Juvenile systemic lupus erythematosus in a adolescent with acquired immunodeficiency syndrome

Nathália de Carvalho Sacilotto[1], Cintia Yukimi Yamashiro[1], Teresa Maria Isaac Nishimoto[2]

RESUMO

Systemic lupus erythematosus juvenile (SLEJ) is a multi-systemic, chronic inflammatory disease, and with autoimmune features. Some clinical manifestations of this disease are similar to those found in Acquired Immunodeficiency Syndrome (AIDS). Coexistence of AIDS with SLEJ is rare, especially in the pediatric population, being described in the literature just 5 patients with congenital HIV infection who developed this rheumatological condition, presenting lupus nephritis as the initial manifestation. We report the case of a 14 year old patient, diagnosed with HIV infection at 8 months of age, with signs and symptoms of SLEJ. This report aims to describe a female patient with AIDS who developed SLE in its classic and forms, but has evolved satisfactorily.

Keywords: juvenile systemic lupus erythematosus, acquired immunodeficiency syndrome, adolescent.

INTRODUCTION

Juvenile Systemic Lupus Erythematosus (JSLE) is a multisystem and chronic inflammatory disease of unknown origin and autoimmune nature.[1] The estimated annual incidence in the juvenile population is 6-20 cases per 100,000 children, predominantly in girls and non-whites.[2] In this age group, the disease is associated with high morbidity and high financial and social impact, causing physical and functional disability, as well as affecting the quality of life of patients and their families.[3] Some clinical manifestations such as fever, myalgia, lymphadenopathy, weight loss, anemia, and leukopenia,[4] and JSLE immunological abnormalities are similar to those found in other diseases such as acquired immunodeficiency syndrome (AIDS).[5] AIDS is a collection of symptoms and infections resulting from specific damage to the immune system. The main target are CD4+ T cells, essential for coordinating the body's defenses.[6] Coexistence of AIDS and JSLE are extremely rare, especially in children. The literature reviewed described only five cases of patients with congenitally acquired HIV infection who developed this rheumatologycal disease and had as primary conditions nephritis or cutaneous vasculitis.[7]

CASE REPORT

TGC, female, 14 years old, diagnosed with HIV infection at 8 months of age. She acquired the disease by maternal-infant transmission and is in antiretroviral therapy since the age of 4. She reports that two months before she developed fever, arthritis in knees and hands, malar rash, and photosensitivity. On the last week, she evolved with dry cough, abdominal pain, nausea and vomiting.

On admission, the patient was taking didanosine, lamivudine, lopinavir, and ritonavir, with undetectable viral load, CD4 lymphocyte count 1524 cells/mm^3, and CD8 T cells 996 cells/mm^3.

On physical examination, she appeared pale 2+/4+, dehydrated+/4+, icteric, febrile (39.5°C), with heart rate of 127 bpm and respiratory rate of 28 bpm, blood pressure 110x60 mmHg, erythematopapulous plaque in the face, chapped lips, aphthous lesions in jugal mucosa, suprasternal retraction, throbbing nose, and jugular stasis. On auscultation, she had crepitant rales in lower thirds, diffuse wheezing, and rhythmic heart sounds without murmurs at two times. The abdomen was distended and painful on palpation, with decreased bowel sounds. Importantly, although the patient had been ill

Received on 03/11/2009. Approved on 05/30/2010. We declare no conflicts of interest.
Hospital Guilherme Álvaro – Centro Universitário Lusíada, Santos, SP, Brazil.
1. Medical Student – Centro Universitário Lusíada.
2. Pediatrician – Secretaria do Estado de São Paulo do Hospital Guilherme Álvaro e da Secretaria de Saúde de Santos.
*Correspondence to:*Nathália de Carvalho Sacilotto. Rua Banco das Palmas, 196. Santana, São Paulo, SP, Brazil. CEP: 02016-020. Phone: 55 (11) 9671-6953. E-mail: nath-cs@uol.com.br.

for a long time and in use of antiretrovirals, she did not have lipodystrophy.

We conducted laboratory tests with the following results: Hemoglobin, 9.3 (12-16g/dL); hematócrito, 27.4 (35-47%); MCV, 85.9 (80-100fL); MCHC, 33.9 (32 -37g/dL); platelets, 223,000 (140-500.000/mm3); WBC, 2720 (3.500-11.000/mm3); VHS, 115 (up to 10 mm/1h); PCR, 26.80 (< 5.0 mg / L); CH50, 30 U/CAE (= 60 U/CAE); C3, 25 mg/dL (90-180 mg/dL); C4, 6.0 mg/dL (10-40 mg/dL); ANA, reagent by indirect immunofluorescence on HEp-2 (1/640, homogeneous nuclear pattern); anti-native DNA (method: indirect immunofluorescence using *Crithidia lucilae* as substrate) and anti-Sm and anti-RNP, reagents (method: double immunodiffusion Ouchterlony; anti-SSA/Ro and anti-SSB/La, non-reagents (method: double immunodiffusion Ouchterlony). Chest x-ray showed opacification of both hemithoraces, and 24-hour proteinúria = 0.96 g (< 0.10 g/24 hours).

JSLE diagnosis was established using the classification criteria proposed in 1982 by the American College of Rheumatology and revised in 1997,[8] as patient presented with malar rash, photosensitivity, nonerosive arthritis, positive ANA, Anti-DNA positive, serositis, and leukopenia.

The patient progressed to diagnosis of pneumonia, acute respiratory distress syndrome, septic shock, and JSLE. She received supportive care, broad-spectrum antibiotics, and pulse therapy with methylprednisolone for four days and then prednisone 40 mg, showing clinical improvement and control of both diseases.

DISCUSSION

Unlike the writing of the current report, according to literature, there are in the pediatric population four published cases of HIV infected children (7 to 37 months old) by congenital transmission that developed lupus nephritis with no manifestations of SLE and one patient (42 months old) who developed cutaneous vasculitis and who met criteria for this rheumatic disease.[7] There are also published in literature a case of congenital HIV patient (9-year-old) that initially had lupus manifestations and developed AIDS years later.[7]

Both diseases are similar and relate to one another, with different assumptions in literature trying to explain the mechanisms by which this occurs.[7]

In addition to the clinical manifestations, a series of laboratory results can occur in both diseases, including leukopenia, lymphopenia, hypergammaglobulinemia, and the presence of antiphospholipid antibodies. Antinuclear antibodies and rheumatoid factor are present, although less frequently in individuals infected by HIV, but the existence of anti-native DNA has been described in patients carrying this virus.[9]

There is evidence that viruses may play a role in the development of rheumatic diseases, such as Epstein-Barr virus, parvovirus, hepatitis virus, and retrovirus.[10] This occurs in different ways; among them there is the surface glycoprotein gp120 present in HIV, which is a major disrupter of the immune system.[5]

HIV infection can improve clinical manifestations of SLE, but there is parallel progression of immunodeficiency by this virus. A class of drugs used to treat AIDS is the protease inhibitors, which can promote the reactivation of SLE and aggravate its clinical course by promoting increased number of circulating CD4+ and restoring the immune function, showing that we must exercise caution with selected drugs for antiretroviral therapy.[4] However, nothing is mentioned regarding the onset of rheumatic disease.

In 1991, Wallace reported that the increased antibody production in SLE eventually becomes protective against HIV infection, which may occur also through the possible effects of antiretroviral drugs found in immunosuppressive agents.[4] On the other hand, there are reports that this class of drugs may decrease this benefit, resulting in subsequent increase in viral load.[4]

Regardless of the organ or system affected, the continued use of antimalarials, such as chloroquine 4 mg/kg/day or hydroxychloroquine sulfate 6 mg/kg/day, is indicated in order to reduce disease activity and attempt to limit the use of corticosteroids. Drug maintenance in controlled patients reduces the possibility of activity resurgence, improves lipid profiles, and reduces the risk of thrombosis.[11]

This report aims to alert the possibility of how the association of both diseases can affect the degree of immunesuppression, and emphasizes the extreme importance of early diagnosis and treatment.

REFERÊNCIAS

REFERENCES

1. Rus V, Hochberg MC. The epidemiology of systemic lupus erythematosus. In: Wallace DJ, Hahn BH (eds.). Dubois lupus erythematosus. 6 ed. Philadelphia: Lippincott Williams & Wilkins 2002; pp. 66-83.
2. Sztajnbok1 FR, Serra CRB, Rodrigues MCF, Mendoza E. Doenças reumáticas na adolescência. J Pediatr 2001; 77(2):234-44.
3. Machado C, Ruperto N. Consenso em Reumatologia Pediátrica Parte II – Definição de Melhora Clínica para o Lúpus Eritematoso Sistêmico e Dermatomiosite Juvenil. Rev Bras Reumatol 2005; 45(1):14-9.

4. Calza L, Manfredi R, Colangeli V, D'Antuono A, Passarini B, Chiodo F. Systemic and discoid lupus erythematosus in HIV-infected patients treated with highly active antiretroviral therapy. International Journal of STD & AIDS 2003; 14(5):356-9.
5. Sekigawa I, Kaneko H, Hishikawa T, Hashimoto H, Hirose S, Kaneko Y *et al*. HIV infection and SLE: Their pathogenic relationship. Clin Exp Rheumatol 1998; 16:175-80.
6. Ministério da Saúde. DST-AIDS. Disponível em http://www.aids.gov.br/data/Pages/LUMIS4A27BE0AITEMID1C6F2BAFF06E4B8BB337AA90A936F967PTBRIE. Acesso em 02 de agosto de 2008.
7. Chalom EC, Rezaee F, Mendelson J. Pediatric patient with systemic lupus erythematosus & congenital acquired immunodeficiency syndrome: An unusual case and a review of the literature. Pediatr Rheumatol Online J 2008; 6:7.
8. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997; 40:1725.
9. Koppelman RG, Zolla-Pazner S. Association of human immunodeficiency virus infection and autoimmune phenomena. Am J Med1988; 84:82-8.
10. Davis P. Viral infections, acquired immunodeficiency syndrome, and rheumatic diseases. Curr Opin Rheumatol 1992; 4:529-33.
11. Sato EI, Bonfá ED, Costallat LTL, Silva NA, Brenol JCT, Santiago MB *et al*. Lúpus eritematoso sistêmico: acometimento cutâneo/articular. Rev Assoc Med Bras 2006; 52(6):384-6.