To Present a Case of the Disseminated Intravascular Large B-Cell Lymphoma Presenting as Fever of Unknown Origin

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Received date: August 03, 2021; **Accepted date:** August 17, 2021; **Published date:** August 24, 2021

Citation: Fidel K (2021) To Present a Case of Disseminated Intravascular Large B-Cell Lymphoma Presenting as Fever of Unknown Origin. J Cancer Diagn, Vol.5 Iss.4 No:1.

Abstract

Background: Intravascular large B-cell lymphoma (IVLBCL) is a rare type of non-Hodgkin's lymphoma (NHL) characterized by the selective growth of neoplastic cells within blood vessel lamina. The precise mechanisms responsible for this distinctive behavior are at the moment largely unknown. By the time of presentation, most patients have advanced, disseminated disease, and often the diagnosis is made at autopsy. Diagnosis requires skin, liver, lung, bone marrow, renal, meningeal, or brain vessel biopsy but is often made only when the illness has progressed or post mortem because early involvement of organs was not evident.

Discussion: We report a case of Intravascular lymphoma who presented as fever of unknown origin. In this case, initial laboratory test results were unremarkable. Computed Tomography of the chest and abdomen as well as bone marrow aspiration and biopsy were negative for malignancy. Patient developed neurologic symptoms and expired due to complications. Autopsy was done which revealed Disseminated Intravascular Diffuse Large B-cell Lymphoma.

Conclusion: Without treatment, intravascular lymphoma is rapidly fatal. Ante-mortem diagnosis is challenging and indefinable. A high index of suspicion followed by biopsy of the organs suspected to be involved, together with early institution of treatment are of utmost importance in approaching these kinds of patients.

Keywords: Disseminated;Intravascular; Lymphoma; B-Cell

Introduction

Intravascular lymphoma (IVL) is an uncommon subtype of lymphoproliferative disorder characterized by the proliferation of neoplastic cells within the lumen of small-caliber blood ves-sels [1]. This type of lymphoma was first reported in 1959 by Pfleger and Tappeiner in Germany as "angio-endothelioma-tosis proliferans systemisata" and was considered to be en-dothelial in origin .This disorder exhibits a life-threatening clinical course of a systemic disease, with predominant neurologic, hematologic, skin, bone marrow, and pulmonary involvement. The course and evolution are unfavorable due to aggressive behavior and late diagnosis. In recent years, the number of patients with IVL diagnosed antemortem has increased, mainly due to bet-ter knowledge of this disease [2]. The IVL diagnosis may be made by biopsies of compromised tissues or by random skin biopsy of visibly una ected skin [3].We describe the case of a 66-year-old white woman with IVL presenting as fever of unknown origin (FUO) of 1-year evolu-tion and a progressive

without neo-plastic cells.Suspecting systemic vasculitis with central nervous system (CNS) compromise, 1000 mg/d IV of methylprednisolone for 3 doses was indicated. However, the patient continued with fever and worsening hemiparesis and she developed dyspnea with hypoxemia; a thoracic CT scan was performed showing bilateral consolidative images. Suspecting infectious pneumonia, antibiotic treatment was started without improvement and progressive hypoxemia de-veloped. Cultures of blood, urine, and bronchoalveolar lavage fluid were negative.Due to progressive neurologic manifestations, pulmonary in-volvement, unremitting fever without evidence of infectious or immunologic disease, and persistently elevated serum LDH, an intravascular lymphoma (IVL) was suspected. To confirm this diagnosis, cutaneous random biopsies were made. These biopsies revealed atypical lymphocytes within the small ves-sels of the dermis and hypodermis. The immunophenotype was consistent with B cell phenotype, showing CD20, PAX5, and BCL2 expression and high proliferation index with Ki67 (80%) (Figures 3, 4).With the confirmed diagnosis of IVLBC, chemotherapy with R-CHOP was started. However, rituximab had to be withdrawn during the first infusion because the patient developed arteri-al hypotension and her hypoxemia worsened.The patient’s evolution was unfavorable, with respiratory in-su ciency and new neurological events, and she wntlJH