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Mantle Cell Lymphoma causing bilateral acute low vision

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INTRODUCTION

Mantle cell lymphoma (MCL) comprises 5% of non-Hodgkin lymphoma. In general, patients are typically Caucasian (about 2:1), male (about 2.5:1), and elderly (median age of onset, 68 years), and they usually present themselves as an extensive disease, including widespread lymphadenopathy, bone marrow involvement, splenomegaly, circulating tumor cells, and bowel infiltration.1 Central nervous system (CNS) involvement is an unusual form of extranodal involvement in the course of MCL. The diagnosis of CNS infiltration was based on clinical findings and the presence of malignant cells in cerebrospinal fluid (CSF), consistent with the diagnosis of MCL. CNS infiltration was more frequently observed in patients with blastoid histology and with Ki-67 expression ≥50% MCL is distinctive by having a poor prognosis. Patients with MCL must be treated aggressively if they can tolerate it. Survival has improved with the addition of rituximab to CHOP therapy, but median survival is still poor at 3–5 years. High mitotic rate and/or proliferation index are associated with poor prognosis and should be evaluated in each case.

We present a rare case of MCL with CNS involvement with bilateral acute low vision and bilateral neuritis. It is a rare case described in the literature.

CASE REPORT

Case of a 74 year-old male patient evaluated in the ophthalmology emergency room sector of São Paulo Hospital with ocular pain for 1 month and acute low vision in both eyes last week. The patient referred pain on eye movements and mild headache 1 month ago. He was diagnosed with lymphoma 8 months ago (Mantle Cell Lymphoma). He was treated with systemic and Intrathecal Chemotherapy for 4 months - last session 4 months ago.

At first evaluation, his best corrected visual acuity of the right eye vision was counting fingers at 1m and the left eye had no light perception. Anterior biomicroscopy was normal with intraocular pressure of 13 mmHg in both eyes, Pupillary Light Reflexes Direct and consensual 4+/4+. Extraocular motility with Direct and consensual 2+/2+ RAPD.

At retina exam (figure 1 and 2), both eyes presented pink optic disc with blurred margins, peripapillary hemorrhages, ingurgitated and tortuous vessels, normal macula and attached retina.

In the ER, screening for syphilis, HIV, toxoplasmosis (negatives) and general exams just a mild anemia (Hb=11,2).Besides this, a Brain, orbit CT and RM scan (image 3) were performed and evidenced with optic nerve thickening in intraorbital segments and densification of retrobulbar adipose tissue and rectification of the walls of the eyeballs. In addition, it was a cerebral spin fluid (CSF) and his immunohistochemical profile. - BCL-2: positive - BCL-6: positive in germinal centers - CD3: positive in paracortical zone - CD5: positive in 80% of lymphocytes - CD20: positive - CD30: negative - Ciclina D1: positive in frequent lymphocytes - *Ki-67: positive in 80% of lymphocytes.*

After some days the patient became a no light perception in both eyes and the conduct was to hospitalize and restart chemotherapy (systemic and intrathecal) with response evaluation through Cerebral Spinal Fluid control. After four applications of intrathecal chemotherapy, there was an improvement of CSF parameters (image 4) and visual acuity; OD: Hands Motion OS: Light Perception.

Unfortunately , the patient had sepsis(pulmonary focus) and died a few days (nadir of chemotherapy) after the treatment.

Material and methods:

It was a case of Mantle Cell Lymphoma with tumoral infiltration of CNS as a cause of bilateral acute low vision. The patient arrived from the emergency room of a university hospital (UNIFESP).

FIGURES

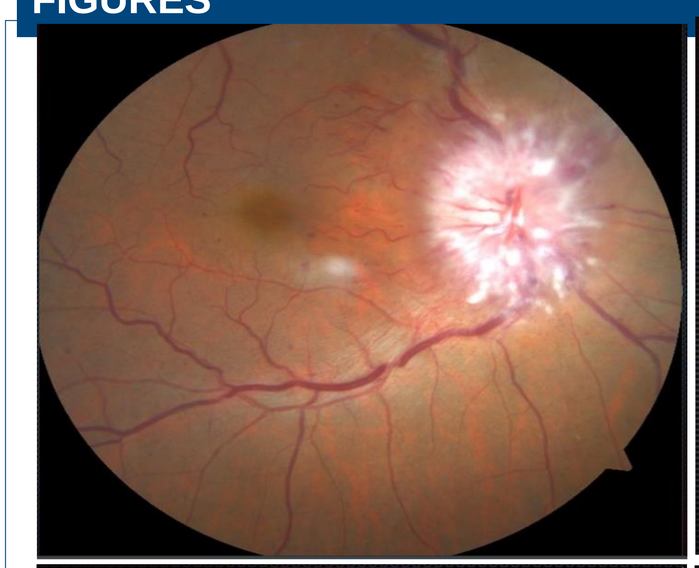

Image 1 (fundoscopy of right eye

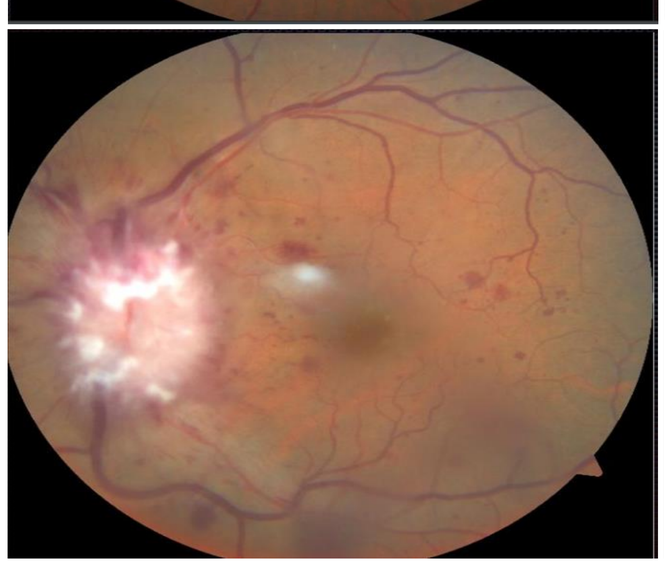

Image 2 (fundoscopy of left eye)

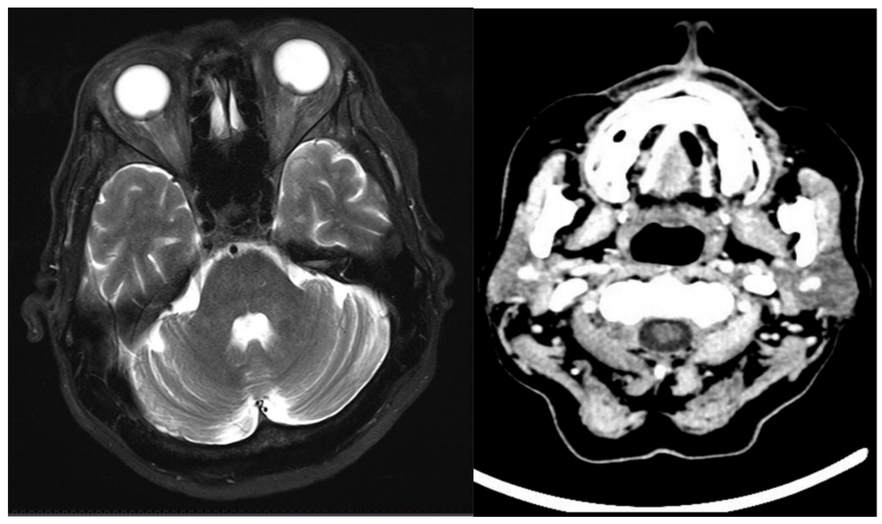

Image 3 (cranial RM and TC)

	White cell count	Diferencial
Reference range	0-4/ mm³	Lymphocytes (50-70) Monocytes (30 - 50%)
DO	238,6	Lymphocytes 87% Monocytes 12%
D4	22,0	Lymphocytes 83% Monocytes 7%
D7	8,3	Lymphocytes 58% Monocytes 15%
D11	0,3	Ø
D13	1,0	Ø

Image 4 (CSF parameters)

DISCUSSION AND CONCLUSION

Optic disc edema may be seen in a number of conditions; increased intracranial pressure (ICP), optic neuritis, tumoral causes (hematologic and metastasis cancer) ischemic optic neuropathy, toxic and nutritional optic neuropathies are possible etiologies. Our patient because of the bilateral impairment is mandatory to exclude ICP first. Metastasis can reach the optic nerve via the choroid, by vascular dissemination, by invasion from the orbit, and through the central nervous system.

Neuroimaging is crucial in patients suspected of infiltrative optic neuropathy due to cancer. Our patient had MRI findings an optic diffuse nerve enlargement that is more common signal in MRI.

MCL is a very aggressive subtype of non-Hodgkin lymphoma and is unique among lymphomas in its clinical, biologic, and genetic properties. Nearly 70% of cases are diagnosed in advanced stages of the disease and most cases show a relatively aggressive course. Median life expectancy ranges from 3 to 7 years. Because of its unresponsiveness to medical treatment as well as its aggressive nature, MCL is generally considered incurable.

REFERENCES

1-Knowles DM, Jakobiec FA, McNally L, Burke JS. Lymphoid hyperplasia and malignant lymphoma occurring in the ocular adnexa (orbit, conjunctiva, and eyelids): a prospective multiparametric analysis of 108 cases during 1977 to 1987. Hum Pathol. 1990 Sep;21(9):959–73.
2-White V, Rootman J, Quenville N, Worth A, Robertson W. Orbital lymphoproliferative and inflammatory lesions. Can J Ophthalmol. 1987 Dec;22(7):362–73.
3-Swerdlow SH. Lymphoma classification and the tools of our trade: an introduction to the 2012 USCAP Long Course. Mod Pathol. 2013 Jan;26(Suppl 1):S1–14.
4-Koban Y, Özlece H, Ayar O, et al. Mantle Cell Lymphoma Presenting with Acute Bilateral Ophthalmoplegia. *Turk J Ophthalmol*. 2017;47(4):235–237. doi:10.4274/tjo.80557