exophthalmos revealing malignant lymphoma

Abstract:

Non-Hodgkin's lymphomas are malignant tumors whose aggressiveness depends on the histological type of the tumor. The orbital localization may be isolated or associated with a systemic lymphoma.

The clinical polymorphism of malignant lymphomas is extreme, posing diagnostic problems and delaying therapeutic management, which is why anatomopathological examination of a tissue biopsy is the only key to diagnosis.

We report the case of a patient who presented with exophthalmos revealing a lymphoma.

key words:exophthalmos - Non-Hodgkin's lymphoma.

Introduction:

Non-Hodgkin's lymphomas (NHL) are monoclonal malignant proliferations of B or T lymphoid cells at various stages of maturation. They usually develop in lymph node territories. Primary extra-nodal localization accounts for less than a third of cases. Orbital localization is rare, and mainly occurs in adults. The most frequent warning sign is exophthalmos. Malignancy is suspected on the basis of clinical and radiological criteria, but only histology confirms the diagnosis.

Clinical case:

This is a 9-year-old female patient with no specific pathological history who presented with a left exophthalmos that had been evolving for 20 days. Visual acuity was preserved, the anterior segment was normal and the FO showed macular folds. After 3 days, the exophthalmos progressed towards exaggeration and the appearance of local inflammatory signs (figure 1).

An orbito-cerebral CT scan revealed an intraconaltissue process in the left orbit measuring 34*21 mm, responsible for stage 3 exophthalmos with no overt signs of aggressiveness, suggestive of a possible lymphoma (figure 2).

Tumour biopsy and pathological study showed the presence of T-type NHL. The extension work-up was negative and the lymphoma was classified as stage I. Induction chemotherapy followed by radiotherapy was indicated after patient consent.

Discussion :

Orbital lymphomas account for 8% of orbital tumours [1] and less than 1% of all non-Hodgkin's lymphomas [2]. They may be isolated, as in our patient's case, or associated with a systemic lymphoma. Lymphomas of the orbit and adnexa affect the eyelids in 5-21% of cases, the conjunctiva in 21-33% or the orbit in 46-74% [2, 3].

These lymphomas mainly affect adults [4,1]. Clinical presentation is highly variable, depending above all on the site of development and the aggressiveness of the disease. Exophthalmos is the most frequent warning sign in cases of posterior development [5]. The diagnosis is suspected on the basis of clinical and radiological findings, but is only confirmed by anatomopathological examination of the tumour biopsy [4]. This histological examination also enables the lymphoma to be typed, thus helping in the choice of treatment protocol. Among lymphomas of the orbit and adnexa, 30-50% present or will present a systemic lymphoma [6,7,8], hence the interest of an extension work-up

involving the superficial and deep lymph node areas, the bone marrow, the cerebrospinal fluid in high-grade forms of malignancy, and other organs depending on the presenting signs [9]. Analysis of the results of the extension work-up and general work-up enables lymphoma to be classified according to the Ann Arbor classification, which will guide treatment and predict the prognosis of the disease.

Treatment depends on the degree of malignancy of the tumor, its aggressiveness and whether or not there is a systemic lymphoma:

- For isolated low-grade orbital lymphoma: radiotherapy alone (30 Gy in 20 sessions over 4 weeks).

- In case of isolated high-grade orbital lymphoma: 3 cycles of CHOP induction chemotherapy followed by radiotherapy of 30 Gy in 20 sessions in case of complete response to chemotherapy, and 36 to 40 Gy in 20 sessions in case of partial response.

- In the case of lymphoma with systemic invasion: polychemotherapy under the supervision of an oncologist.

In the case of aggressive lymphomas, a combination of chemotherapy and radiotherapy is indicated, given that the relapse rate is fairly high after radiotherapy alone (9 to 20%) and that the survival rate is better in the case of a combination. This chemotherapy makes it possible to deliver lower doses ofradiotherapy [2, 6, 10, 11, 7, 8].

Local control of orbital lymphomas by radiotherapy is excellent, ranging from 89% to 100%, with a distant metastasis rate of 0-25% for high-grade lymphomas and 5% of low-grade lymphomas [4].

The prognosis depends on the anatomopathological characteristics of the tumour (type T, poor prognosis), the size of the tumour mass (extension work-up+++) [12] and the patient's condition. However, this prognosis can be improved by early management [13].

Conclusion:

Orbital lymphoma is a tumour characterized by extreme clinical polymorphism, which can lead to misdiagnosis and delayed therapeutic management. Diagnosis is based on anatomopathological examination of the lesion. Treatment depends on the type and stage of the lymphoma, and includes radiotherapy preceded or not by induction chemotherapy in the case of stage I-II, and polychemotherapy in the case of systemic lymphoma.

figure 1: profile photograph of exophthalmos with inflammatory signs in the left eye.

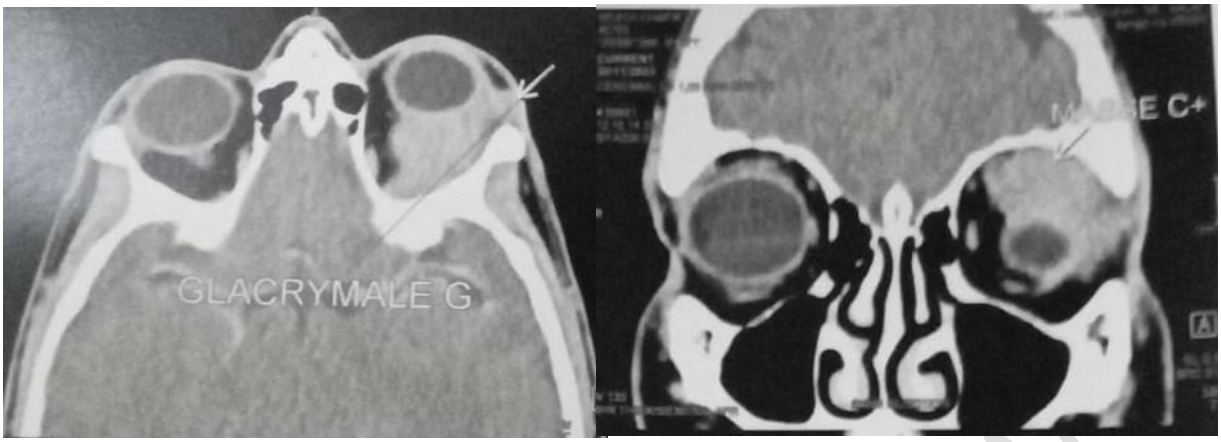

figure 2: scans showing an intraconical tissue process in the left orbit measuring 34*21 mm, responsible for stage 3 exophthalmos with no overt signs of aggressiveness.

Conflicts of interest:

All authors declare that they have no conflicts of interest

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