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Disseminated intravascular diffuse large B-cell lymphoma presenting as fever of unknown origin, diagnosed on autopsy

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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. Objective: To present a case of disseminated intravascular large B-cell lymphoma presenting as fever of unknown origin. Method & Result: 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. Intravascular lymphoma (IVL) is a rare lymphoproliferative disorder characterized by the proliferation of large B lymphoma cells within the lumen of small-caliber blood vessels. Clinical features are nonspecific, presenting as a systemic disease with fever and may be life-threatening. Antemortem diagnosis is difficult but may be made with biopsies of affected tissues or with random skin biopsies. 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 better knowledge of this disease [1–3].

The IVL diagnosis may be made by biopsies of compromised tissues or by random skin biopsy of visibly unaffected skin [4].

We describe the case of a 66-year-old white woman with IVL presenting as fever of unknown origin (FUO) of 1-year evolution and a progressive behavior with predominantly neurologic and pulmonary compromise

A 66-year-old woman was admitted for fever and left hemiparesis. One year before, she had FUO and pericardial effusion with a pericardial biopsy showing unspecified chronic pericarditis. The patient continued with recurrent fever in the last 12 months. One day before admission, she developed left hemiparesis and was admitted to our institution.

Physical examination on admission revealed fever (38–38.5°C), skin pallor, and mild left hemiparesis. No lymphadenopathy, hepatosplenomegaly, cardiac murmurs, pulmonary abnormal sounds, or cutaneous lesions were present.

Laboratory evaluations were: hemoglobin (Hb) 9.6 gr/dL with a mean corpuscular volume of 88 fland reticulocytes of 1%. White blood cells were 4.7×109/L (neutrophils 80%, lymphocytes 12%, and monocytes 8%) and platelet count 240×109/L. Serum C-reactive protein (CRP) was 8 mg/dl and the erythroid sedimentation rate was 129 mm/h. Serum AST and ALT was slightly elevated and serum lactic dehydrogenase (LDH) was severely elevated (1692 UI/L). Serum ferritin was 1650 mg/dl. The total serum protein was decreased, as were albumin and gammaglobulin, without paraprotein. Urinalysis was normal. Blood and urine cultures were negative. An HIV antibody test was negative, as were HBsAg, HCV, Epstein-Barr virus IgM, Huddleson test, VDRL, toxoplasmosis antibodies, cytomegalo-virus (CMV) antibodies, and CMV-polymerase chain reaction. The antinuclear antibody test was negative, as were anti-DNA antibody, antineutrophil cytoplasmic antibodies, rheumatoid factor, antiphospholipid antibodies, cryoglobulins, and serum complement.

Reference :

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2. 8. DiGiuseppe JA, Nelson WG, Seifter EJ, et al. Intravascular lymphomatosis: A clinicopathologic study of 10 cases and assessment of response to chemotherapy. J Clin Oncol. 1994;12:2573–79. [PubMed] [Google Scholar]
3. 9. Murase T, Nakamura S. An Asian variant of intravascular lymphomatosis: An updated review of malignant histiocytosis-like B-cell lymphoma. Leuk Lymphoma. 1999;33:459–73. [PubMed] [Google Scholar]

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