



# Exuberant primary cutaneous marginal zone B-cell lymphoma with orbital invasion

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## INTRODUCTION

Primary cutaneous marginal zone B-cell lymphoma (PCMZL) is the second most common cutaneous B-cell lymphoma. It usually presents as solitary or clustered plaques, nodules or tumors mostly on trunk, arms or face. It rarely exhibits extracutaneous involvement and have an excellent prognosis, with a 98% to 100% 5-year survival rate.

## CASE REPORT

A 63-year-old male presented red nodules on the right face that resolved after a short period of prednisone. Two years later, he started presenting proptosis and edema of the right orbit. The magnetic resonance showed expansive lesion posterior to the right orbit, involving all extrinsic ocular muscles and the optic nerve, causing elongation of the right eyeball and moderate proptosis. He was treated as an orbital pseudotumor with prednisone up to 60 mg with good response, but the lesion relapsed after every drug reduction. One year later, after the withdraw of the prednisone, he presented with red, deep-seated, tumoral lesions on the right temporal and right earlobe. Histology of the skin demonstrated centrocyte and centroblast types lymphocytes infiltration and immunohistochemistry showed positivity for PAX-5 and CD10 and negativity for BCL-2, compatible with primary cutaneous marginal zone B-cell lymphoma with posterior invasion of the orbit. The patient was treated with R-CHOP, with complete remission.

## CLINICAL FINDINGS

Fig.1. Proptosis and edema of the right eye; Fig. 2. Red, deep-seated, tumoral lesions on the right temporal and right earlobe.

Fig.3/4. Complete remission after 6 cycles of R-CHOP and 3 cycles of intrathecal chemotherapy.

## HISTOLOGICAL FINDINGS

Fig1. Diffuse lymphocytic infiltration of the dermis; Fig2. Diffuse lymphocytic infiltration of periorbital striated muscle; Fig.3. Atypical lymphocytes: irregularity of the nuclear contour and predominance of centrocyte-type lymphocytes, with cleaved nuclei.

Fig. 4. PAX-5 positive; Fig. 6. CD10 positive; Fig 7. BCL2 negative.

## DISCUSSION

Primary cutaneous marginal zone B-cell lymphoma is an indolent low-grade B-cell cutaneous lymphoma with a tendency to recur in the skin, but extracutaneous involvement is rarely seen. Nodal dissemination and a large cell transformation are the most common reported.

The largest case series from Hoefnagel et al reported fifty patients with PCMZL where only 2 patients presented extracutaneous spread. None of the patients died of the lymphoma.

Our patient presented orbital spread of the lymphoma with complete remission after six cycles of R-CHOP and 3 cycles of intrathecal chemotherapy.

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