International Journal of Advanced Multidisciplinary Research
ISSN: 2393-8870
www.ijarm.com
DOI: 10.22192/ijamr Volume 5, Issue 9 -2018

Case Report
DOI: http://dx.doi.org/10.22192/ijamr.2018.05.09.001

"Presentation of a case of diffuse infiltrative lymphocytic syndrome associated with HIV in a school-age patient"

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Keywords

Human immunodeficiency virus, diffuse infiltrative lymphocytic syndrome (DILS), Sjögren's syndrome (SS), adenopathy, parotid.

Abstract

The infection caused by HIV causes a cytopathic effect in the CD4 + T lymphocytes, during the initial phase of the infection the CD8 + T cells proliferate, in response against the viral antigens. However, when the expansion of CD8 + lymphocytes is persistent, it results in peripheral CD8 T lymphocytosis associated with lymphocyte infiltration in various organs[5]. The diffuse lymphocyte infiltration syndrome (DILS) is characterized by seropositivity for HIV, bilateral growth of the salivary glands or persistent xerostomia for more than six months, lymphocytic infiltration of CD8 + of the lacrimal or salivary glands in the absence of granulomatous or neoplastic tissue12. This growth is accompanied in 60% of cases of dry symptoms. Knowledge of the rheumatological manifestations of HIV infection in pediatrics can help clinicians to recognize early cases of infection before diagnosing them when it has progressed devastatingly in the patient.

Introduction

In 2016 it was estimated that around 36.7 million people were infected with HIV worldwide, of which 2.1 million corresponded to children under 15 years of age[1], only 43% of children had access to treatment. The United Nations Organization estimates that 370,000 children are infected every year, the equivalent of six infected children per minute[2]. The infection caused by HIV causes a cytopathic effect in CD4 + T lymphocytes, increasing the risk of patients to acquire opportunistic infections[3,4]. During the initial phase of the infection CD8 + T cells proliferate, as part of the immune response triggered by the host against viral antigens, this response is not durable and is followed by the cellular decrease of both types of lymphocytes throughout the natural history of the disease[4]. However, when the expansion of CD8 + lymphocytes is persistent, it results in peripheral CD8 T lymphocytosis associated with a lymphocyte infiltration in various organs[5], these findings suggest that HIV may be the antigen responsible for these clonal expansions, which represents an immune response against viral antigens[6].

Diffuse lymphocyte infiltration syndrome (DILS) was defined for the first time 30 years ago[7]. The presence of persistent bilateral parotid growth and, in some cases, of the lacrimal gland, xerostomia and keratoconjunctivitis, accompanied by persistent lymphocytosis with infiltration to different organs[8] should lead us to make a differential diagnosis between DILS, Sjögren's syndrome (SS) and lymphoma. The difference between DILS and SS lies in the frequent appearance of extra-glandular sites of lymphocytic infiltration in DILS, the nature of infiltrating lymphocytes, that is, CD8 in DILS, CD4 in SS and the presence of positive autoantibodies is rare in DILS (anti-Ro, anti-La)[9].

There are HLA associations (HLA-B45, B49, B50, DR5 and DRw6)[10, 11] that allow the first condition to be differentiated from the second. The following diagnostic criteria have been proposed for DILS: seropositivity for HIV, bilateral growth of the salivary glands or persistent xerostomia for more than six months, lymphocytic infiltration of CD8 + of the lacrimal or salivary glands in the absence of granulomatous or neoplastic tissue[12]. This growth is accompanied in 60% of cases of dry symptoms. The average time between the diagnosis of HIV infection and the onset of DILS symptoms was 3.4 years [13].

The prevalence of DILS is variable in each geographic region, a range between 0.85% to 3%[14], has been reported in African population it is up to 7.8% [4]. The rate of opportunistic infections decreases in patients with DILS, the main hypothesis for this "protection mechanism" is that lymphocyte infiltration participates in the suppression of viral replication in infected cells[15].

Case Report

An 8-year-old female patient was referred to the Infectology Department for bilateral parotid adenopathy of four years of evolution (Review image 3), without other aggregate symptoms, for study protocol.

The physical examination revealed a right parotid gland approximately 10x15cm in diameter, left parotid 8x5cm in diameter, both painless on palpation, with soft, depressible, mobile features with regular edges; anterior cervical adenopathies of 1x1.5cm in diameter, without data of airway compromise, inguinal adenopathies of 1x1cm in diameter. The patient had a history of a single hospitalization due to resection of a left cervical adenopathy with a biopsy that reported chronic sialoadenitis and a history of acute rhinopharyngitis of two to four episodes a year, with no history of blood transfusion.

Fine-needle aspiration biopsy of right parotid was performed with a report of abundant lymphocytic infiltrate. The patient was given a viral panel with a reactive result for HIV, a Western Blot test with positive proteins: GP 160, GP 110/120, GP 41, P 68/66, P 52/51, P 34/31, P 55, P 40, P 24/25, P 18/17. Adoptive parents with ELISA for non-reactive HIV, biological mother terminated by unspecified complications of AIDS, this information was revealed by the adoptive parents after the result of the minor.

The viral genotype resulted in HIV-1, viral load 27, 200 copies / ml, CD4 197.23 Cells / uL (201-3201), CD8 1, 176.27 Cells / uL (81-1821), ratio CD4 / CD8 0.17 (higher a 1), CD 19 and CD 20 positive, ANA by positive indirect immunofluorescence with mottled coarse pattern 1:80, AC Anti SSA / Ro negative and AC Anti SSB / Negative, was also reported, IgG 2659 (572-1474) with Protein electrophoresis compatible with polyclonal gammopathy. A simple and contrasted phase tomography study was performed (Image 1 and 2) with both parotid findings with a notable increase in size with a heterogeneous pattern due to the presence of multiple cysts of 3 to 8 mm on average, with random distribution. In the post-contrast phase, homogeneous reinforcement of the residual parotid parenchyma is seen. There are ganglionic growths of all the cervical chains with an average diameter of 11 mm. Biopsy with finding of serous acini and ducts with inflammatory infiltrate that focally erases the acinar structure, infiltrate composed of B and T lymphocytes, plasma cells, lymphoid aggregates with active germinal center, fibrous septa, sclerosis and hyalinization of the parenchyma with few remaining remnant ducts, some ducts showed squamous metaplasia; follicular hyperplasia pattern.

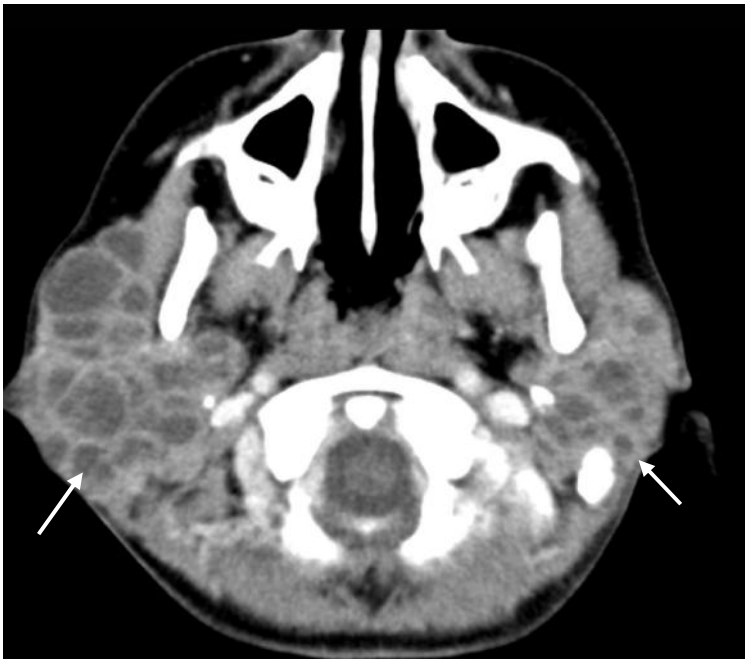

Image 1. Axial section with contrast with multicystic appearance.

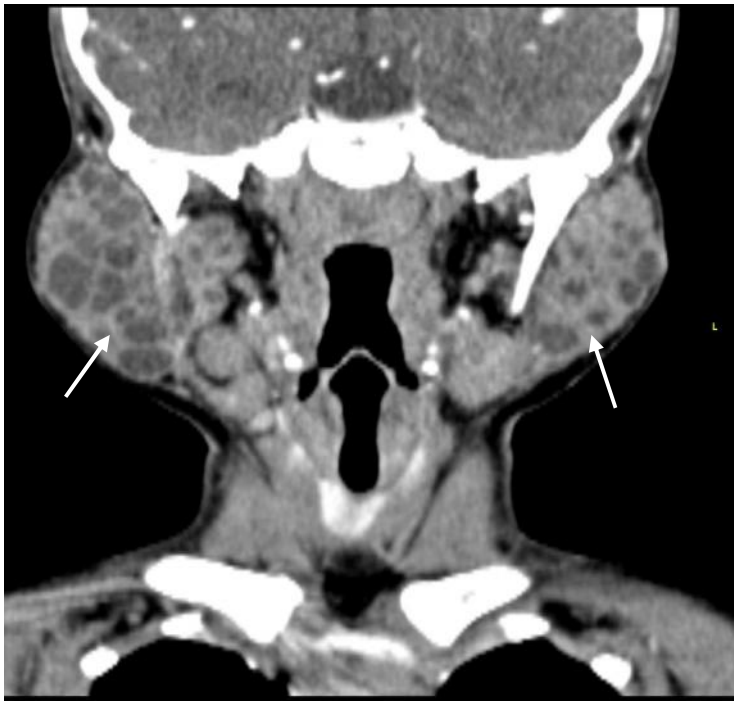

Image 2. Coronal section in arterial phase, increase of both parotid and cervical ganglion chains.

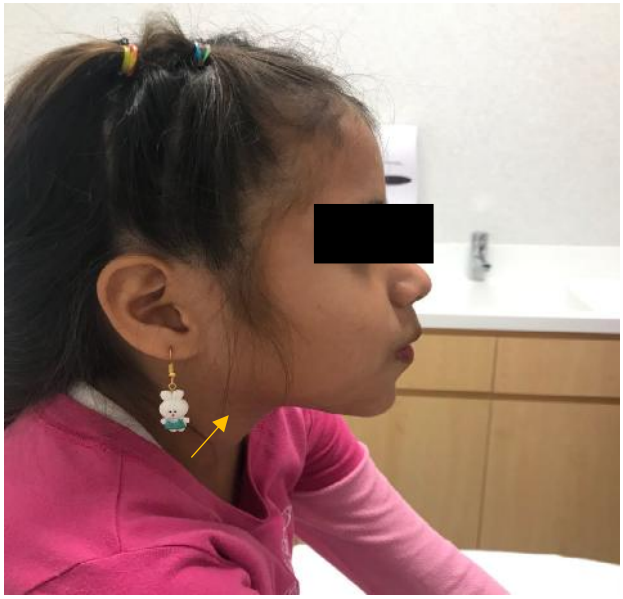

Image 3. Increase in volume of parotid gland.

Discussion

The clinical manifestations of HIV infection have a very varied spectrum; this is one of the main factors for the clinical picture to be diverse and individual of each patient. In some cases the immune system intensifies its work to be threatened by different infections, this response, a tool that limits the agent's progress and damage to the body, "freezing" at the expense of increasing the number of some cell lines in favor to preserve balance. It is in these situations in which diagnosis can be a challenge.

Currently rheumatological manifestations of HIV are widely recognized in the literature, but not in clinical practice where it is challenging to confront a patient with dry symptoms, persistent parotid growth or musculoskeletal disorders, without a history of opportunistic recurrent infections and thinking within the Differential diagnoses, in a condition that has become chronically destructive of the immune system, seems to be a paradoxical response to this infection.

HIV infection in children has a different course than adults, because the infection tends to be acquired earlier in life, this in the former, combined with the development of the immune system. The oral symptom most frequently described in patients with HIV who do not have DILS is xerosis and the most frequently associated lesions are: herpes simplex infection, linear gingival erythema, recurrent aphthous stomatitis, hairy leukoplakia and Kaposi's sarcoma rarely. They are observed in children infected with HIV[16].

Our patient has no history of other symptoms or opportunistic infections or any other oral injury. A theory for the lack of dry symptomatology may be due to the parotid enlargement that compensates with a sufficient salivary production to not give clinical manifestations in an initial way. However, the lymphocytic infiltration of the salivary glands can lead to the deterioration of its function at some point. The lack of salivary production increases the risk of opportunistic infections and caries, as well as having a negative impact on the quality of life due to the difficulty to chew, swallow and savor food. In the present case, we believe that the context of autoimmunity has been determinant for the clinical picture, the positive ANA and the polyclonal gammopathy presented in the patient. Antibodies and hypergammaglobulinemia seem to be present, especially at the beginning of infection, by polyclonal activation of B cells, it is very rare to find positive antibodies in cases of DILS[17].

DILS biopsy is more frequently to find cystic lymphoid hyperplasia and periductal CD8 + lymphocytic infiltration with acinar atrophy, ductal ectasia and fibrosis. The histopathology of the presented case has important fibrosis and inflammatory infiltrate consisting of B and T lymphocytes, which may be due to the time of evolution, the histopathological pattern can also be influenced by the viral load[18].

Conclusion

Oral manifestations are common and prevalent in pediatric HIV infection and have been found as the first indicators of HIV infection. The use of oral lesions as predictors of disease progression could be of great importance. Knowledge of the rheumatological manifestations (although infrequent) of HIV infection in pediatrics can help clinicians to recognize seropositive cases early, before the manifestations are devastating in patients. The awareness that HIV infection can have very diverse and infrequent clinical presentations can help identify cases of DILS. Likewise, this case reminds us of the importance of making a detailed clinical history and keeping in mind the social and family environment of children, inspire and give confidence with a broad sense of confidentiality for families is essential to arrive at an effective diagnosis from the patients.

Acknowledgments

We thank the professors who provided us with their advice for the drafting of our report Dr. Claudia Fernández, Dr. Piedras of the Department of Parology, Dr. Susana Pavón of the Department of Immunology and Allergies.

Competing interests

The authors declare that they do not have any type of conflict of interest, contribute to the exchange of knowledge.

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Quick Response Code

DOI:10.22192/ijamr.2018.05.09.001

How to cite this article:
D. Michelle Cruz, Iris E. Alonzo, Claudia P. Fernández-Luna. (2018).Presentation of a case of diffuse infiltrative lymphocytic syndrome associated with HIV in a school-age patient. Int. J. Adv. Multidiscip. Res. 5(9): 1-6.
DOI: http://dx.doi.org/10.22192/ijamr.2018.05.09.001