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Sezary Syndrome: A Clinico-biological Study of 5 Cases

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Abstract: *Background*: Sézary syndrome (SS) is a rare erythrodermic and leukemic variant of cutaneous T-cell lymphoma (CTCL) that belongs to the group of non-Hodgkin's lymphomas (NHL) resulting from malignant proliferation of skin-homing T cells. We report through a series of 5 cases, the experience of the hematology laboratory in the diagnosis of the syndrome of Sézary. *Methods*: This is a retrospective study of 5 cases of Sezary syndrome collected in the dermatology department of Marrakech. *Results*: five patients were identified with the clinicopathological criteria of SS. At the time of diagnosis, all 5 patients had erythroderma and generalized lymphadenopathy in both superficial and deep stations. The white blood cell count was elevated (>10,000 WBC/mm^3) in all 5 patients with a mean value of 18,120 WBC/mm^3. The blood smear showed the presence of 75% (27 G/l) of small to medium-sized cells with a high nucleocytoplasmic ratio and cerebriform nuclei typical of Sezary cells and suggests the diagnosis of SS. *Conclusions*: The diagnosis of SS remains a challenge in many situations, the pathophysiology and definition of SS have evolved significantly over the past decades.

Keywords: Sezary Syndrome, Lymphoma, T-cell, Blood Smear

1. Introduction

Sézary syndrome (SS) is a rare erythrodermic and leukemic variant of cutaneous T-cell lymphoma (CTCL) that belongs to the group of non-Hodgkin's lymphomas (NHL) resulting from malignant proliferation of skin-homing T cells [1]. SS and mycosis fungoides (MF) are the most common forms of CTCL, accounting for approximately 65% of cases, while SS accounts for approximately 3% of all CTCL [2]. CTCLs are assumed to be predominantly male and the median age of onset is between the fifth and sixth decade [3].

In order to establish the diagnosis of SS, clinical signs, a blood smear with cerebriform atypical nuclei T cells, and evidence of T cell clones in the skin, lymph nodes and blood should be considered [4].

We report through a series of 5 cases, the experience of the hematology laboratory in the diagnosis of the syndrome of Sézary.

2. Material and Methods

This is a retrospective study of 9 cases of Sezary syndrome collected in the dermatology department of Marrakech.

The data collection was carried out from registers and patient files of the Dermatology Department. The patients were admitted in the Dermatology department. We have performed a blood sample for the purpose of complete blood count and blood smear with May-Grunwald-Giemsa (MGG) staining method. The blood analyse has been performed in the Military Hospital Inb Tofail in Marrakech – Morocco.

3. Results

Five patients were identified with the clinicopathological criteria of SS. There were 3 males and 2 females (M/F=1.25), mean age at diagnosis of SS was 62.1 years (52-73).

At the time of diagnosis, all 5 patients had erythroderma

and generalized lymphadenopathy in both superficial and deep stations.

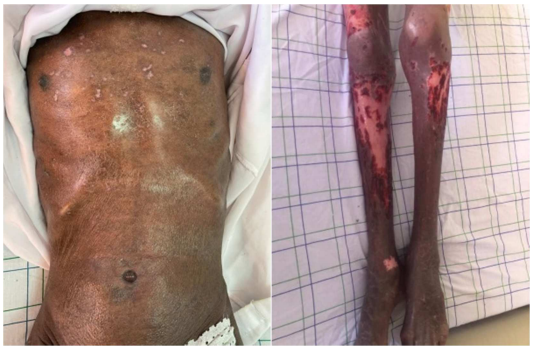

Figure 1. *Generalized erythroderma.*

The white blood cell count was elevated (>10,000 WBC/mm^3) in all 5 patients with a mean value of 18,120 WBC/mm^3.

The blood smear showed the presence of 75% (27 G/l) of small to medium-sized cells with a high nucleocytoplasmic ratio and cerebriform nuclei typical of Sezary cells and suggests the diagnosis of SS.

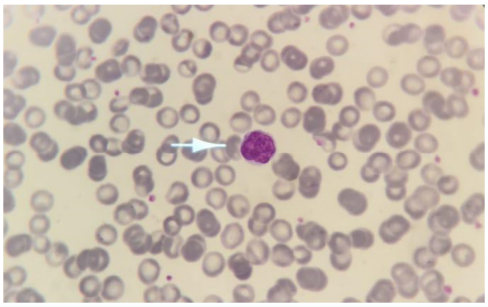

Figure 2. *Sezary cell on peripheral blood smear, MGG staining (x1000).*

Lymphocyte immunophenotyping was performed and showed a CD4/CD8 ratio greater than 10 in all cases.

Cytological examination of the skin biopsy showed malignant lymphocytic proliferation.

Thoracoabdominopelvic CT scan showed bilateral iliac lymphadenopathies.

4. Discussion

The first description of Sézary syndrome (SS) was by Sézary and Bouverain in 1938. They described a disseminated form of cutaneous lymphoma associating erythroderma and the presence of "monstrous" cells in the blood. [5]

Sézary syndrome (SS) is one of the cutaneous lymphomas whose pathophysiology remains poorly understood. It is a primary cutaneous T-cell lymphoma, which means that the skin is the first or only organ affected. These lymphomas are composed of memory T cells with cutaneous tropism, which is conferred by the expression on their surfaces of specific markers such as CLA (cutaneaous lymphocyte antigen) which interacts with the E-selectin receptor present in the post-capillary venules of the dermis. These mature T cells are usually recruited to the skin during skin infections to prepare a specific immune response. [6]

SS and MF are the most frequent forms of CTCL. It is a rare syndrome that represents between 3 and 5% of primary cutaneous T lymphomas. It generally affects adults aged 50 years on average. It is a rare and aggressive lymphoma, characterized by the triad of erythroderma, generalized polyadenopathy, and the presence of Sezary cells in the blood [7].

It is a primary cutaneous T-cell lymphoma that develops primarily in the skin, without extracutaneous involvement at diagnosis, i.e., no lymph node, visceral, or bone marrow involvement.

The ISCL/EORTC system has well determined the diagnosis of SS, thus requiring a rigorous clinicopathological and molecular cohesion. Its diagnosis is based on clinical, biological, and histological criteria. [8]

The biological workup includes a blood count, which is a non-specific test and most often shows normal hemoglobin, platelets, and leukocytes at the time of diagnosis.

Hyperlymphocytosis and hypereosinophilia may be present and are most often associated with a poor prognosis.

In patients with suggestive clinical signs, even a moderate lymphocytosis should prompt the diagnosis of SS. In some cases, signs of bone marrow involvement, such as normocytic normochromic anemia and severe cytopenia, may be found [9, 10], a CBC must be associated with a blood smear.

Sézary cells (SCs) have a characteristic morphology:

They are small to medium sized cells approximately 10 to 12 µm in diameter, with a high nucleo-cytoplasmic ratio, a very irregular nucleus with fairly dense, clear, mature chromatin without a nucleolus, and which has one to two "nail-bitten" grooves giving it a cerebriform appearance, sometimes described as a "wet sheet" appearance. [11, 12]

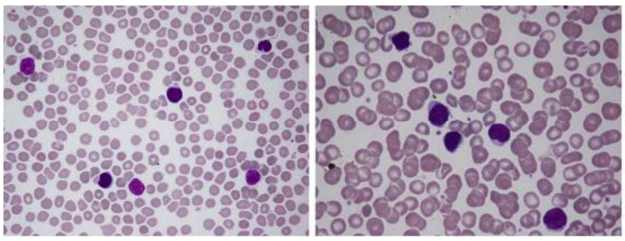

Figure 3. *Sézary cell on blood smear stained with May-Grünewald Giemsa [13].*

The blood smear distinguishes two types of Sezary cells:

1. Lutzner-Flandrin cells with a size of less than 12µm
2. Sezary-Lutzner cells whose size is greater than 12 µm

These 2 cell types have the same nuclear characteristics described above. The existence of large Sezary cells is the majority of cases related to a poor prognosis. [12]

A precise microscopic examination of the blood smear is essential for any patient presenting clinical signs suggestive of SS. Other more efficient techniques have been developed and allow a more objective diagnosis, in particular the immunophenotyping technique by flow cytometry [4, 15].

Immunophenotyping by CMF during SS allows to find several abnormalities that are part of the diagnostic criteria defined by the International Society of Cutaneous Lymphomas (ISCL) and the EORTC since 2007. There is a T cell hyperlymphocytosis (>1750 CD3+/uL lymphocytes) composed of cells with the following phenotype: CD2+, CD3+, CD4+, CD5+, CD8-, CD45RO+, CD45RA-, associated with the increase of CD4/CD8 ratio that is greater than 10 that was found in 48% to 88% of SS patients [16, 17].

5. Conclusion

The diagnosis of SS remains a challenge in many situations, the pathophysiology and definition of SS have evolved significantly over the past decades. Research continues to develop better management of this syndrome. The performed study showed the complementarity between biological analyse and clinic information in the process of SS diagnosis.

References

[1] Campbell JJ, Clark RA, Watanabe R, Kupper TS. Sezary syndrome and mycosis fungoides arise from distinct T-cell subsets: a biologic rationale for their distinct clinical behaviors. Blood. 5 août 2010; 116 (5): 767-71.

[2] Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, Swerdlow SH, et al. WHO-EORTC classification for cutaneous lymphomas. Blood. 15 mai 2005; 105 (10): 3768-85.

[3] Imam MH, Shenoy PJ, Flowers CR, Phillips A, Lechowicz MJ. Incidence and survival patterns of cutaneous T-cell lymphomas in the United States. Leuk Lymphoma. avr 2013; 54 (4): 752-9.

[4] Klemke CD, Booken N, Weiss C, Nicolay JP, Goerdt S, Felcht M, et al. Histopathological and immunophenotypical criteria for the diagnosis of Sézary syndrome in differentiation from other erythrodermic skin diseases: a European Organisation for Research and Treatment of Cancer (EORTC) Cutaneous Lymphoma Task Force Study of 97 cases. Br J Dermatol. juill 2015; 173 (1): 93-105.

[5] Hwang ST, Janik JE, Jaffe ES, Wilson WH. Mycosis fungoides and Sézary syndrome. The Lancet. 2008; 371 (9616): 945-57.

[6] Kim EJ, Hess S, Richardson SK, Newton S, Showe LC, Benoit BM, et al. Immunopathogenesis and therapy of cutaneous T cell lymphoma. J Clin Invest. avr 2005; 115 (4): 798-812.

[7] Willemze R, Kerl H, Sterry W, Berti E, Cerroni L, Chimenti S, et al. EORTC Classification for Primary Cutaneous Lymphomas: A Proposal From the Cutaneous Lymphoma Study Group of the European Organization for Research and Treatment of Cancer. Blood. 1 juill 1997; 90 (1): 354-71.

[8] Moins-Teisserenc H, Daubord M, Clave E, Douay C, Félix J, Marie-Cardine A, et al. CD158k is a reliable marker for diagnosis of Sézary syndrome and reveals an unprecedented heterogeneity of circulating malignant cells. J Invest Dermatol. janv 2015; 135 (1): 247-57.

[9] Moriarty B, Whittaker S. Diagnosis, prognosis and management of erythrodermic cutaneous T-cell lymphoma. Expert Rev Hematol. avr 2015; 8 (2): 159-71.

[10] Querfeld C, Rosen ST, Guitart J, Kuzel TM. The spectrum of cutaneous T-cell lymphomas: new insights into biology and therapy. Curr Opin Hematol. juill 2005; 12 (4): 273-8.

[11] Flandrin G, Brouet JC. The Sezary cell: cytologic, cytochemical, and immunologic studies. Mayo Clin Proc. août 1974; 49 (8): 575-83.

[12] Vonderheid EC, Sobel EL, Nowell PC, Finan JB, Helfrich MK, Whipple DS. Diagnostic and prognostic significance of Sézary cells in peripheral blood smears from patients with cutaneous T cell lymphoma. Blood. août 1985; 66 (2): 358-66.

[13] Kubica AW, Pittelkow MR. Sézary Syndrome. Surgical Pathology Clinics. juin 2014; 7 (2): 191-202.

[14] Olsen E, Vonderheid E, Pimpinelli N, Willemze R, Kim Y, Knobler R, et al. Revisions to the staging and classification of mycosis fungoides and Sezary syndrome: a proposal of the International Society for Cutaneous Lymphomas (ISCL) and the cutaneous lymphoma task force of the European Organization of Research and Treatment of Cancer (EORTC). Blood. 15 sept 2007; 110 (6): 1713-22.

[15] Hristov AC, Tejasvi T, Wilcox RA. Mycosis fungoides and Sézary syndrome: 2019 update on diagnosis, risk-stratification, and management. American Journal of Hematology. 2019; 94 (9): 1027-41.

[16] Rappl G, Muche JM, Abken H, Sterry W, Tilgen W, Ugurel S, et al. CD4(+)CD7(-) T cells compose the dominant T-cell clone in the peripheral blood of patients with Sézary syndrome. J Am Acad Dermatol. mars 2001; 44 (3): 456-61.

[17] Vonderheid EC, Bernengo MG. The Sézary syndrome: hematologic criteria. Hematol Oncol Clin North Am. déc 2003; 17 (6): 1367-89, viii.