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Orbital Localization Of A MALT-Type Lymphoma: A Case Report

M.Yousfi, Z.Hammoumi, G.Daghouj , L.Elmaaloum, B.Allali, A.Elkettani
Pediatric Ophthalmology Department, August 20, 1953 Hospital, Morocco

Abstract:
Lymphomas Of The Orbit And Periocular Tissues Are Rare Primary Orbital Tumors. The Most Common Histological Type Is MALT (Mucosal Associated Lymphoid Tissue) Lymphoma. It Is Usually Localized And Has A Good Prognosis. Ocular Involvement Is Rare, And Most Often Involves The Orbit, Conjunctiva And Lacrimal Glands. We Report Here The Case Of A Patient With Conjunctival MALT Lymphoma.
The Patient Was 49 Years Old, With No Previous Pathological History, And Presented To The Ophthalmological Emergency Department With Mild Palpebral Edema Of The Right Eye, Which Had Appeared One Month Prior To The Consultation. Ophthalmological Examination Of The Right Eye Revealed A Corrected Visual Acuity Of 10/10, While Examination Of The Adnexa Revealed A Firm, Salmon-Pink Nodular Lesion Developed At The Expense Of The Inferior Conjunctival Sac. Ocular Ultrasound Revealed A Well-Limited, Evenly Contoured, Heterogeneous, Oval Mass In The Inferolateral Soft Tissue Of The Right Eye, Vascularized By Doppler. The Conjunctival Mass Was Biopsied In The Operating Theatre Under Local Anaesthetic. Histological Examination Of The Biopsy Fragment With Immunohistochemical Study Revealed A Morphological And Phenotypic Appearance Of A Small-Cell B Lymphoma, Suggestive Of A B Lymphoma Of The Marginal Zone Of The Mucosa-Associated Lymphoid Tissue (MALT). The Patient Was Referred To The Haematology Department, Where She Received 6 Courses Of Intravenous Chemotherapy. The Conjunctival Mass Regressed Completely.

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I. Introduction:

Lymphomas of the orbit and periocular tissues are rare primary orbital tumors. The most frequent histological type is MALT (Mucosal Associated Lymphoid Tissue), an extra-ganglionic B lymphoma of the marginal zone, the lymphoid tissues associated with the mucosa. It is usually localized and has a good prognosis, characterized by curability and slow progression with non-aggressive treatments, which does not exclude regular long-term follow-up. [1][2] Ocular involvement is rare, and most often involves the orbit, conjunctiva and lacrimal glands [3].

II. Clinical case:

A 49-year-old female patient with no previous pathological history presented to the ophthalmological emergency department with mild palpebral edema of the right eye, which had appeared one month prior to the consultation.

Ophthalmological examination of the right eye revealed a corrected visual acuity of 10/10, while examination of the adnexa revealed a firm, salmon-pink nodular lesion developed at the expense of the inferior conjunctival sac **(Photo 1**). The rest of the examination was unremarkable, and the ophthalmological examination of the left eye was normal.

Photo 1: nodular lesion of the salmon-pink inferior conjunctival CDS (right eye).

Ocular ultrasound revealed a well-limited, evenly contoured, heterogeneous, oval, Doppler-vascularized, inferolateral soft tissue mass measuring 1.83*1.31 cm in the right eye **(Image A)**, and no abnormalities in the left eye **(Image B)**.

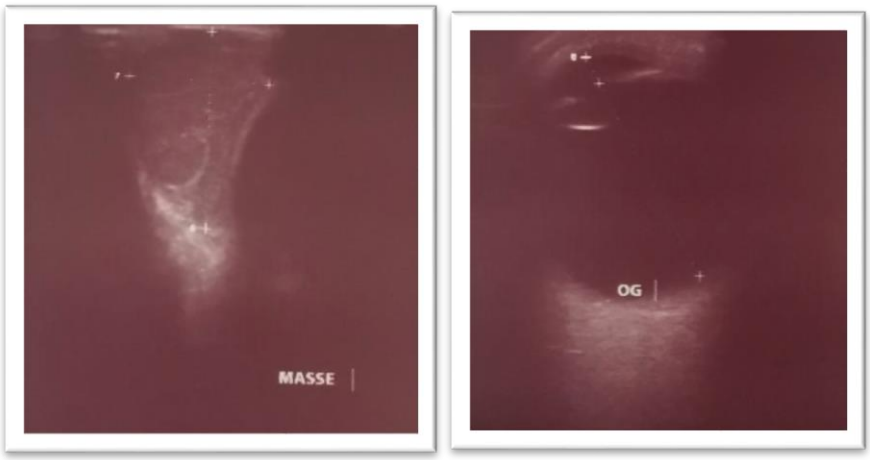

Image A **Image B**

A biopsy of the conjunctival mass was performed in the operating room under local anesthetic. Histological examination of the biopsy fragment of the conjunctival mucosa revealed a chorion massively infiltrated by a diffuse, vaguely nodular, malignant lymphoid tumor proliferation. Immunohistochemistry revealed a morphological and phenotypic appearance of a small-cell B lymphoma, suggesting a B lymphoma of the marginal zone of the mucosa-associated lymphoid tissue (MALT).

The patient was referred to the hematology department, where she received 6 courses of intravenous chemotherapy.

Progression was marked by total regression of the conjunctival mass.

III. Discussion:

Orbital and conjunctival localizations of MALT lymphomas are uncommon. In the two largest series available, orbital and conjunctival involvement accounted for 13 and 18 cases respectively out of 180 cases, and 7 and 1 case respectively out of 73 cases [4], [5]. Other ocular locations include orbital connective tissue, eyelids and lacrimal glands. In patients with conjunctival localization, the symptoms described in the literature are non-specific and discreet, ranging from photophobia to moderate ocular redness associated with ocular irritation. Bilateral involvement is frequent (15-23%), independently of any distant dissemination, which has been explained by a "homing" phenomenon of mucosal lymphocytes. Some authors had suggested that MALT might represent a normal component of the human conjunctival fornix [6], [7]. This hypothesis has been invalidated by other authors, who explain the acquisition of MALT during life as a consequence of external antigenic stimulation or a specific infectious reaction. MALT was thus found in 30 patients from 88 with conjunctival post-mortem samples [7]. For a long time, there was confusion between proliferations reacting to an infectious stimulus of the eyelids (chlamydia, Epstein-Barr virus, adenovirus...) and lymphoid proliferations. Histological and immunohistochemical analysis is currently used to clarify the difference.

Classically, MALT-type lymphomas have a good prognosis, with a tendency to remain localized. Bilateral lesions may be a factor in recurrence [8]. MALT-type lymphomas can occur simultaneously in several anatomical sites, and these multiple localizations appear to be more frequent in extradigestive MALT-type lymphomas [8].

The current treatment approach most often adopted for MALT lymphomas strictly localized to the orbit is radiotherapy, with recommended doses of 25 to 35 Gy in 10 to 15 fractions of 1.5 - 1.8 Gy, delivered over 2 or 3 weeks. The only side effects described at these doses are a mild dry syndrome and, more rarely, cataracts [9].

In cases where radiotherapy is contraindicated or refused by the patient, the therapeutic alternatives are surgery and/or chemotherapy. Chemotherapy gives good results, but its toxicity limits its use in view of the good results obtained with radiotherapy alone; surgery alone appears to be inadequate and prone to relapse [10], [11].

IV. Conclusion:

Orbital and conjunctival localizations of MALT-type lymphomas are uncommon. Clinical appearance is often suggestive, but biopsy should always be proposed to confirm the diagnosis. The most common treatment for MALT-type lymphomas strictly localized to the orbit is radiotherapy.

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