

Exuberant primary cutaneous centrofollicular B lymphoma with orbital invasion

INTRODUCTION

B-cell lymphomas represent 25% of all primary cutaneous lymphomas and can only be diagnosed when there is no evidence of extracutaneous disease at initial presentation. Lesions tend to have indolent behaviour, with slow growth.

However we report here a case of exuberant presentation with local invasion of the orbit and complete remission after treatment.

CASE REPORT

A 53-year-old man was referred by ophthalmology clinic due to the appearance of nodular lesions on the right hemiface 2 months before with severe pain.

In the previous history, patient reported that 2 years previously he had been ongoing systemic corticosteroids every 3 months for similar, but smaller, lesions on the face interpreted as allergic dermatitis.

On examination, he presented with a tumorous, infiltrated and erythematous lesion of 3 cm in right zygomatic region, approximately 1 cm in right ear lobe and medial right epicanthus.

Presence of exophthalmos and significant right eyelid edema was noted as well as high myopia, ophthalmoplegia and ocular enanthema. There was no change in the pupillary reflex and no lymph node enlargement.

Histopathological and immunohistochemical studies showed neoplastic cells exhibiting predominantly centrocyte morphology and expression of CD20, PAX-5 and CD10. Most of them did not express Bcl-2. Aggregates of CD23 positive cells accompany the neoplastic infiltrate. The set of findings supports the diagnosis of centrofollicular lymphoma with a diffuse growth pattern.

Magnetic resonance imaging demonstrated expansive formation with signal of intermediate intensity on T1 and T2, in the retro and infralobar, intra and extraconal region. PET CT had no sign of visceral or lymph node involvement.

Due to evolution, associated with pathological and immunohistochemical profile, final diagnosis was of a primary orbital large B-cell lymphoma with local orbital invasion.

Patient underwent treatment with R-CHOP for 6 months and maintenance afterwards with cytarabine and methotrexate for 3 months, with complete remission of the disease.

DISCUSSION

Primary cutaneous centrofollicular lymphoma is considered an indolent neoplasm, representing around 60% of cases of primary cutaneous B-cell lymphomas. It occurs more frequently in men, with an average age of 50 years, exactly like our patient.

It can manifest as plaques, nodules or tumors, growing slowly and rarely ulcerating. They occur more in head and neck region, with multiple lesions occurring in 80% of patients.

On histopathological examination, weit is observed irregular nodules that resemble follicles or sheets of medium B cells with a germinal center phenotype. There is also a loss of polarization, with an imperceptible mantle zone.

Treatment can be carried out with radiotherapy, surgery, radiotherapy, rituximab or chemotherapy.

The reason for presenting this case is the exuberance of lesions, including orbital involvement, a behavior that is not typical of this type of tumor, as well as highlighting the importance of evolution to define the diagnosis between primary cutaneous versus systemic with secondary invasion of the skin.