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Successful Use of Rituximab and Chlorambucil in a Patient with Primary Lacrimal Mantle Cell Lymphoma

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Abstract **Case Report**

Primary lacrimal mantle cell lymphoma is extremely rare and constitutes a challenge in diagnostic and therapeutic management. We report the case of a patient with a lacrimal localization of mantle cell lymphoma who responded well to a combination of immuno-chemotherapy.

Keywords: Rituximab, Chlorambucil, immuno-chemotherapy.

INTRODUCTION

Orbit lymphomas represent 10% of the extranodal locations of non-Hodgkin's lymphomas. We present a rare localization of mantle cell lymphoma (MCL) in the orbit.

CASE REPORTS

He is a 71-year-old patient with no specific medical history who presented with progressive progressive palpebral swelling in four months, without impaired visual acuity and with good general condition.

The clinical examination found visual acuity at 10/10. Upon inspection, there was a right palpebral ptosis. The motility test objectified a limitation of the elevation. With the slit lamp, there is a swelling of the right upper conjunctival fornices (Fig-1). The fundus was normal. The biological assessment showed a blood count, normal renal and hepatic functions.

The LDH level was normal and the beta 2-microglobulin raised to 3.60 mg / l (N: 0.7– 1.80).

Magnetic resonance imaging (MRI) revealed an increase in the volume of the lacrymal glands bilaterally and symmetrically, which are 9 mm thick on each side. The eyeballs are of normal morphology with respect for intra-orbital fats (Fig-2).

An anatomopathological examination of the biopsy of the lacrymal gland showed a diffuse proliferation of medium-sized lymphomatous cells, presenting a nucleus with mature chromatin and of cleaved outline.

Immunohistochemistry confirmed the diagnosis of LCM by showing CD5 + CD20 CD20 phenotype B cells negative for CD10 and CD23 and a 15% Ki67. Cyclin D1 immunostaining was positive (Fig 3a, 3b, 3c).

The cervico-thoraco-abdomino-pelvic CT scan did not show any deep lymphadenopathy. The bone marrow biopsy does not show lymphomatous infiltration.

In total, our patient had bilateral lacrimal stage IE (Ann Arbor classification) mantle cell lymphoma. It is an intermediate risk mantle cell lymphoma with an index of 6.1 according to the MIPI (Mantle cell lymphoma International Prognosis Index).

To date, the patient has received 6 courses of Rituximab-Chlorambucil poly chemotherapy. The evolution was marked by the achievement of a complete remission (Fig-4). Maintenance treatment with Rituximab is in progress.

Fig-1: Swelling of the upper right conjunctival sac

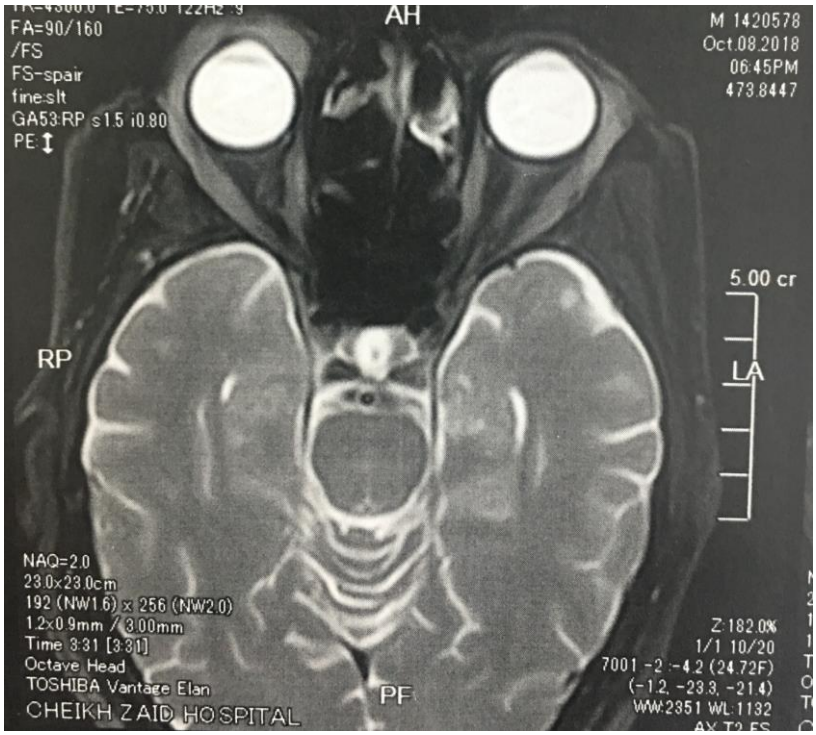

Fig-2: Orbitocerebral MRI: infiltration of the lacrimal glands

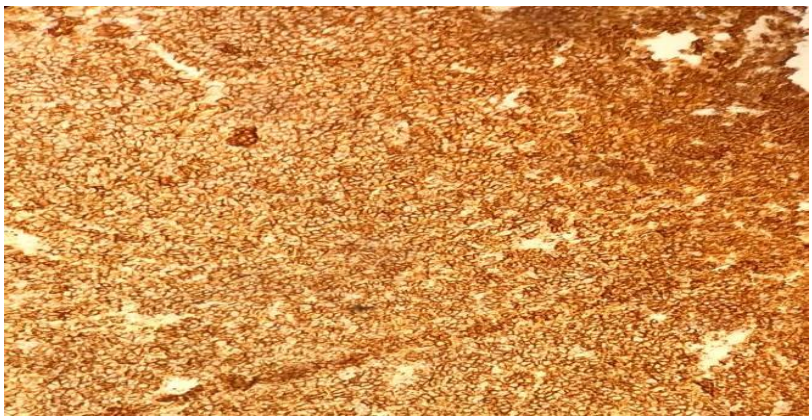

Fig-3: a) HE: View of average power of tumor infiltration by small monotonic lymphocytes. (hematoxylin-eosin, × 25)

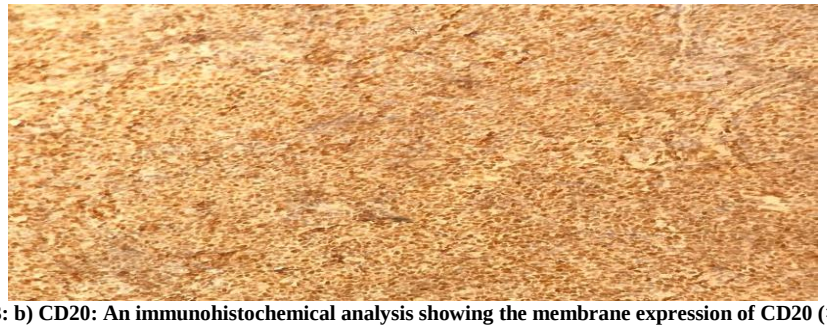

Fig-3: b) CD20: An immunohistochemical analysis showing the membrane expression of CD20 (× 40)

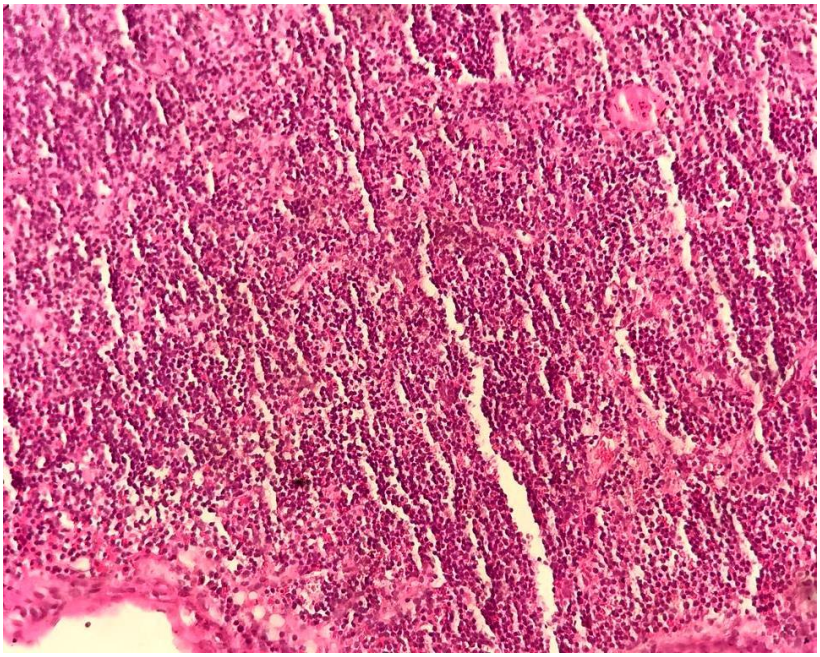

Fig-3: c) Cycin D1: An immunohistochemical result showing nuclear staining of cyclin D1 (BCL1) (× 40)

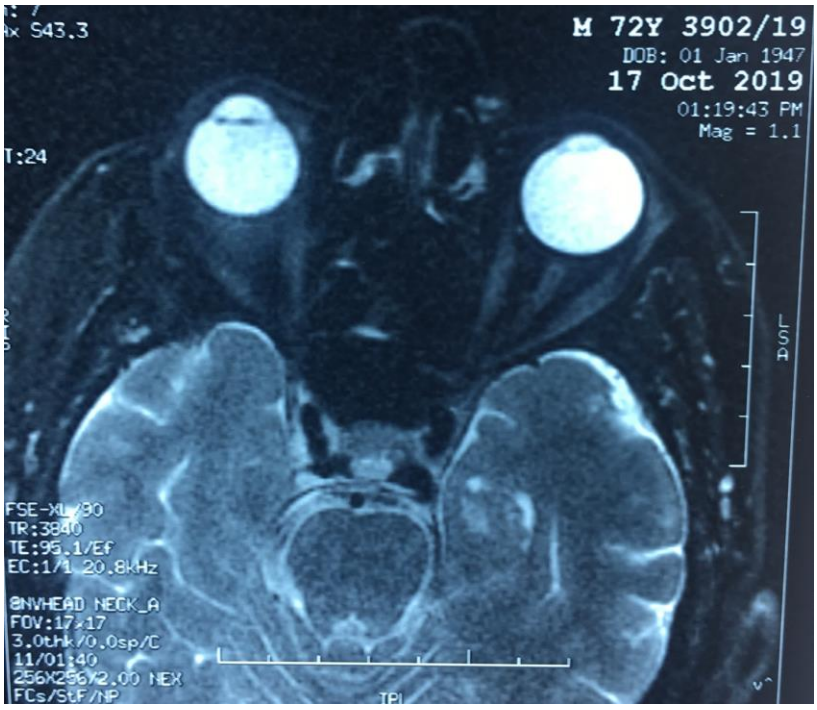

Fig-4: Orbital MRI showing complete remission

DISCUSSION

Mantle cell lymphoma (MCL) is a rare type of non-Hodgkin's lymphoma (NHL) that originates in B cells in the inner mantle area. It is characterized by chromosomal translocation t (11,14) (q13; q32), which leads to dysregulation and hyperexpression of BCL-1, the gene which codes for the protein cyclin D-1 [1, 2].

LCM accounts for approximately 9% of all NHL cases. The annual incidence of this subtype has increased in recent decades to 1 to 2 per 100,000 people [2].

It's a disease of the elderly (median age 71) and affects more men than women with a sex ration of 3 [1, 3].

The clinical course of LCM is generally aggressive; he usually presents a disseminated disease and characterized by frequent relapses and a low survival rate [1-3].

The diagnosis can be easy when there are other localizations on the other hand it is laborious in front of an isolated ophthalmological attack.

The ocular appendix LCM accounts for 2% to 7% of all non-Hodgkin's lymphomas [4, 5]. In the past, treatment was limited to palliative care. Which consisted mainly of chemotherapy including anthracyclines [6, 7].

In recent decades, new strategies combining chemotherapy such as cyclophosphamide, hydroxydoxorubucin, vincristine and prednisone (CHOP) with Rituximab (anti-CD20 monoclonal antibody) (R-CHOP) have shown improved response rates and rates of survival [8, 9].

For the elderly, various protocols combining Rituximab (R-thalidomide, R-Chlorambucil, R-Lenalidomide, R-Bortézomib-Dexamethasone) have achieved overall response rates of 50 to 92% [8].

CONCLUSION

Mantle cell lymphoma is currently a well-defined entity with rare orbital location. Immuno-chemotherapy seems to improve the prognosis of this pathology.

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