Bilateral Orbital Mantle Cell Lymphoma

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Received: 03.11.2010 Accepted: 18.02.2011 A 75-year-old Moroccan man without a medical history of interest was admitted to our department for 10 months history of bilateral exophtalmia. He reported loss of the vision and weight loss of 15 Kg. The physical examination revealed pulsating bilateral exophtalmia (Figure 1), palpebral oedema, bilateral palpebral ptosis along a tumor in the region of the right cheek muscle, multiple enlarged cervical and inguinal lymph nodes. The spleen was not palpable and the patient was afebrile. Laboratory investigations showed haemoglobin of 8 (normal range 12-16) g/dL, mean cell corpuscular volume (MCV) of 83 (80-98) fL, white cell count of 3,900/L (4,000-10,000/L) and platelets of 13,000/L (150,000-300,000/L). Peripheral blood film was normal. Erythrocyte sedimentation rate was 14 (0-16) mm/hr and C-reactive protein was 5 (0-5) mg/L. The lactate dehydrogenase was 600 U/l (200-400). The serologies for Human Immunodeficiency virus (HIV), hepatitis B and C viruses were all negative. Computed tomography of the head showed a bilateral intra-orbital extraconal tumor without extension to the eyeballs (Figure 2). Histologic analysis of an inguinal lymph node showed mantle-cell lymphoma. A sample obtained on bone marrow biopsy also revealed infiltration by lymphoma. Chest-X ray, abdominal and pelvic ultrasound scans were normal. After initiating chemotherapy including cyclophosphamide, vincristine and prednisolone, the orbital tumor regressed significantly with loss of the vision but his condition deteriorated rapidly. The patient died afterward. Mantle cell lymphoma (MCL) is a unique lymphoma entity under the World Health Organization (WHO) classification of lymphomas, characterized by t (11;14) (q13;q32) and cyclin D1 over-expression (1). MCL represents 2.5-10% of all non-Hodgkin's lymphomas. It occurs in elderly patients with an average age of 60 years and predominantly in males. Seventy percent of patients present the disease in advanced stages. Its biological behaviour is very aggressive, with a mean survival of 3-4 years (2). Clinically, MCL is a lymphoma with multiple lymph-node involvement and also bone-marrow involvement. The clinical evolution of patients with MCL is relatively aggressive with a poor response to the treatment. In the orbital and adnexal region MCL represents 1-5% of all lymphomas (3). Clinicopathological reports

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Figure 1. Bilateral exophtalmia with ptosis

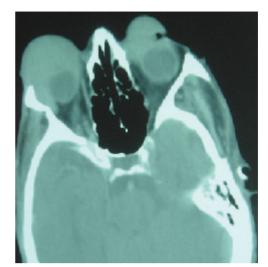


Figure 2. Head CT scan showed bilateral intra-orbital extraconal tumor

of MCL in this region have only been published in a few studies with a limited number of cases (4,5).Lymphoma should always be considered in patients with exophtalmia especially in the elderly population, to avoid a delay in diagnosis.

Key words: Mantle cell, lymphoma, orbit

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