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Oculo-Orbital Non-Hodgkin's Lymphoma: A Retrospective Study of 9 Patients

Ghorbel Asma*, Abidi Rim, Keskes Aicha, Yousfi Amani, Yahyaoui Safia, Zarraa Semia, Mahjoubi Khalil, Belaid Asma and Nasr Chiraz

Department of Radiotherapy, Salah-Azaiz Institute, Bab Saadoun, Tunis, Tunisia

Abstract

Background: Oculo-orbital non-Hodgkin lymphoma is a rare presentation of lymphoma. The objective of our study was to clarify the clinical features of non-Hodgkin's oculo-orbital malignant lymphoma and to establish the place of radiotherapy in the treatment.

Case: This is a retrospective study including 9 patients treated at the Salah Azaiez Institute between 2007 and 2018.

Results: There were 5 women for 4 men with an average age of 65 years. Four LMNHs were located on the right, three on the left and two bilateral. The time between the appearance of functional signs and the diagnosis was less than 6 months. The diagnosis is confirmed by biopsy, resulting in several histological varieties. A full extension assessment was performed in all patients. Muscle and/or optic nerve extension was noted in 6 patients and bone damage was observed in two patients. Six patients had chemotherapy. The 9 patients were treated with orbital radiotherapy with a dose that varied between 30.6 and 40 Gy. The duration of patient follow-up was from 2 months to 10 years. Seven patients were in complete remission. Two patients had insufficient follow-up and one patient died one month after the end of treatment.

Conclusion: The interest of radiotherapy in oculo-orbital LMNH has been clearly demonstrated in several studies, but its place, its methods and its tolerance profile remain to be defined in many situations.

Keywords: Oculo-orbital tumor • Malignant Non-Hodgkin's lymphoma • Chemotherapy • Radiotherapy

Introduction

Oculo-Orbital non-Hodgkin's lymphoma is a rare presentation that accounts for 1 to 2% of all lymphomas, 1 to 8% of non-Hodgkin's lymphomas, and 10% of extra-nodal non-Hodgkin lymphoma. But it remains the most common malignant orbital tumor in adults [1-3]. The objective of this study was to clarify the clinical and therapeutic particularities of this lymphoma and to establish the role of radiotherapy in the treatment of oculo-orbital non-Hodgkin's lymphomas.

Case Study

We conducted a retrospective study of oculo-orbital lymphomas diagnosed and treated at the Salah Azaiez Institute in Tunis between 2007 and 2018. The data collected were gender, age, different clinical and therapeutic characteristics as well as clinical evolution of patients.

Results

The charts of nine patients, four men and five women, were studied. The age at diagnosis ranged from 43 to 84 years with an average of 65 years. The symptomatology which led to the consultation was in the majority of cases (6 patients) the progressive appearance of a painless orbital mass. Exophthalmos, ulcerated lump with inflammatory signs and total and brutal blindness were also reported. The time between the appearance of functional signs and the anatomohistochemical diagnosis was less than 6 months. Four tumors were located on the right, three on the left and two cases of LMNH were bilateral. The diagnosis was confirmed by histologic biopsy with one large B cell lymphoma, one marginal zone B lymphoma, two small B cell lymphomas, one follicular B lymphoma, one NK type T lymphoma, two MALT lymphomas and one Coat lymphoma. All patients underwent CT scan and orbital MRI and a full extension evaluation. Muscle and / or optic nerve extension was noted in six patients, bone involvement was observed in two patients, and spinal cord infiltration occurred in one patient. Therapeutically, six patients received primary CHOP-type chemotherapy. All patients were treated with orbital radiotherapy with a dose ranging from 30.6 to 40 Gy at a fraction of 1.8 or 2 Gy per session. Average patient follow-up ranged from 2 months to 10 years. The evolution was marked by the total regression of the tumor in seven patients who were in complete clinical and radiological remission. However, some patients have retained post-radiation sequelae such as cataracts, optic neuropathy and cutaneous-mucous membrane toxicity. Two patients were lost to follow-up and one patient, in whom the disease was metastatic, died one month after the end of treatment, having a very poor general condition

Discussion

In our small series, the sex ratio was 0.8. In the literature, some publications also report a female preponderance and others an inverted sex ratio. These lymphomas mainly affect adults [4,5]. The average age at diagnosis was 60 years. This does not imply that the disease always spares young people. Recently, a 15-year-old patient was reported [6,7]. In our series, the symptomatology which led to the consultation was in most cases (6 patients) the progressive appearance of a painless orbital mass. Exophthalmos, total and brutal blindness were also reported. In the literature, it has been described that the clinical presentation is very variable, it generally depends on the site of development and the aggressiveness of the disease. Exophthalmos is the most frequent call sign in posterior development. Slow-growing, insidious lesions are often found, and biopsy can be difficult. This could explain the

***Address for Correspondence:** Ghorbel A, Department of Radiotherapy, Salah-Azaiz Institute, Bab Saadoun, Tunis, Tunisia, E-mail: ghorbelasma.med@gmail.com*

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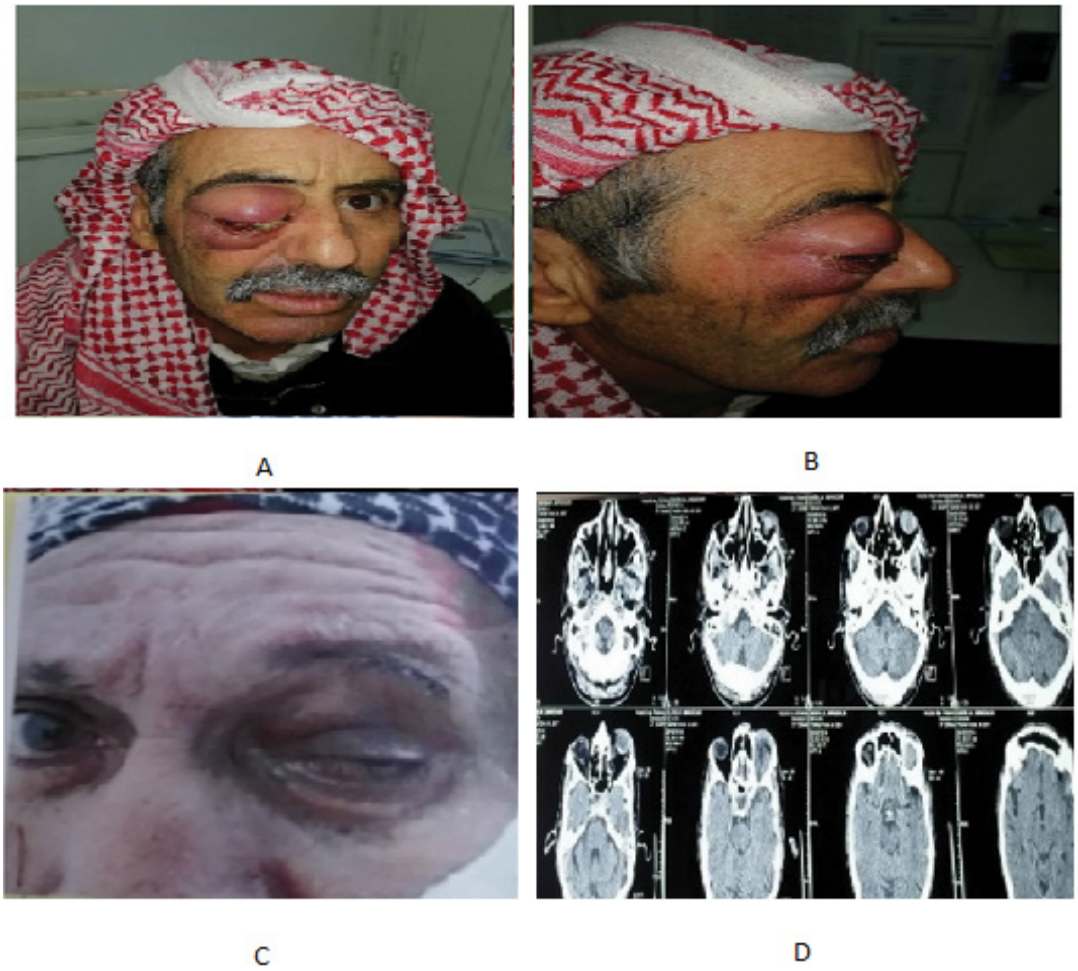

Figure 1. (A, B) NK lymphoma in a 53-year-old male patient with swelling, ulceration and erythema. (C) Extranodal lymphoma of the marginal zone in an 80-year-old woman presenting with swelling and facial asymmetry. (D) Computed Tomography (CT) in the same patient showing a diffuse mass in the inferior and anterior orbital region without bony destruction.

late diagnosis in several cases. The diagnosis is suspected based on clinical and radiological elements, but the confirmation is pathological by tumor biopsy [8]. The histological examination allows the lymphoma to be typed and helps in the choice of the therapeutic protocol. The first differential diagnosis is inflammatory pseudotumor [9]. This tumor disease is characterized by its histological diversity and its locoregional aggressiveness, making treatment difficult and mutilating. Among lymphomas of the orbit and adnexa, 30 to 50% present or will present a systemic lymphoma [10], hence the importance of the extension assessment which must be complete to guide treatment and predict the prognosis of each patient. The treatment depends on the aggressiveness of the tumor and the existence or not of a systemic lymphoma. For isolated low-grade lymphomas, radiotherapy alone at a dose of 30 Gy is indicated (Figure 1).

For symptomatic or high-grade forms but without distant metastasis, the treatment combines 3 cycles of induction chemotherapy of the CHOP type followed by radiotherapy of 30 Gy in 20 sessions in the event of a complete response to chemotherapy and 36 to 40 Gy in 20 sessions in case of partial response. In cases of lymphoma with systemic invasion, multidrug therapy or chemoradiotherapy may be indicated. Chemotherapy allows smaller doses of radiotherapy to be delivered. The benefit of radiotherapy has been demonstrated in several studies on the different histological types, but its place, its modalities and its safety profile remain to be defined in many situations. New radiotherapy techniques (IMRT, proton therapy) can help reduce toxicity in irradiated areas. Side effects of this treatment include corneal ulcers, retinal vasculopathy, optic neuropathy, and cataracts. Local control of orbital lymphomas by radiotherapy is excellent, ranging from 89 to 100%, with a relapse rate after radiotherapy alone of 9 to 20% [11], In our series, complete clinical and radiological remission at the expense of post-radiation toxicity was observed in seven patients. The prognosis depends on the histological features (type T poor prognosis), the extent of the tumor and the general condition of the patients. In our study, a patient died one month after the end of treatment, having very poor general condition.

Conclusion

Oculo-orbital non-Hodgkin's malignant lymphoma is characterized by its clinical polymorphism. Treatment is based on chemotherapy and radiotherapy. The interest of radiotherapy in oculo-orbital LMNH has been clearly demonstrated in several studies, but its place, its modalities and its tolerance profile have yet to be defined in many situations.

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