

Case Report

Non-Hodgkin's Lymphoma (NHL) Associated to Systemic Lupus Erythematosus (SLE) at National Hospital of Niamey (Niger): A Case Report and Literature Review

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Abstract

Introduction: SLE is a well-known systemic autoimmune disease associated with an increased risk of malignancies, particularly lymphoma. The pathophysiological mechanism of non-Hodgkin's lymphoma in patients with SLE is not yet well understood.

Case Presentation: We report a new case of SLE in a 27-year-old female patient with a history of recently diagnosed non-Hodgkin's lymphoma (NHL). She presented an intermittent night-time fever accompanied by joint pain predominating in the large and small joints, without joint inflammation, progressive alopecia predominating at the hairline, malar erythema, mouth ulcerations, photosensitivity, Raynaud's phenomenon and a productive cough with mucous sputum. The diagnosis of Systemic Lupus Erythematosus was made on the basis of ACR criteria (7 out of 11) and SLICC.

Conclusion: SLE is often associated with non-Hodgkin's lymphoma (NHL), although the pathophysiological mechanism remains unexplained. The course of the disease is marked by phases of relapse and remission, with other metabolic and infectious complications compromising and complicating the clinical picture.

Keywords: Non-Hodgkin's lymphoma; Systemic Lupus Erythematosus; HNN-Niger

Introduction

Systemic lupus erythematosus is an autoimmune connective-tissue disorder with a wide range of clinical features, which predominantly affects women, especially from certain ethnic groups. Diagnosis is based on clinical assessment supported by investigations, including the finding of autoantibodies [1-4]. The clinical manifestations of Systemic Lupus Erythematosus (SLE) are frequent and polymorphic. SLE is a well-known systemic autoimmune disease associated with an increased risk of malignancies, particularly lymphoma [5-8]. The pathophysiological mechanism of non-Hodgkin's lymphoma in patients with SLE is not yet well understood.

Case Presentation

We report a new case of SLE in a 27-year-old female patient with

a history of recently diagnosed non-Hodgkin's lymphoma (NHL). She underwent a course of Cyclophosphamide-Hydroxydoxorubicin-Oncovin-Prednisolone (CHOP) chemotherapy.

After the courses of chemotherapy, the patient was in complete clinical remission, with no adenopathy, splenomegaly or hepatomegaly.

The 6-month course was marked by the progressive onset of intermittent night-time fever accompanied by joint pain predominating in the large and small joints, without joint inflammation, progressive alopecia predominating at the hairline, malar erythema, mouth ulcerations, photosensitivity, Raynaud's phenomenon and a productive cough with mucous sputum.

No neuropsychiatric or hematological disorders, indeed no cytopenias on blood count.

Radiological and sputum examinations ruled out pleurisy or pulmonary tuberculosis.

On laboratory examination, the patient had a frank inflammatory syndrome, lymphopenia and proteinuria of 0.3 g/24 hours.

The anti-nuclear antibody title was positive at 1/1280, with negative native anti-DNA and positive anti-Sm antibodies.

The diagnosis of Systemic Lupus Erythematosus was made on the basis of ACR criteria (7 out of 11) and SLICC.

After a three-day mini-bolus of corticoids, the patient was put on Hydroxychloroquine, with a favorable outcome.

There was complete remission of both NHL and lupus.

At 6 months, the patient was readmitted to the medical ward for a lupus relapse. She presented with fever, physical asthenia, chronic cough and mucous sputum.

Paraclinical investigations led to a diagnosis of slow adrenal insufficiency and pulmonary tuberculosis with positive bacilloscopy, positive sputum microscopy (BAAR++) and a Gene Xpert rt-PCR of sputum detected as moderately sensitive to rifampin.

Anti-tuberculosis treatment and hydrocortisone supplementation were instituted.

After two weeks of treatment, the patient was readmitted with respiratory distress, severe anemia and associated malaria. The patient died in this situation.

Discussion

This case report concerns a 27-year-old female patient with an association of NHL and SLE.

Many studies or clinical cases have reported the association of SLE and NHL in subjects of similar age. The majority of cases reported in the literature follow chemotherapy as part of the management of NHL, with no direct link between chemotherapy and the onset of SLE. This is not the case for our patient. In fact, many of the lymphoma cases were not exposed to any immunosuppressant medications before the onset of the malignancy [9-11].

On the other hand, there was a trend for a higher proportion of lymphoma cases being exposed to cyclophosphamide versus the cancer-free controls.

In addition, there is no clear relationship between SLE disease activity and lymphoma risk [12,13].

A priori, one might suspect that high disease activity in SLE, which is associated with heightened lymphocyte proliferation, might also heighten lymphoma risk. However, it is also known that the immune system has important roles in deleting abnormal cells. Multiple studies have demonstrated an increased risk for all hematologic malignancies in SLE, particularly for non-Hodgkin's lymphoma (NHL) [13-16]. A large multinational study involving 16,409 patients primarily from the Systemic Lupus International Collaborating Clinics (SLICC), the Canadian Network for Improved Outcomes in Systemic Lupus, and other international collaborators, demonstrated a slightly increased overall risk of malignancy in patients with SLE [12].

Therefore, an increased risk to develop B cell malignancies among SLE patients raise several questions, including: Do the female sex hormones (for example, estrogen) and increased levels of the type I IFNs contribute to an increased risk to develop B cell malignancies in SLE patients [16]

Conclusion

Although rare, SLE is often associated with non-Hodgkin's lymphoma (NHL), although the pathophysiological mechanism remains unexplained. The course of the disease is marked by phases of relapse and remission, with other metabolic and infectious complications compromising and complicating the clinical picture.

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