Acta Clin Croat 2007; 46 (Suppl 1): 113-116 Case Report

PRIMARY ORBITAL NON-HODGKIN'S LYMPHOMA: CASE REPORT

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SUMMARY – An 81-year-old male presented with rapidly progressive loss of right eye vision, pain, mechanical ptosis, extraocular motility problems, and binocular diplopia. Orbital image obtained by computed tomography scanning detected right orbital tumor and cytological examination showed non-Hodgkin's lymphoma (small diffuse cells with low grade malignancy). The patient was treated with 12 cycles of chemotherapy (CHOP protocol) over a 2-year period, with good clinical response.

Key words: *primary orbital non-Hodgkin's lymphoma, low grade malignancy, binocular diplopia, cytology, chemotherapy*

Introduction

Primary orbital non-Hodgkin's lymphoma (NHL) is a rare presentation of extranodal non-Hodgkin's lymphoma accounting for less than 1% of cases (2 cases *per* million)[1]. It affects primarily the lacrimal glands, conjunctiva, eyelids and orbit[1,2]. Primary orbital non-Hodgkin's lymphoma affects all ages and is presented with symptoms of binocular diplopia, extraocular motility problems, proptosis, and mechanical ptosis from tumor pushing down the lid. It is characterized by gradual onset, slow progression and absence of pain[2,3].

Complete ophthalmologic and internist examination, ultrasound, computed tomography (CT) scan, cytologic and histologic examinations were used for diagnosis.

Case Report

An 81-year-old man presented with complaints of rapidly progressive loss of right eye vision, gradual redness of the right eye for several months, pain, restriction of eyeball movements, and diplopia. In May 2002, the patient was admitted to our department for complete work-up. Ophthalmologic examination revealed visual acuity of 0.3 on the right eye and 0.9 on the left eye, mechanical ptosis from tumor pushing the lid, conjunctival chemosis and corneal edema of the right eye, extraocular motility problems, elevation problems on the right eye with right hypotrophy on primary gaze, binocular diplopia, intraocular pressure of 28 mm Hg right

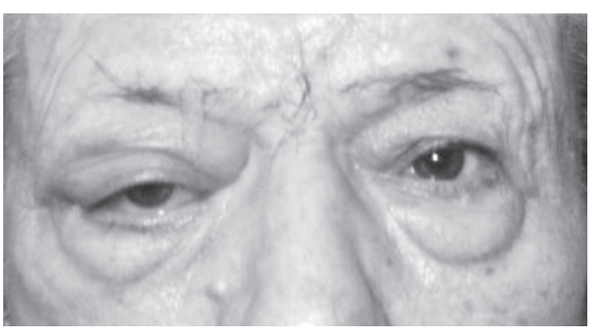

Fig 1. A 81- year old patient with primary orbital NHL before the treatment-ptosis, conjunctival chemosis, exophthalmus of the right eye.

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Received January 30, 2007, accepted March 26, 2007

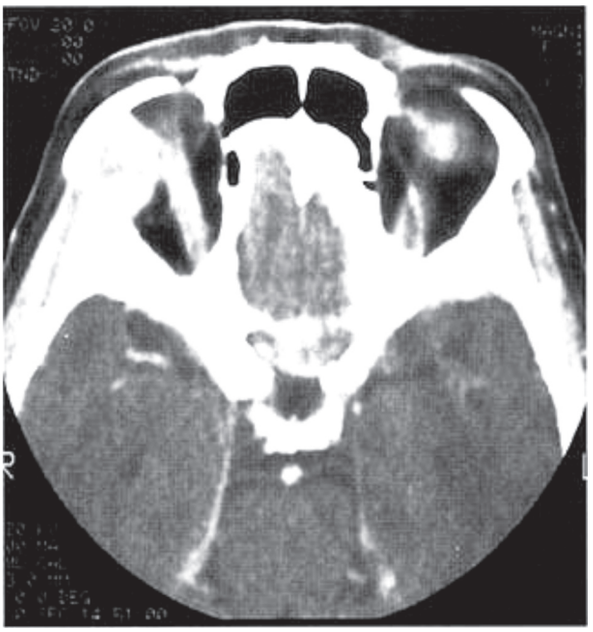

Fig 2. CT scan from 2002 y. of head and orbit shows right orbital tumor

eye/18 mm Hg left eye, Hertel (base 113) 20 mm right eye/16 mm left eye; retinal examination showed normal macula, vessels and periphery on both eyes.

Non-Hodgkin's lymphoma (small diffuse cells of low grade malignancy) was verified by cytology. Aspiration was performed with a 23-G needle through the right upper eyelid.

Internist examination revealed poor physical condition, hypertension, cardiopulmonary compensation; there was no organomegaly or lymph gland enlargement. Abdominal CT scan showed lymph node enlargement, with normal structures of all abdominal organs. Due to his poor physical condition and hypertensive heart failure, the patient was not a candidate for radical tumorectomy or radiotherapy. These two options were not possible to perform at our hospital and the patient was not capable to travel. The only solution available for his condition was systemic chemotherapy. In consultation with a haematologist, the patient was treated with chemotherapy according to the CHOP protocol (cyclophosphamide, doxorubicin (or Adriamycin), vincristine (or Oncovin), and prednisone). Clinical response was excellent; the tumor mass started to decline after chemotherapy and the patient's condition was good, showing cardiopulmonary compensation, with no pretibial edema or organomegaly, and lymph nodes stayed negative throughout the treatment period. After the 12th cycle of chemotherapy, there was no residual tumor and no metastases. The patient was followed up for 3 years, when he died from unrelated cause.

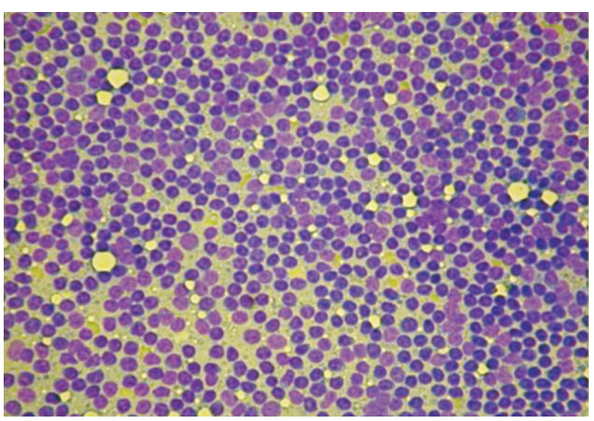

Fig 3. Diffuse small cell lymphoma. Low grade malignancy

Discussion

Ocular adnexal lymphomas represent the malignant end of the spectrum of lymphoproliferative lesions that occur in the conjunctiva, eyelids, lacrimal glands and orbit[1,5]. Orbital lymphoma is a rare presentation of extranodal non-Hodgkin's lymphoma.

On differential diagnosis of primary orbital non-Hodgkin's lymphoma, we have to take in consideration some more common orbital tumors seen in adults such as cavernous hemangioma, lymphoid lesions of the orbit (benign reactive lymphoid hyperplasia, lymphoma,

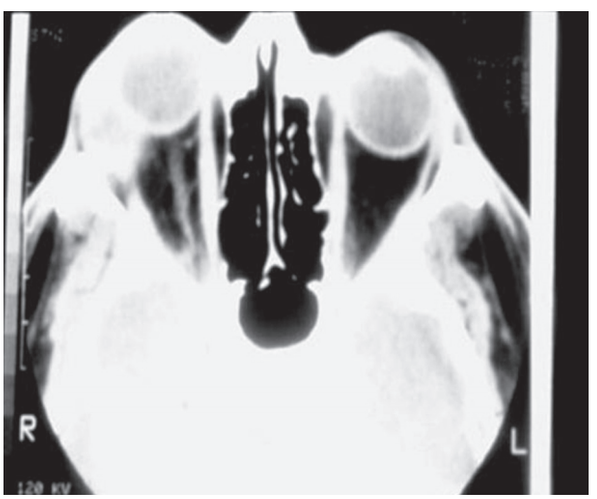

Fig 4. CT orbital image after 4- cycles of chemotherapy 2003y. – tumor mass is declining

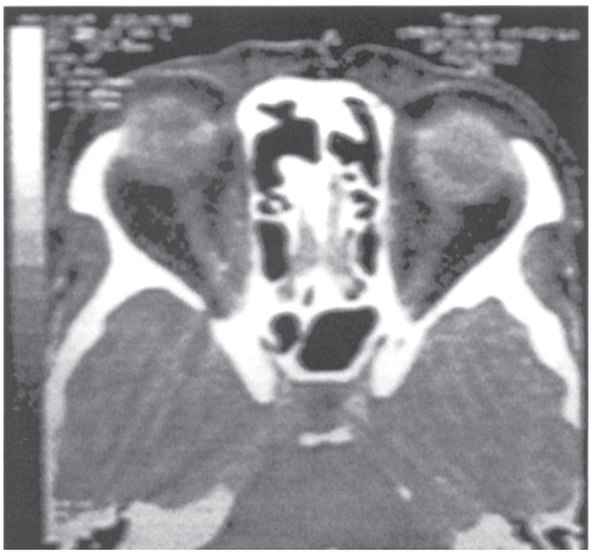

Fig 5. CT image after 12- cycles of chemotherapy 2005y. – there is no residual tumor left

atypical lymphoid hyperplasia), optic nerve meningioma, orbital metastasis, neurofibroma, neurilemoma (a.k.a. benign schwannoma), fibrous histiocytoma, hemangiopericytoma, lymphangioma, mucocele, and thyroid disease[2]. To confirm the diagnosis we need complete ophthalmologic and internist examination, ultrasound, CT orbital image, cytologic and histopathologic examination. It is rare to confirm the diagnosis only with cytologic examination as in our case; however, the patient's condition was so poor that we were not able to perform biopsy. Yet, the finding of diffuse small cells with low grade malignancy was so indicative of non-Hodgkin's lymphoma that we could rely on the finding and were able to start treatment with certainty in the diagnosis[6,7].

The treatment depends on the type and extension of tumor. If there is no systemic involvement, as in our patient, currently recommended therapy is radiotherapy[3,4,8,9], which we could not conduct due to the lack of radiotherapy at our hospital; disseminated disease is treated with chemotherapy[3,8,10].

Our experience showed that CHOP protocol has good tumor and clinical response. The patient was in good clinical condition throughout the period of chemotherapy and the tumor mass disappeared after the 12th cycle. The patient was followed up for three more years and there was no sign of tumor recurrence. Although usually demonstrating an indolent course, non-Hodgkin's lymphomas are known for recurrence at extranodal sites, including other ocular adnexal sites. Long-term follow-up with half-year examinations is therefore recommended[11]. The major prognostic criteria for ocular adnexal lymphomas include patient age, anatomical location of the tumor, stage of the disease at first presentation, serum lactate dehydrogenase level at the time of diagnosis, lymphoma subtype as determined according to WHO lymphoma classification, and tumor cell growth rate[7].

Although primary orbital non-Hodgkin's lymphoma is a rare diagnosis, it should be taken in consideration when finding orbital masses. In our patient, we demonstrated an unusual way of diagnosing and treatment that proved to yield good results.

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Sažetak

PRIMARNI NE-HODGKINOV LIMFOM ORBITE: PRIKAZ SLUČAJA

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Prikazuje se slučaj 81-godišnjeg bolesnika koji je primljen s brzo progredirajućim gubitkom vida na desnom oku, bolovima, mehaničkom ptozom, problemima u ekstraokularnom motilitetu i dvoslikama na oba oka. Kompjutorizirana tomografija orbite otkrila je tumor desne orbite, dok je citološka pretraga ukazala na ne-Hodgkinov limfom (difuzne male stanice niskog stupnja maligniteta). Bolesnik je primio 12 ciklusa kemoterapije (protokol CHOP) kroz dvije godine, uz dobar klinički odgovor.

Ključne riječi: *primarni ne-Hodgkinov limfom orbite, nizak stupanj maligniteta, obostrane dvoslike, citologija, kemoterapija*