

Bilateral Exophthalmia, Revealing a Lymphoma not Hodgkinien of the Coat Type

Mouzari Y, Ould Hamed MA, Zerrouk R, Elasri F, Reda K and Oubaaz A

Ophthalmology Service, Military Hospital, Mohammed V University, Morocco.

*Correspondence:

Mouzari Y, Ophthalmology Service, Military Hospital, Mohammed V University, Morocco, E-mail: ys.mouzari@gmail.com.

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ABSTRACT

Introduction: The Non hodgkinien lymphoma (NHL) orbital location is a rare tumor, difficult to diagnose. The mantle cell lymphoma represents 6% of cases of LNH B. We report a patient of 60 years with bilateral exophthalmia revealing a mantle cell lymphoma.

Clinical Observation: A man of 60 years ran for ophthalmological emergencies in an array of orbital cellulitis of the right eye. This swelling was that day, painful, inflammatory, axile, irreducible, and non-pulsatile, with limited horizontal movement of the eyeball. In general terms, the patient has axillary lymph nodes and supraclavicular without splenomegaly. A biopsy of the orbital mass produced by trans-conjunctival, concluded in a non-Hodgkin lymphoma B type mantle (CD20 +, CD5 +, cyclin D1 +).

The assessment of extension realized in the Hematology Clinic department shows that this is a mantle cell lymphoma NHL B stage (nodes above and below the diaphragm, bilateral eyelid damage, spinal cord and location). After 3 treatments Rituximab-CHOP and DHAP-Rituximab treatments 3, obtaining a complete remission.

Discussion: The mantle cell lymphoma represents about 6% of NHL. Its incidence is estimated between 0.07 and three cases per 100 000 population per year. The geographical area and ethnicity shows a higher frequency in Caucasians than in African Americans. Ocular involvement in the NHL is a rare event, as evidenced by the limited number of cases reported in the literature. It may be opening that is the case of our patient, or alter the course of lymphoma known.

Conclusion: The eye location of mantle cell lymphoma is a rare entity. The clinical picture of this location can be misleading, particularly that of orbital cellulitis. The tumor biopsy confirms the diagnosis. Despite the treatment regimens, the prognosis of this type of lymphoma is reserved.

Keywords

Lymphoma B-type mantle, Bilateral exophthalmos, Non-Hodgkin lymphoma, Orbital cellulitis.

Introduction

Non-Hodgkin's lymphoma with orbital location is a rare tumor, difficult to diagnose. Coat-type B lymphoma accounts for 6% of B [1,2], NHL cases. It is characterized by a chromosomal translocation t (11; 14) (q13; 32) responsible for overexpression of cyclin D1.

We report the case of a 60-year-old patient with bilateral exophthalmos, revealing mantle-type orbital non-Hodgkin's lymphoma.

Observation

A 60-year-old man presented to ophthalmic emergencies in a table of bilateral orbital cellulitis with a sudden, larger installation on the right side. The interrogation finds a progressive evolution of an exophthalmia, without associated pain or redness, since about 2 years for the right eye and less than a year for the left eye. However,

the sudden appearance of significant pain at the level of the OD, 6 days ago, justified its consultation.

On inspection, there is a larger bilateral palpebral swelling (Figure 1) on the right, taking the entire upper eyelid, with a pseudo ptosis hiding the eyeball. This tumefaction was painful, inflammatory, axile, non-reducible, and non-pulsatile, with a limitation of the horizontal and vertical movements of the eyeball.



Figure 1: Bilateral palpebral swelling.

In the left eye, the swelling was of less volume, less painful and non-inflammatory, taking the inner third of the upper eyelid. The eye movements were, nevertheless, preserved on this side.

The visual acuity of the OD, after a difficult elevation of the upper eyelid, was 4/10. It is 7/10 in the left eye. The bio-microscopic examination finds on the right secretions at the level of the conjunctival cul-de-sacs, a diffuse conjunctival hyperemia which has no perkeric circle; the anterior chamber was optically empty. The ocular tone was 18mmHg; in the back of the eye, there is diffuse chorioretinal senile atrophy, with papillary excavation of 3/10. The bio-microscopic examination of the left eye was without particularities. In general, the spleno-ganglionic examination objectified the presence of axillary adenopathies, sup clavicular, without splenomegaly. The rest of the somatic examination was normal.

Given this clinical picture, the patient was put on broad-spectrum oral antibiotics: protected amoxicillin 1g3 times/day, and topical antibiotic eye drops. The biological assessment found hyper lymphocytosis at 6500/mm³, GB at 9300/mm³ with 16% circulating lymphomatous cells; there was no anemia or thrombocytopenia.

MRI performed (Figure 2), showed a bilateral soft tissues thickening under the palpebral tissues, predominant on the right, with a hyper signal T2 and a hypo signal T1 strongly raised after the injection of contrast product. This lesion process invades the right internal and external muscles on the left and the outer right. It extends bilaterally into intra-conical periorbital fat, encompassing both eyeballs without endocular invasion. It also engages both optical nerves and measures 13mm thick on the right and 8mm on the left.

A biopsy of the orbital mass, performed transconjunctivally, concluded in a small cell NHL B. The immunohistochemical study of tumor cells is in favor of mantle-type NHL (CD20 +, CD5 +, and cyclin D1 +).

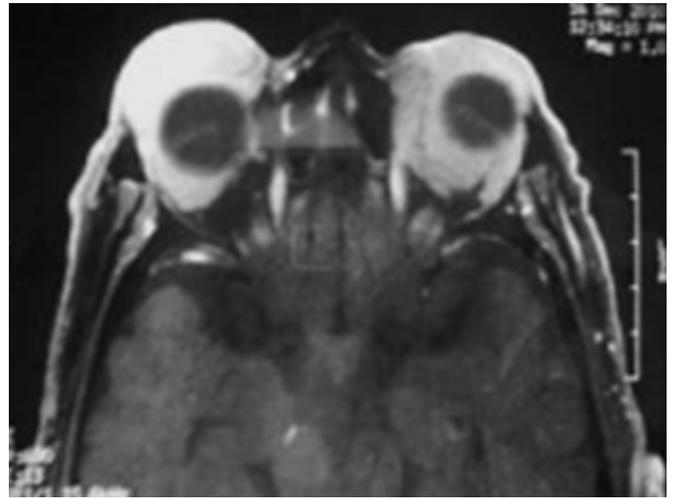


Figure 2: Initial cerebral orbito MRI.

Osteomedular biopsy showed bone marrow invasion.

The thoracoabdominal CT scan (Figures 3a and 3b) showed:

- Multiple mediastinal, axillary and supraclavicular lymphadenopathies.
- Large adenopathies of the stomachic coronary chain, hepatic and splenic, peri-pancreatic, coelio-mesenteric and inter-aortico-caves, iliac and inguinal hiles.
- Dense tissue nodules from the extra-pericardial anterior retrosternal space.

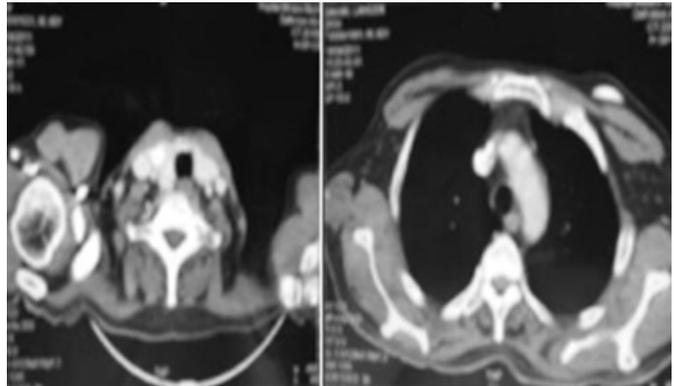


Figure 3a: Thoracoabdominal CT showing mediastinal jugulo-carotid adenopathies.

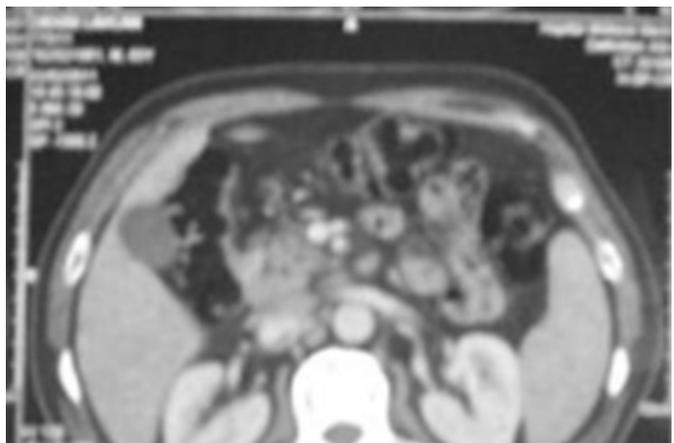


Figure 3b: Thoracoabdominal CT showing inter-aortic and cell-mediated lymphadenopathy.

In conclusion, our patient presents a B-type coat of bone at the stage IV medullary, revealed by a table of orbital cellulitis.

The evolution was spectacular with a clear clinical and radiological regression of the tumor on both sides and free ganglionic areas (Figures 4-6) after 3 courses of R-CHOP protocol (Rituximab, Cyclophosphamide, Adriamycin, Oncovin and Prednisone) performed 3 weeks apart in the Clinical Hematology Department. The reassessment balance is in favor of a complete remission.

Three consolidation courses R-DHAP type (Rituximab, Dexamethasone, Cytarabine, Cisplatin) were performed. The last ophthalmologic examination is normal, with a visual acuity of ODG corrected by 8/10, and P2 with addition to +250.



Figure 4: Regression of palpebral tumor after treatment.

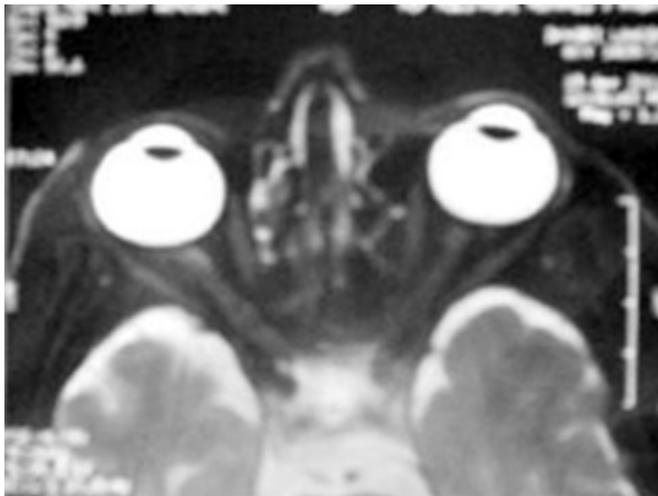


Figure 5: Cerebral orbito MRI after treatment.

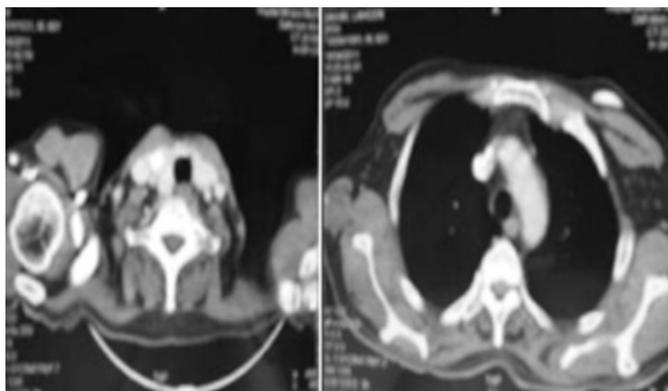


Figure 6: Thoracoabdominal CT after treatment showing disappearance of lymphadenopathy.

Discussion

Non-Hodgkin's lymphoma (NHL) accounts for 35% of all hematological malignancies. Mantle cell lymphoma (LCM) is a newly described entity [2-4]. Indeed, in the early 1990s, two authors described and characterized cytologically, then cytogenetically, a particular form of NHL whose tumor cell derives from the area of the mantle that will be at the origin of the term "mantle cell lymphoma" [5-7] This represents 6% of non-Hodgkin lymphoma cases.

The tumor cell derives from a so-called naïve B cell (cell that has not accessed the germinal center) and thus has not undergone the somatic mutation stage, and which originated in the mantle zone (zone surrounding the germinal center) [8].

This anatomico-clinical entity is currently recognized as a serious form of lymphoma, whose median survival is 3 to 4 years.

The tumor cell conventionally expresses a CD19 +, CD20 +, CD5 +, CCND1 +, CD10-, CD23-, Bcl-2 + phenotype with a surface immunoglobulin (most often IgM type). The other characteristic of Mantle Lymphoma is the presence of a reciprocal translocation between chromosomes 11 and 14. The t (11; 14) positions the CCND1 gene under the control of the gene coding for the immunoglobulin heavy chain. The consequence is the increase in the expression of the cyclin D1 protein, inducing the disruption of the cell cycle.

Mantle lymphoma accounts for about 6% of NHLs. The median age at diagnosis is 68 years with a male predominance (sex / ratio of 3 to 1) [9,10]. Its incidence is estimated to be between 0.07 and 3 cases per 100 000 population per year, with differences by age, geographical area and ethnicity showing greater frequency among Caucasians than among American blacks [9].

An increase in the incidence of LCM was noted between 1992 and 2004, which could suggest the existence of environmental factors favoring the appearance of LCM [10]. Ocular involvement in LMNH is a rare event as evidenced by the limited number of cases reported in the literature [10]. It may be inaugural, as is the case in our patient, or enamel the evolution of a known lymphoma. It most often concerns the orbit and, to a lesser degree, the conjunctiva, the eyelids and the lacrimal glands.

The blood count shows hyperlymphocytosis in 20 to 50% of patients, which is the case of our patient, but the presence of a circulating tumor population is more frequent (> 50%), which was also found in our patient. Anemia, classically normochromic normocytic and arterenative, is present in 40 to 50% of cases. It is most often moderate just like thrombocytopenia.

Our patient is classified as stage IV according to the classification of Ann Arbor, this is found in more than 80% of patients. Poly

chemotherapy provides about 60% of patients with a response, but it remains insufficient to achieve complete and prolonged remission. The contribution of Rituximab (monoclonal antibody directed against CD20) has precisely its interest to increase the rate of complete remission.

In the LCM, two randomized trials, conducted on a small sample, confirmed the benefit of Rituximab. The complete remission rate was 7% in the CHOP arm versus 34% in the R-CHOP arm. In the youngest patients whose age limit can be set at around 65 years, it is therefore recommended induction by poly chemotherapy, then consolidation by autograft.

Conclusion

Mantle lymphoma is a rare entity in NHL. Its ocular starting location is, again, more rare. The clinical manifestation of this localization can be misleading, especially that of orbital cellulitis. Tumor biopsy, guided by imaging results, confirms the diagnosis. Despite recently revised treatment regimens, the prognosis for this type of lymphoma remains unclear.

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