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Case Report

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Exophthalmos and Blindness Revealing a Non-Hodgkin's Malignant Lymphoma Type B with High Grade of Malignancy

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Abstract

Background: *Non-Hodgkin's lymphomas represent a heterogeneous group of haematological disorders characterized by a malignant monoclonal proliferation of the lymphoid system (B or T cells). Orbicatory lolisation is rare, it is seen mainly in adults.*

Case Report: *We report the case of a 79-year-old patient with a high-grade NHML, revealed by blindness and left exophthalmos evolving for 3 months. Ophthalmological examination revealed left blindness with axil exophthalmos grade 1. MRI cranio-orbital objectified a left orbital processe lesion intra and extra conical associated with a mucosal thickening of the cavum with left necrotic jugulocarotidian adenopathies. A biopsy of the cavum and ADP objectified a NHML of type B with high grade of malignancy.*

Result: *The patient was put on induction chemotherapy first before being placed on radiotherapy. The evolution was marked by the regression of the left exophthalmia.*

Conclusion: *The NHML is characterized by an extreme clinical polymorphism, especially in the cervicofacial localizations, make the diagnosis difficult and lead to a delay in management.*

Keywords: Exophthalmos, Blindness, Non-hodgkin's malignant lymphoma, Chimiotherapy, Radiotherapy, Orbit

Introduction

Non-Hodgkin's lymphoma is a heterogeneous group of hematologic disorders characterized by a malignant monoclonal proliferation of the lymphoid system (B or T cells) that tend to invade the entire body. The mode of revelation of the NHML is ganglionnaire in 2/3 of the cases and extra-ganglionnaire of 1/3 of the cases. The orbital location is rare, it is mainly seen in adults. The most common calling sign is exophthalmia.

The purpose of our work is to report the case of exophthalmitis and unilateral blindness revealing an NHML and to study the diagnostic, therapeutic and prognostic of this cancer.

Case report

We report the case of a 79-year-old patient with NHML with high grade of malignancy, revealed by blindness and left exophthalmos evolving for 3 months.

Ophthalmologic examination revealed left blindness with grade 1 axile exophthalmos, associated with a significant chemosis and purulent secretions and ophthalmoplegia (Figure 1).

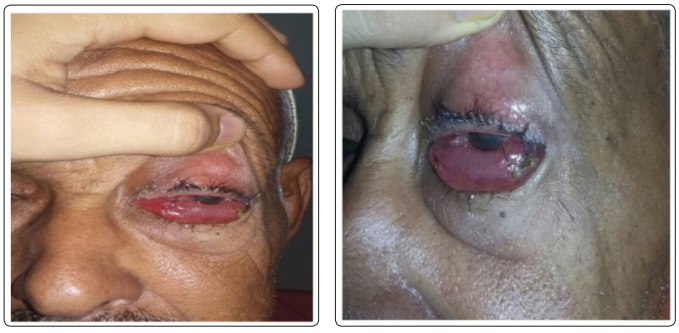

Figure 1: Left grade 1 exophthalmia and significant chemosis

Examination of the contralateral eye, found a visual acuity at 3/10 in the right eye, secondary to a senile cataract. Fundus examination revealed only diffuse chorioretinal atrophy in both eyes.

Initial cranio-orbital CT showed a left-handed retro-ocular process filling the retro-conical space with Grade 1 exophthalmos (Figure 2).

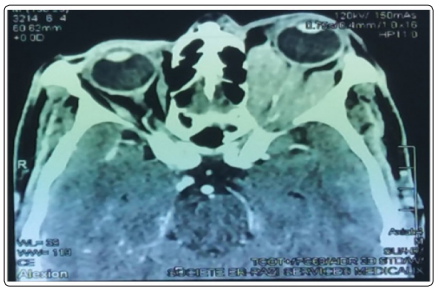

Figure 2: Cranio-orbital CT showing a left-handed retro-ocular process filling the retro-conical space with Grade 1 exophthalmos

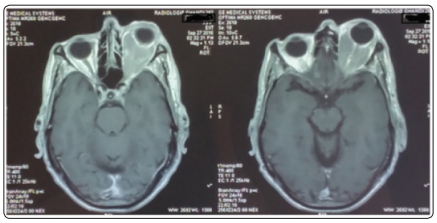

Figure 3: Cranio-orbital MRI showing an intra-orbital left orbital lesion process

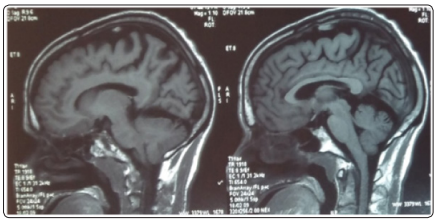

Figure 4: Mucosal thickening of the cavum with necrotic left jugulocarotidian adenopathies (ADP)

A biopsy of the cavum and ADP with anatomopathological study was carried out and objectified a non-Hodgkin malignant lymphoma type B with high grade of malignancy. The rest of the extension assessment was without particularity.

The patient was placed under poly induction chemotherapy session first before being placed on radiotherapy.

The evolution was marked by the regression of the left exophthalmia (Figure 5).

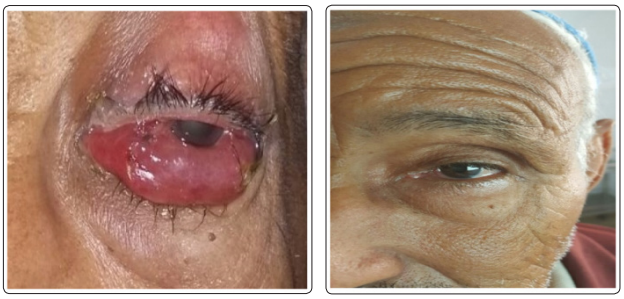

Figure 5: Regression of exophthalmia (right picture: before chemotherapy, left picture: after chemo-radiotherapy)

Discussion

The NHML is characterized by a clinical, morphological and biological diversity, that reflects the apparent complexity of modern histopathological classifications. Ophthalmological manifestations are rarely the revealing mode of these lymphomas (1.5%). The special interest in NHL is due to the increase in their incidence. A better knowledge of their development and the therapeutic progress can lead to a cure in a certain number of cases.

The clinical presentation is highly variable, depending mainly on the site of development and the aggressiveness of the disease. Diagnosis is suspected in clinical and radiological evidence, but it is confirmed only by histopathological examination. This histological examination help to type the lymphoma and choice the therapeutic protocol.

30 to 50% of patient with orbital lymphomas present or will present a systemic lymphoma, hence the interest of an extension assessment interesting the superficial and deep ganglionic areas, the bone marrow, the cerebro-spinal liquid in high-grade.

The diagnosis of lymphoma always results from the histopathological examination of the biopsy of tissue. Other examinations are only guiding. The histological examination will be carried out before any treatment likely to cause histological modifications hindering the interpretation of the cuts including a corticotherapic treatment. This examination can assert the malignant nature of lymphoid proliferation as well as the precise typing of the lymphoma which will be useful for the therapeutic decision.

The treatment depends on the degree of malignancy of the tumor, its aggressiveness and the existence or not of a systemic lymphoma:

- In the case of isolated low grade malignant orbital lymphoma: radiotherapy alone.
- In cases of isolated high grade malignant orbital lymphoma: 3 cycles of induction chemotherapy followed by radiotherapy.
- In the case of lymphoma with general invasion: multidrug therapy under the supervision of an oncologist.

Local control of orbital lymphomas by radiotherapy is excellent ranging from 89 to 100%. With a rate of distant metastasis of 0 to 25% in high grade malignancy lymphomas and 5% for low grade malignancy lymphomas.

Conclusion

The NHML is characterized by an extreme clinical polymorphism, especially in the cervicofacial locations, making the diagnosis difficult and lead to a delay in management; The diagnosis results from careful examination with biopsy of suspicious lesions with pathological evidence.

Its treatment is based on radiotherapy, which tends to be associated with chemotherapy, which seems to increase the rate of complete remission. The prognosis depends on several clinical and biological criteria, among which the Ann Arbor classification, the age, the LDH serum level, the general state, the number of extra-ganglionic sites reached [1-8].

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