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Physiology and Anatomy

Elixir Physio. & Anatomy 124 (2018) 52100-52101

ISSN: 2229-712X

A superior eyelid and Orbit localisation of Malt Lymphoma. A Case Report at the Clinical Hématology Department of Yopougon National Hospital. (Abidjan–Cote d'ivoire)

Packo S, Danho NC, Meite N, Kouakou B, Botti RP, Kamara I , Silue DA, Bokoum S and Sanogo I

ARTICLE INFO

Article history:
Received: 17 September 2018;
Received in revised form:
20 October 2018;
Accepted: 01 November 2018;

Keywords
Malt lymphoma,
Ocular.

ABSTRACT

Non-Hodgkin's malignant lymphomas are malignant haemopathies characterized with clinical and anatomy diversity. Mycosis associated lymphoma tissue (MALT) is part of marginal zone B-cell lymphoma. The stomach is the most common site. We report in this study, a case of a 57 year old woman with no particular médical history who presented a right superior eyelid and orbit tumor with the exophthalmos. Context of fever noted in the progression. The immunohistochemistry of the biopsy of this tumor had concluded with a MALT lymphoma. Nowadays such a case has seldom been published. After using Rituximab, cyclophosphamide, vincristine and prednisone, all the tumefactions disappeared. This case was interesting because of the rarity localisation of this lymphoma, and shows the interest of a first biopsy before any surgical excision of tumor.

1. Introduction

MALT lymphoma is a form of extra nodal marginal zone lymphomas of lymphoid tissue whose cells are arising of B lymphocytes and develop in lymphoid tissue associated mucosa [1, 2]. The localizations are mostly associated with microbial pathogens. Helicobacter pylori is associated with gastric MALT lymphomas; Borrelia burgdorferi for cutaneous MALT lymphomas; Campylobacter jejuni for intestinal MALT lymphoma and Chlamydia psittaci for ocular appendages MALT lymphoma [3, 4, 5].

Epidemiologically, it's the most frequent of extra nodal marginal zone lymphoma with frequency between 50% and 70% and represents 5-8% of all B-cell lymphomas. It would be mainly the infection of old subject to 60-year-old [6, 7, 8, 9]. Stomach is the most common site of MALT. Other sites are the rares disease [2]. The ocular appendages site is rarely described. We report in our study a case of ocular appendices MALT lymphoma, which for our knowledge constitutes the first of the kind described in our department.

2. Observation

Mrs. I.L 57 year old, with no particular pathological history was referred to us by the ophthalmology department of the "Polyclinic Farah" of Abidjan for ocular lymphoma. According to the Anamnesis the symptom started since 2015 to Nigeria by the occurrence of a headache of increasing intensity without triggering factor, or sedation. It was accompanied by an ocular tumor with increasing in volume, obstructing the eye gradually. She consulted the ophthalmology department of "polyclinique Farah" where aye's tomography noted oculomotor muscles injury and exophthalmos. Magnetic Resonance Imaging, was requested but the patient disappeared. In November 2017, she returned for an ophthalmic consultation at the same clinic for worsening symptoms in a context of physical asthenia, weight loss and intermittent fever. A surgical exeresis of the tumor was performed. The immunohistochemical examination showing a lymphomatous proliferation consisting of small lymphocytic cells with a hyperchromatic rounded nucleus with cytoplasmic rim. It was later referred to our department where the immunohistochemical examination found the same histological appearance with lymphoma cells that were positive for CD20 and CD79a. They were negative for CD23, CD10, Cyclin D1. The morphological image was consistent with MALT marginal zone lymphoma.

Clinically, the general condition is average with a SPI to 2, in a patient with a temperature at 38 ° C, moderate pallor mucocutaneous. There was no jaundice. There was a tumor syndrome made of a fixed right orbito-palpebral mass, painless, about 7cm difficult to assess because of mass effect on ocular orbit with the eye in total exophthalmia. At the level of the external palpebral region, the skin opposite is smooth, glistening, traversed by some venous lacing and covering the right eye. There was no peripheral lymphadenopathy. The rest of the exams were peculiar.

Paraclinically, the blood count was strictly normal, the multiparametric biochemical assessment showed renal and hepatic function was normal. Hepatitis B, C and HIV were negative, as was HTLV1 serology. The inflammatory assessment noted a positive C-reactive protein C at 192 mg / L and a Lactodehydrogenase (LDH) level = 481 IU / L. MRI orbito-cerebral MRI noted tissue mass and hypointense T2, hypersignal diffuse ADC low, contrast enhancement with gadolinium injection of right intra-orbital seat measuring 36mm / 24mm / 57mm. This mass seems developed at the expense of the right rectus muscle and throwing back the right eyeball 7 mm right and optic nerve. There was a mass effect on the lateral orbital wall without image of bone lysis. There was the integrity of the optic nerves, no abnormality of the occulomotor muscles and the retro-orbital fat and the integrity of the chicsma.

The scanner was normal. The requested endoscopic assessment could not be realized because of lack of financial means. The pre-therapy assessment noted normal heart function with FEV 65%.

Tele: 0022589542209
E-mail address: stcyrpacko@yahoo.fr

The osteomedullary biopsy does not aim at medullary invasion. RCVP chemotherapy had led to a regression of tumor syndrome after two courses of treatment and currently

Discussion

MALT lymphoma has been recently described and first published by Isaacson in 1983 [10]. Gastric localization is the most common. The other localizations are little observed and represent 15% among which the localization at the level of the ocular appendages [2, 11]. It is a low grade lymphoma of malignancy whose evolution is indolent and often localized [12]. This corroborates the slowness of evolution in our patient found in the anamnesis with a gradual start since 2015. The symptomatology revealing the picture of our patient was dominated by exophthalmos and a palpable mass. The latter sign is found in 80% of cases according to White [13]. However other signs can be found such as palpebral edema, ptosis, oculomotor disorders [13, 14]. The other clinical signs of MALT lymphoma vary according to their location. The gastric-type gastric spur-like signs in the gastric gastrointestinal tract show signs of abdominal pain, diarrhea, constipation and dyspepsia in the intestinal localization [14]. The absence of its signs in the clinical course of our patient excludes the possible possibility of these localizations and suggests a single lymphoma that was the right orbito-palpebral muscles. Diagnostically, immunophenotyping on peripheral blood or immunohistochemistry on the affected organ biopsy specimen allows the diagnosis of MALT lymphoma as in all other small cell lymphoproliferative B syndromes. In our study, immunohistochemistry on the biopsy piece of the orbito-palpebral mass found a lymphomatous proliferation consisting of small cells positive for CD 20, CD79a, and negative for CD10, CD23, cyclin D1. This aspect allowed us to confirm the diagnosis. These membrane markers of B lymphocytes are similar to data from the literature [15]

Conclusion

The interest of our observation lies in the rarity of the location of this lymphoma and in our context was to encourage practitioners to systematically perform a biopsy before any evocative clinical signs.

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