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Case report

Pyomyositis in childhood-systemic lupus erythematosus

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ABSTRACT

Pyomyositis is a pyogenic infection of skeletal muscle that arises from hematogenous spread and usually presents with localized abscess. This muscle infection has been rarely reported in adult-onset systemic lupus erythematous and, to the best of our knowledge, has not been diagnosed in pediatric lupus population. Among our childhood-onset systemic lupus erythematous population, including 289 patients, one presented pyomyositis. This patient was diagnosed with childhood-onset systemic lupus erythematous at the age of 10 years-old. After six years, while being treated with prednisone, azathioprine and hydroxychloroquine, she was hospitalized due to a 30-day history of insidious pain in the left thigh and no apparent trauma or fever were reported. Her physical examination showed muscle tenderness and woody induration. Laboratory tests revealed anemia, increased acute phase reactants and normal muscle enzymes. Computer tomography of the left thigh showed collection on the middle third of the *vastus intermedius*, suggesting purulent stage of pyomyositis. Treatment with broad-spectrum antibiotic was initiated, leading to a complete clinical resolution. In conclusion, we described the first case of pyomyositis during childhood in pediatric lupus population. This report reinforces that the presence of localized muscle pain in immunocompromised patients, even without elevation of muscle enzymes, should raise the suspicion of pyomyositis. A prompt antibiotic therapy is strongly recommended.

Piomiosite no lúpus eritematoso sistêmico juvenil

Palavras-chave:
Piomiosite
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RESUMO

A piomiosite é uma infecção piogênica da musculatura esquelética, decorrente da disseminação hematogênica e geralmente acompanhada de formação de abscesso localizado. Esta infecção da musculatura é raramente descrita em adultos com lúpus eritematoso sistêmico (LES) e, até onde se sabe, ainda não o foi em pacientes com LES juvenil (LESJ).

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De nossos 289 pacientes com LESJ, uma apresentou piomiosite. Diagnosticada com LESJ aos 10 anos de idade e após seis anos de tratamento com prednisona, azatioprina e hidroxicloroquina, a paciente foi hospitalizada em razão de um histórico de 30 dias de dor insidiosa na coxa esquerda, sem relato algum de trauma aparente ou febre. O exame físico mostrou músculos sensíveis e com endurecimento lenhoso. Os exames laboratoriais revelaram anemia, aumento de reagentes de fase aguda e enzimas musculares normais. A tomografia computadorizada da coxa esquerda mostrou coleção no terço médio do vasto intermédio, sugerindo estágio purulento de piomiosite. Iniciou-se tratamento com antibiótico de largo espectro, que levou à resolução clínica completa. Em suma, descreveu-se o primeiro caso de piomiosite em pacientes com LESJ encontrado neste serviço. Este relato reforça que a presença de dor muscular localizada em pacientes imunocomprometidos, ainda que sem aumento de enzimas musculares, deve sugerir o diagnóstico de piomiosite. Recomenda-se tratamento imediato com antibióticos.

Introduction

Pyomyositis is a pyogenic infection of skeletal muscle that arises from hematogenous spread and usually presents with localized abscess.[1] Infectious myopathy is associated with immunocompromised conditions, especially human immunodeficiency virus (HIV) infection, diabetes mellitus, malignancy, immunosuppressive drugs and rheumatic diseases.[1–7]

Of note, infections are one of the main causes of morbidity and mortality in adult systemic lupus erythematosus (SLE) and childhood-onset SLE (C-SLE) patients,[8,9] and may involve the skeletal muscle. Pyomyositis has been rarely reported in adult SLE,[3–7] and to the best of our knowledge, it has not been diagnosed during childhood in pediatric lupus population.

We reviewed our data from January 1983 to July 2013 and included 5593 patients of Pediatric Rheumatology Unit of Instituto da Criança da Faculdade de Medicina da Universidade de São Paulo. We identified 289 (5.1%) C-SLE patients that fulfilled the American College of Rheumatology (ACR) classification criteria.[10] One (0.34%) of them had a pyomyositis episode after C-SLE diagnosis was established and was described herein.

Case report

A 10-year-old girl was diagnosed with C-SLE based on discoid skin rash, pericarditis, psychosis, lymphopenia, thrombocytopenia, proteinuria, antinuclear antibodies (ANA) 1/1280 (nuclear homogenous pattern), and presence of anti-double-stranded DNA (anti-dsDNA) antibody, fulfilling the ACR classification criteria for SLE.[11] At that moment, the SLE Disease Activity Index 2000 (SLEDAI-2K) score[11] was 28 based on the following findings: psychosis, proteinuria, cellular casts, alopecia, pleuritis, pericarditis, low levels of C3 and C4, presence of anti-dsDNA, fever and thrombocytopenia. She was treated with prednisone (2.0 mg/kg/day) that was progressively tapered to 2.5 mg/day, hydroxychloroquine (5.0 mg/kg/day) and azathioprine (2.0 mg/kg/day). At the age of 16 years, she was hospitalized in our service with a 30-day history of insidious pain in the middle of left thigh, without cramps, apparent trauma or fever. Her physical examination showed localized muscle tenderness and woody induration. She did not present skin erythema and local heat. At that time, she was on prednisone (0.95 mg/kg/day), azathioprine (2.25 mg/kg/day) and hydroxychloroquine (5.0 mg/kg/day) due to severe alopecia, discoid skin rash, photosensitivity and oral ulcers. Laboratory tests revealed hemoglobin 9.3 mg/L, hematocrit 29%, white blood cell count 8700 mm^3 (71% neutrophils, 20% lymphocytes and 8% monocytes), platelet count 202,000 mm^3, C-reactive protein 46.9 mg/L (reference value <5 mg/L), erythrocyte sedimentation rate 54 mm/1st hour (normal range 0–20 mm/h), proteinuria 0.12 g/24 h, urea 41 mg/dL (normal range 15–45 mg/dL) and creatinine 0.36 mg/dL (normal range 0.6–0.9 mg/dL), C3 75 mg/dL (normal range 90–180 mg/dL) and C4 9.9 mg/dL (normal range 10–40 mg/dL). Muscle enzymes tests performed showed: aspartate aminotransferase 31 U/L (normal range 10–34 U/L), alanine aminotransferase 34 U/L (normal range 10–44 U/L), creatine kinase 64 U/L (normal range 24–204 U/L). Anti-dsDNA was positive, and SLEDAI-2K was 4. Computed tomography (CT) of the left thigh showed collection on the middle third of the *vastus intermedius* muscle, dimensions 5.0 cm × 3.0 cm × 2.0 cm, strongly suggestive of purulent stage of pyomyositis. The patient was submitted to an ultrasound-guided puncture, but no fluid was obtained. Blood culture did not identify any organisms. Treatment with intravenous antibiotics, such as cefepime (150 mg/kg/day) and clindamycin (40 mg/kg/day), was administered for a seven-day period. She was then discharged with oral clindamycin (40 mg/kg/day) for more seven days, resulting in a complete clinical resolution.

Discussion

Pyomyositis had never been reported in C-SLE population followed in our tertiary University Hospital in the last 30 years. The presence of localized muscle pain with indurations in an immunosuppressed patient suggested this diagnosis.

This infectious myopathy was first described in 1885 by Scribba[12] and is a rare pyogenic infection of muscle tissue. In childhood, the most affected regions are thighs and hips.[2,13]

It is usually caused by *Staphylococcus aureus*, *Streptococcus* sp. and *Escherichia coli*.[2,7] However, in more than two thirds of the cases, the etiological agent is not identified in blood cultures,[14] as observed in our patient.

Pyomyositis comprises three clinical stages: invasive (insidious and nonspecific symptoms); purulent (deep abscess formation and inflammatory signs); and late stage (high fever, severe pain, muscle fluctuation and occasionally septic shock).[2,7] Purulent stage is observed in up to 90% of patients at diagnosis,[15] as evidenced herein.

Of note, the clinical presentation and the laboratory exams of pyomyositis are nonspecific. White blood cell count can be elevated in 50–60% of cases; acute phase reactants are usually high[15] and muscle enzymes are often normal,[2] as observed in this case. Therefore, early imaging tests are imperative for the diagnosis of pyomyositis. X-ray studies can be used to rule out bone lesions. Ultrasonography is an accessible, low-cost and quick tool, but not sensitive enough, particularly at the onset of the infection.[14] Importantly, CT or magnetic resonance imaging confirms the diagnosis, and may reveal muscle enlargement, abscess extension, joint effusions and fluid collections.[15]

This disease can also occur in healthy patients after local mechanical trauma,[13,15] and can be associated with parasitosis and desnutrition, also known as tropical pyomyositis. The atypical or nonspecific presentation of pyomyositis may delay the diagnosis and treatment in immunosuppressive and/or chronic diseases, such as HIV infeccion,[13–15] HTLV infeccion,[7] leukemia,[13–15] diabetes,[13–15] scleroderma,[13] and dermatomyositis.[15] To the best of our knowledge, there are only five cases described of pyomyositis in adult SLE patients[3–7] and only one with childhood-onset lupus who presented pyomyositis at the age of 32.[4] Therefore, the first case of pyomyositis in pediatric lupus population was described herein.

Differential diagnosis of pyomyositis affecting the thigh includes hematoma, osteomyelitis, septic arthritis, deep venous thrombosis, cellulitis and malignant tumors.[2,15]

The treatment depends on the stage of disease. In the purulent and late stages, intravenous broad-spectrum antibiotics are the main therapy. Moreover, percutaneous drainage or surgical incision may be necessary in patients refractory to antibiotics.[2] Our patient had complete recovery after clinical treatment.

In conclusion, we described a case of pyomyositis in C-SLE population. This report reinforces that the presence of localized muscle pain in immunocompromised patients, even without elevation of muscle enzymes, should raise the suspicion of pyomyositis. A prompt antibiotic therapy is strongly recommended.

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Conflicts of interest

The authors declare no conflict of interest.

REFERENCES

1. Lopez FA, Lartchenko S. Skin and soft tissue infections. Infect Dis Clin North Am. 2006;20:759–72.
2. Mitsionis GI, Manoudis GN, Lykissas MG, Sionti I, Motsis E, Georgoulis AD, et al. Pyomyositis in children: early diagnosis and treatment. J Pediatr Surg. 2009;44:2173–8.
3. Dede H, Ozdocan H, Dumankar A, Aktuglu Y, Tabak F, Yazic H. Tropical pyomyositis in a temperate climate. Br J Rheumatol. 1993;32:435–6.
4. Claudepierre P, Saint-Marcoux B, Allain J, Montazel JL, Larget-Piet B, Chevalier X. Clinical images: value of magnetic resonance imaging in extensive pyomyositis. Arthritis Rheum. 1996;39:1760.
5. Ravindran V, Duke O. Non-tropical pyomyositis in a patient with systemic lupus erythematosus. Lupus. 2009;18:379–80.
6. El Baaj M, Tabache F, Modden K, Hassikou H, Safi S, Khalid R, et al. Pyomyositis: an infectious complication in systemic lupus erythematous. Rev Med Intern. 2010;31:4–6.
7. Souza HC, Carvalho BN, Morais MG, Monteiro GZ, Emori FT, Latorre LC. Tropical pyomyositis in a patient with systemic lupus erythematosus and HTLV 1/2 infection. Rev Bras Reumatol. 2011;51:97–103.
8. Faco MM, Leone C, Campos LM, Febrônio MV, Marques HH, Silva CA. Risk factors associated with the death of patients hospitalized for juvenile systemic lupus erythematosus. Braz J Med Biol Res. 2007;40:993–1002.
9. Silva MF, Ribeiro AS, Fiorot FJ, Aikawa NE, Lotito AP, Campos LM, et al. Invasive aspergillosis: a severe infection in juvenile systemic lupus erythematosus patients. Lupus. 2012;21:1011–6.
10. Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum. 1997;40:1725.
11. Gladman DD, Ibañez D, Urowitz MB. Systemic lupus erythematosus disease activity index 2000. J Rheumatol. 2002;29:288–91.
12. Scriba J. Beitrag zun aetiologie der myositis acute. Dtsch Z Chir. 1885;22:497–502.
13. Nikolopoulos DD, Apostolopoulos A, Polyzois I, Liarokapis S, Michos I. Obturador internus pyomyositis in a young adult: a case report and review of the literature. Cases J. 2009;2:8588.
14. Moran GG, Duran CG, Albiñana J. Imaging on pelvic pyomyositis in children related to pathogenesis. J Child Orthop. 2009;3:479–84.
15. Drosos G. Pyomyositis. A literature review. Acta Orthop Belg. 2005;71:9–16.