CASE REPORT

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Orbital lymphoma with isolated occult bilateral adrenal involvement: report of an extremely rare case

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Abstract

Orbital lymphoma is very rare malignant neoplasm, usually diagnosed in early stage of disease as primary lymphoma, but dissemination occurs in approximately 33% of cases. Isolated bilateral adrenal lymphomatous involvement is extremely rare, described in 0.83% of cases. We present autopsy case of a 63-year-old man with bilateral orbital diffuse large cell lymphoma, clinical stage IEA, successfully treated by one cycle of chemo- and radiotherapy, but after administration of the second cycle, the patient developed signs of gastrointestinal hemorrhage and died two months after the diagnosis. Autopsy findings exclude lymphoma involvement of any organ except histopathological infiltration of both adrenal glands without evidence of a mass lesion.

Keywords: extranodal lymphoma, orbital lymphoma, adrenal lymphoma, autopsy.

Introduction

Orbital lymphoma (OL) is very rare malignant neoplasm, comprising only 1% of all non-Hodgkin's lymphomas (NHL) [1] and 5–14% of all extra-nodal lymphomas [2]. It is usually diagnosed in early stage of disease as primary lymphoma, but dissemination occurs in approximately 33% of cases [2]. Bilateral OL is observed in 7% to 24% of patients [2, 3]. The predilection for site of extra-orbital disease was described: lymph nodes 29%, skin 13%, bone marrow 8%, spleen 6%, temporalis fossa 5%, salivary gland, sinuses and lung 4% each and other organs less than 3% [3]. Isolated bilateral adrenal involvement is extremely rare, described in 0.83% of cases [4].

In this article, we present autopsy case of a 63-year-old man with bilateral orbital diffuse large cell lymphoma, with isolated occult bilateral adrenal involvement.

Case presentation

A 63-year-old man with no previous history of chronic disease or malignancy presented with slowly enlarging painless lesions of both orbits, with proptosis and ocular adnexal soft tissue swelling with eyelid ptosis, proptosis and livid discoloration of both eyelids of eight months duration. These symptoms were associated with headaches and fatigue. His family history was noncontributory. He was admitted to Clinic for Ophthalmology for further diagnosis of an orbital lesion. A magnetic resonance imaging (MRI) scan of endocranium showed deep orbital tumor of the right side and deep orbital and eyelid infiltration of the left side, without extra-orbital involvement. Right chronic pansinusitis was also presented. Diagnosis was obtained by incisional biopsy of the left orbital tumor. A histopathological analysis of tumor showed diffuse and dense eyelid infiltration by uniform population of blue round neoplastic cells (Figure 1A). The neoplastic cells are middle-sized and large lymphoid cells with scant cytoplasm, large nuclei, with one to three peripherally positioned nucleoli (centroblasts and rare immunoblasts) with scattered small non-neoplastic lymphocytes (Figure 1B). A histopathological analysis with immunohistochemistry of the tumor specimen confirmed the presence of non-Hodgkin's lymphoma (NHL), diffuse large B-cell lymphoma (DLBCL), centroblastic, germinal center (GC) subtype, with CD20 (Figure 1C) and CD10 positivity (Figure 1D) and a moderate proliferative index (50% of tumor cells were Ki-67 positive) (Figure 1E). Tumor cells were negative for CD3, CD5, Bcl-2, Bcl-6 (Figure 1F), MUM-1 (Figure 1G), CD23, CD43 and cyclin D1.

The patient was admitted to Clinic for Hematology for staging and therapy purpose. The patient was afebrile, without "B" symptoms. Examination revealed *Eastern Cooperative Oncology Group* (ECOG) Performance Status 1, normal blood pressure, no lymphadenopathy and no hepatosplenomegaly. Electrocardiogram showed no signs of acute coronary syndrome or arrhythmia. Complete blood count was in the normal reference ranges (hemoglobin 141 g/L; 8.6×10^9 leukocytes/L; and 191×10^9 platelets/L). Other laboratory tests revealed the following: erythrocyte sedimentation rate 24 mm/h, fibrinogen 59.9 mg/dL, prothrombin time (PT) 107%, partial thromboplastin time (PTT) 24 s, C-reactive protein (CRP) 19.20 mg/L, β_2-microglobulin 3.92 mg/L, normal level of immunoglobulins (IgG 12.10 g/L, IgA 1.42 g/L, IgM 0.69 g/L), albumin 38 g/L,