

Ocular Involvement and Contribution in Diagnosing a Case of Non-Hodgkin's T Cell Lymphoma

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Abstract

The objective is to report a case of non-Hodgkin's and HTLV lymphoma with ocular manifestation at the Puc Campinas University Hospital. A patient with skin lesions, as well as lymph node and pulmonary alterations, especially with hyperemia and persistent eye pain with no signs of inflammatory disease, trauma or apparent triggering factor. Progressing with scleral nodules being biopsied: the result of anatomopathological suggested a lymphoid tumor. From there, the patient undertook other approaches and diagnostic tests that confirmed the hypothesis and settled diagnosis. Patient undergoing chemotherapy on the CHOP regimen. Purpose is to expose an rare case in the ophthalmological routine.

Keywords: Ocular Surface Lymphoma; Scleral Nodules; Scleral Necrosis; Non-Hodgkin's Lymphoma; Uveal Exposure

Introduction

Adult T-cell leukemia/lymphoma (ATL) is a malignancy of mature CD4+ T-cells caused by human T-cell lymphotropic virus type 1 (HTLV-1). Twenty million people are believed to be infected throughout the world. It is classified in the following clinical forms: acute, lymphoma, primary cutaneous tumoral, chronic (favorable and unfavorable) and smoldering (leukemic and non-leukemic) [1]. Although it is considered an aggressive disease, there are cases with a long progression. Ocular manifestations have yet to be well documented due to its rarity. The mechanisms that underlie ocular involvement in ATL patients remain poorly understood [2,3]. We report a case of a patient with tegumentary, ocular and multiple organs involvement. The diagnose was oriented after a scleral biopsy.

Case Report

JNVS, 34 years old, brown, alcoholic, smoker, marijuana and cocaine user with psoriasis came for a routine consultation in the dermatology department, referring a worsening of the dermatological condition associated with ocular hyperemia and pain on eye movement, especially on the right side. He also reported persistent afternoon fever, headache and cough for 60 days (Figure 1). Therefore, the patient was admitted for diagnostic investigation under the hypothesis of Lyme disease, tuberculosis, psoriasis decompensation and paraneoplastic syndrome. Laboratory tests and imaging to screening were requested. During hospitalization, an ophthalmological consultation was requested.

At the ophthalmic examination: visual acuity 20/20 OU, preserved extrinsic eye movement, tonometry 12 mmHg OU, biomicroscopy of anterior segment of both eyes showed hyperemia of bulbar conjunctiva 2+, consensual and photomotor reflex preserved, absence of anterior camera reaction or previous signs of uveitis; fundoscopy within the normal pattern; ocular USG without changes. The management was conservative and combined with dermatology.

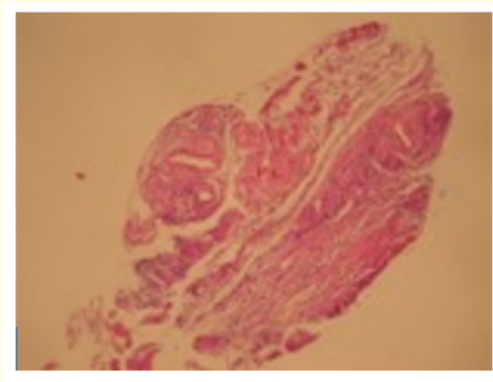


Figure 1

During the same hospitalization, a cutaneous biopsy was performed, revealing a mild chronic inflammatory process and lymphocytic infiltration in the dermis/epidermis interface. In addition to the following results of laboratory tests and serology: blood count: Hb 13.3; Ht 43%, leucogram: Basts 6%, segmented 67%, lymphocytes 20%, monocytes 6%; protein electrophoresis with increased gamma globulin, increased Beta 2 microglobulin contrast with bone marrow examination in the absence of active hematological disease. Serum HTLV 1 and 2 positive, ANA +, BAAR culture and other serologies negative. Computed tomography showed: nodules in the right subpleural region, lymph node changes in the inguinal and axillary chains; an excisional biopsy was carried out with an inconclusive result regarding the neoplasm but evidenced undifferentiated proliferation of large and intermediate cells. Prednisone 60 mg/day and methotrexate was introduced, with clinical improvement and consequent hospital discharge.

He maintained clinical stability for months with exacerbation of the condition in the attempts to regress the corticosteroid dose. After 7 months, he returned for ophthalmologic evaluation, referring worsening of hyperemia and ocular pain, presenting diffuse endothelial infiltrate and scleral nodules in both eyes (Figure 2-4), it was decided to perform a biopsy of the scleral lesion, which showed lymphocytic proliferation (Figure 3); being suggested by our pathology department to perform new skin biopsy for diagnosis of the case. This new biopsies showed: proliferation of lymphoid cells of intermediate size, compromising the dermis and superficial and deep epidermis, with an immunohistochemistry panel: CD3, CD4, CD5, CD8, CD45Ro diffuse positivity; Ki 67 positive expression in about 25-30% of the cellularity of interest (Figure 5 and 6); ALK-1, BCL-6, CD10, CD30, CD56, EMA, CD20 negative; BCL-2, CD79a, MUM-1 negative and positive internal control. Such findings established diagnostic criteria for immunophenotype T non-Hodgkin's lymphoma with extensive skin involvement.

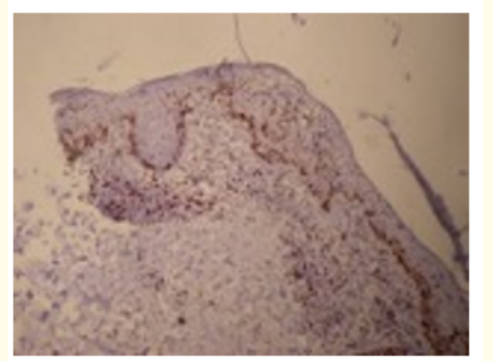


Figure 2



Figure 3

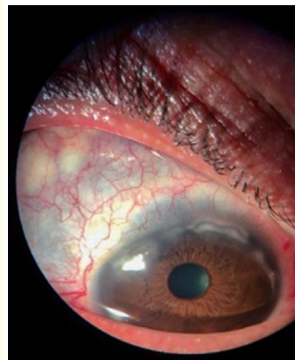


Figure 4



Figure 5

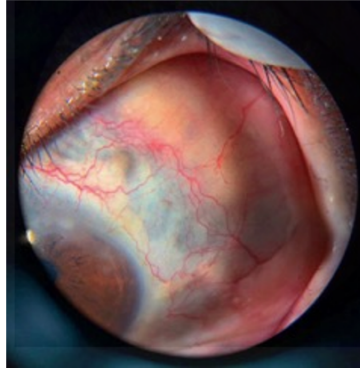


Figure 6

The patient was referred to the onco-hematology sector to start chemotherapy under the CHOP regimen.

Discussion

Ocular manifestations of this entity remain poorly documented. It has been linked as a common cause of uveitis in certain areas of Japan. Most patients present intermediate uveitis with moderate or heavy vitreous opacities [4]. ATL cells have the potential to infiltrate various tissues in the eye such as the orbit, conjunctiva, lacrimal glands, cornea, vitreous humor, uvea, retina, choroid and optic disc [4,5].

In this case our patient presented an atypical chronic presentation, classified as non-uveitic [4], reinforced by the fact that his visual acuity remained unchanged. He presented scleral infiltration of lymphocytic proliferation confirmed by the biopsy and it was not described in previous case reports. Although he didn't present active ocular inflammation during our evaluations, he presented diffuse endothelial infiltrate which could indicate previous inflammation. According to our service protocol, the patient is under treatment with the CHOP regimen, remains clinically stable and without recurrences of ophthalmologic manifestations.

Conclusion

Due to multidisciplinary follow up, we were able to get the rare diagnose. With the development of novel therapies, the longer survival times of ATL patients have raised, such as great quality of vision. Hematologists must be aware of ocular manifestations of ATL, mainly uveitis and always refer to an ophthalmologic evaluation [6].

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