Short communication

Orbital involvement by non-Hodgkin lymphoma NK T cells

A. Hervás-Ontiveros\textsuperscript{a,*}, E. España-Gregori\textsuperscript{a,b}, P. Hernández-Martínez\textsuperscript{a}, F.J. Vera-Sempere\textsuperscript{a,c}, M. Díaz-Llopis\textsuperscript{a,b}

\textsuperscript{a} Servicio de Oftalmología, Hospital Universitario La Fe, Valencia, Spain
\textsuperscript{b} Departamento de Cirugía, Unidad Docente de Oftalmología, Universidad de Valencia, Hospital Universitario La Fe, Valencia, Spain
\textsuperscript{c} Departamento de Anatomía Patológica, Universidad de Valencia, Hospital Universitario La Fe, Valencia, Spain

\textbf{A R T I C L E   I N F O}

Article history:
Received 6 October 2012
Accepted 28 October 2013
Available online 27 November 2014

Keywords:
Lymphoma
T-Cell/pathology
Eye neoplasms/diagnosis
Eye neoplasms/therapy
Exophthalmos
Natural killer T-cells/pathology

\textbf{A B S T R A C T}

Clinical case: The case is presented of 37-year-old male with a history of nasal obstruction with right rhinorrhea, headache, hearing loss and right exophthalmos of 4 months progression. The MRI revealed that the ethmoidal and maxillary sinuses contained inflammatory tissue extending into the orbital region. The biopsy confirmed a non-Hodgkin lymphoma of natural killer (NK) T cells.

Discussion: Non-Hodgkin’s T NK lymphoma is a rare tumour in the orbital area that requires an early detection and multi-disciplinary care to ensure appropriate monitoring and treatment.

© 2012 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.

\textbf{Afectación orbitaria por linfoma no Hodgkin de células T NK}

\textbf{R E S U M E N}

Caso clínico: Un varón de 37 años se presenta por una historia de obstrucción nasal con rinorrea derecha, cefalea, hipoacusia y exoftalmos derecho de 4 meses de evolución. La RM revela la ocupación de senos etmoidales y maxilares por tejido inflamatorio con extensión hacia la región orbitaria. La biopsia confirma un linfoma no Hodgkin de células T natural killer (NK).

Discusión: El linfoma no Hodgkin T NK es un tumour infrecuente en el área orbitaria que precisa de una precoz detección así como de una atención multidisciplinaria para asegurar un seguimiento y tratamiento adecuados.

© 2012 Sociedad Española de Oftalmología. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.


\*\* This case was presented in part as a communication during the 88th Congress of the SEO (Sociedad Española de Oftalmología [Spanish Society of Ophthalmology]), Barcelona, Spain, from 26 to 29 September 2013.

\* Corresponding author.
E-mail address: anheront@gmail.com (A. Hervás-Ontiveros).

2173-5794/$ – see front matter © 2012 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.
Introduction

Extranodal non-Hodgkin lymphomas are tumours usually derived from B-cells (65%) and, in most cases, they have a favourable prognosis.

Natural killer T (NKT) cell lymphomas account for 15% of all non-Hodgkin lymphomas and are located in the nasal fossae and maxillary sinuses. They occur between 50 and 60 years of age, preferably in male patients (2:1). They show an aggressive clinical course, defined by extensive tissue destruction and fatal prognosis, with a poor response to treatment. There is high association with the Epstein Barr virus (EBV). In this paper, we present the case of a patient with orbital involvement of NK T-cell non-Hodgkin lymphoma of nasal sinus origin.

Clinical case

A 37-year-old male patient who is admitted due to a 4-month history of bilateral nasal obstruction with right-sided rhinorrhoea, headache, impaired hearing and right-sided exophthalmos. His personal history includes asthma in treatment with salbutamol. Due to lack of improvement with antibiotic treatment, a sinus CT scan and a maxillofacial MRI were performed reporting diffuse occupation by homogenous inflammatory tissue of the maxillary and ethmoid sinuses next to the nasal fossae, more predominantly on the right side. This process extends towards the lacrimal sac and the pre-septal region of the right orbit (Fig. 1). As an atypical feature, there is a mass invasion of soft tissue from the medial side of the right orbit with thinness of the plane between the lamina papyracea and the internal rectus muscle.

A biopsy of the nasal fossae revealed a lymphoid infiltrate predominantly of T cells, plenty of mast cells and eosinophils with a local perivascular lymphocytic infiltrate of a destructive nature (Fig. 2A–D). The immunohistochemical analysis showed lymphocytic activity of T cells, with immature large cells, CD 117+, CD 45RO+, CD43+. The PCR was negative for EBV.

Fig. 1 – Coronal section of orbit CT scan showing a solid mass with erosion of the lamina papyracea and intraorbital component of soft tissue, fossa of lacrimal sac and right-sided pre-orbital septum region.

Fig. 2 – Histological sections on haematoxylin–eosin and CD3 from the sample obtained from the patient’s paranasal sinuses. (A) 10× section showing an infiltrate of lymphoid cells stained with haematoxylin–eosin and plenty of mast cells and eosinophils. (B) 20× section on haematoxylin–eosin showing those lymphoid cells with round nucleus, intermediate size and little cytoplasm. (C) 40× section on haematoxylin–eosin showing the lymphocytic infiltrate in greater detail. (D) 40× section showing CD3 staining.
The clinical findings and complementary tests led to a diagnosis of NK T-cell non-Hodgkin lymphoma and local radiotherapy treatment (RT) (initial dose of 35 Gy) was scheduled. A thoracic-abdominal-pelvic CT scan and a brain MRI were performed as complementary tests, which did not reveal any abnormal findings.

Cervical adenopathies were detected over the course of treatment and biopsies were performed on them. The tests showed clonal rearrangement of T cells with expression of CD43, CD2, CD7 and CD4>CD8. Under these conditions, intensive chemotherapy (ChT) was administered under a SMILE (steroids, methotrexate, ifosfamide, L-asparaginase, and etoposide) regimen and posterior RT was given in the facial region. By the fourth cycle, the patient presents periorbital oedema with hyperaemia and intense conjunctival chemosis that improves with topical antibiotic treatment. The patient shows favourable progress and a partial remission is seen in the CT scan (Fig. 3).

After one month, the patient showed progressive deterioration of his general condition associated with facial oedema and severe local pain, increased proptosis and reduced visual acuity. Upon examination, a right-sided orbital mass is palpated and palpebral malocclusion is noticed (Fig. 4).

The MRI/CT scan of the orbit revealed progression of the neoplastic process, involving the posterior inferomedial portion, and thickening of all the paranasal sinuses. The eyeball and the extrinsic muscles remained intact (Figs. 5 and 6). Upon progression, a new biopsy was performed showing recurrence of disease. Further intensive ChT treatment was administered under an ESHAP (etoposide, methylprednisolone, cytarabine and cisplatin) regimen.

An improved clinical picture is achieved with all the steps mentioned above, with decreased inflammation and periorbital oedema, and the neoplastic process is stabilized at the local level.

In the following months our patient experienced many exacerbation episodes in the setting of neutropenia and thrombocytopenia due to a neoplastic infiltration of the bone marrow. The patient died nine months after the initial diagnosis.

Discussion

The clinical case presented is of great interest for its rare nature and for compromising the patient’s life.

Fig. 3 – CT, coronal section: an apparent absence of mass is seen in the right orbital nasal area. Recovery of anatomical planes from the previously infiltrated right internal orbital portion.

Fig. 4 – Eye proptosis and intense chemosis on the right side that result in palpebral malocclusion indicating progression of the neoplastic process.

Fig. 5 – CT, coronal section: recurrence of the neoplastic process is seen with diffuse mucous thickening of all the paranasal sinuses.

Fig. 6 – T1-weighted MRI with contrast and fat suppression; axial section with posterior orbital compromise in its inferomedial part.
The NK T-cell lymphoma is characterized by the involvement of the structures in the midline of the face and may affect the orbit by contiguity. Coupland et al. described two cases of NK T-cell lymphomas with extension to the orbit. The differential diagnosis should include autoimmune, infectious and inflammatory processes, centrofacial ulcerative lesions and a great variety of tumours, both primary orbit tumours and metastatic ones. Within the diagnosis of extranodal lymphomas, the typical characteristics of its midfacial clinical presentation and the pathology enable a differential diagnosis versus the rest of lymphomas. On the other hand, even though its association with the EBV has been described, in our case the PCR was negative for that virus.

NK T-cell lymphomas are radiosensitive tumours with a poor response to ChT; local control of the disease is possible but recurrence is the norm in most of the published series. In our patient, RT was the initial treatment of choice due to the tumour location of the lesion. ChT was added afterwards upon occurrence of adenopathies.

Conflicts of interest

The authors declare that they do not have any conflicts of interest.

REFERENCES