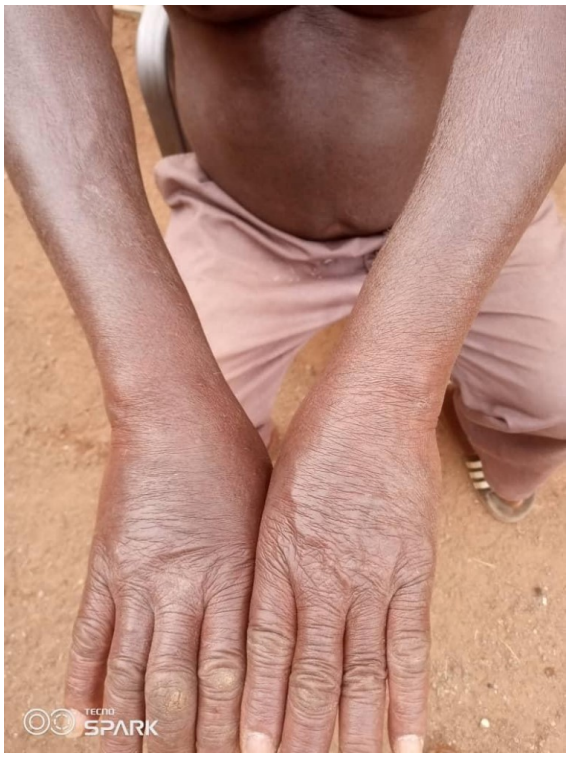

Figure 5: Back aspect of hands and forearms, front aspect of abdomen.

Treatment with dermocorticoids had improved the scaly lesions, which had reappeared on D21 of treatment, hence the use of dermocorticoids on D20 of each course of treatment.

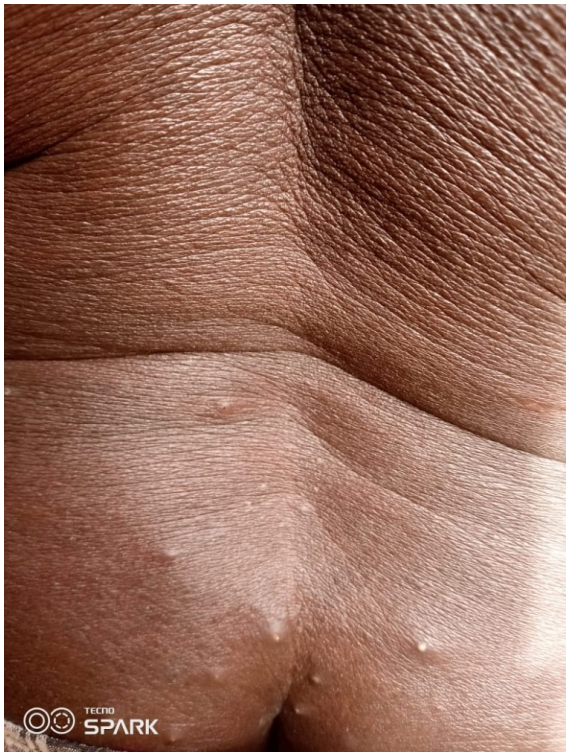

Figure 6: Posterior aspect of chest and buttocks with pustules.

On D136 of treatment, there was a weight loss of 3 kg.

On D224, Sezary cells were 8% in the blood smear, i.e. 3885.68 Sezary cells/µL (white blood cells 48.57 Giga/L, lymphocytes

35.94 Giga/L).

Discussion

The limitations of our study included the absence of evidence of a blood T cell clone detected by Southern blot or PCR, the lack of evidence of chromosomal abnormalities of the tumor T clone, and the absence of GeneScan analysis (GS or standardized BIOMED2-GS method), a recent very sensitive and accurate analysis method for detecting the presence of a blood and skin T clone (from at least two skin biopsies in two different sites and one blood sample). GS analysis differentiates clonal heterogeneity (coexistence of 2 or more T cell clones in several samples from the same patient) from clonal homogeneity. Clonal homogeneity is associated with epidermotropism, a denser histological infiltrate, and a higher number of circulating tumor cells, suggesting a poor prognostic factor in CTCL (cutaneous T-cell lymphomas). Our patient did not undergo KIR3DL2 testing. KIR3DL2 is a member of the KIR (killer immunoglobulin-like receptor) family of natural killer (NK) lymphocyte receptors, whose genes lie on chromosome 19 at q13.4 [9]. This KIR3DL2 is highly specific to Sézary syndrome because it is not significantly present on the surface of CD4+ cells in healthy subjects or patients with erythroderma. This KIR3DL2 can differentiate inflammatory erythroderma from true Sézary syndrome and mycosis fungoides from Sézary syndrome, both currently regarded as two sides of the same pathology. Clonal heterogeneity is more frequent in SS (80%) than in MF (48%). GS analysis would also identify patients who respond to treatment, particularly extracorporeal photochemotherapy (ECT) [8].

Sézary syndrome is a rare disease accounting for 2-3% of all lymphomas and 70% of CTCLs. The incidence of Sézary syndrome in France is 50-60 new cases per year [9].

Our patient was 80 years old. DIAKITE M. et al. had diagnosed Sézary syndrome in a 63-year-old man [10]. The disease predominates in men [9, 10,11]. Our patient developed erythroderma, adenopathies, palmoplantar keratoderma, onychodystrophy, alopecia, and intermittent pruritus consistent with the literature [1, 10, 11]. DIAKITE M. et al. found no adenopathy, onychodystrophy, or alopecia [9].

Our case had a hyperlymphocytosis of 69.87 Giga/L, compared with a discrete hyperlymphocytosis of 5.3 Giga/L in the case of DIAKITE M et al. [9]. At diagnosis of Sézary syndrome, there is no hyperleukocytosis or hyperlymphocytosis, even when the number of Sézary cells exceeds 1,000/mm3[9].

Immunophenotyping of our patient's blood lymphocytes revealed T-cell hyperlymphocytosis expressing markers CD3, CD4, and CD5 with loss of CD7, accounting for around 98% of lymphocytes, i.e. 60 Giga/L (= 60,000 Sézary cells/mm3). DIAKITE M et al. found the following phenotypic profiles: CD2+, CD3+, CD4+, and very low CD20[9]. In Sézary syndrome, pan-T CD7 antigen (CD4+ / CD7-) [2]. CD7 is a membrane glycoprotein of the immunoglobulin family found on 85-90% of blood T cells in healthy adults, thymocytes, and many NK lymphocytes. CD7 is a co-stimulator of T cells. Its loss results in resistance to apoptosis and survival of tumor cells. The cut-off point for significant antigen loss was set at 40% of CD4+ lymphocytes by the International Society for Cutaneous Lymphoma: ISCL [2]. This threshold is never reached in inflammatory dermatoses except in 55% of Sézary syndromes.

Our 80-year-old patient was classified T4N3M0B2 by the TNMB classification and stage IVA2 by the EORTC. He underwent treatment with chlorambucil combined with prednisone every 21 to 28 days. From day 20 of each course of chlorambucil-prednisone, the erythroderma reappeared, and the dermocorticoid was applied morning and evening. Before treatment, Sézary cells were 60,000/µL (white blood cells 77.46 Giga/L, lymphocytes 69.87 Giga/L). At D224 after treatment, Sézary cells were 3,885.68/µL (white blood cells 48.57 Giga/L, lymphocytes 35.94 Giga/L). The indicated treatments for Sézary syndrome were unaffordable for him: extracorporeal photochemotherapy, combined possibly with subcutaneous peginterferon α or bexarotene *per os* [11]; second-line, intravenous mogamulizumab and brentuximab vedotine [11], and romidepsin, a histone deacetylase in-

hibitor [11] were also unaffordable.

Conclusion

Sézary syndrome is a rare cutaneous epidermotropic T lymphoma with a poor prognosis, as there is currently no curative treatment. Our case involves an 80-year-old patient at an advanced stage of the disease who had progression-free survival on the chloraminophen-prednisone protocol until D224 of treatment.

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