Case Report

Cutaneous polyarteritis nodosa – case report*
*Poliarterite nodosa cutânea – relato de caso**

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Abstract: Cutaneous polyarteritis nodosa is the vasculitis affecting medium-sized vessels in the deep dermis and panniculus. It is characterized by tender nodules which may ulcerate, preceded, accompanied or followed by livedo reticularis. Its diagnosis is confirmed by clinical and histopathological findings. A case of a patient presenting a chronic form of the illness is described.
Keywords: History; Polyarteritis nodosa; Vasculitis/therapy

***Resumo**: Poliarterite nodosa cutânea é vasculite que acomete vasos de médio calibre na derme profunda e panículo adiposo. É caracterizada por nódulos dolorosos que podem ulcerar, antecedidos, acompanhados ou seguidos de livedo reticular, sendo seu diagnóstico clínico e histopatológico. Descreve-se caso de paciente com evolução arrastada da enfermidade.*
Palavras-chave: História; Poliarterite nodosa; Vasculite/terapia

INTRODUCTION

Cutaneous polyarteritis nodosa is a rare disease of chronic behavior, not having the same ominous prognosis of the systemic form of vasculitis.

Currently, it is classified as a form of septal panniculitis with vasculitis, affecting arteries and arterioles, sometimes associated with systemic complaints, although generalized involvement by vasculitis is a rare occurrence. Its diagnosis must be thought in patients with inflammatory nodules and livedo reticularis, despite being uncommon.

Its etiology is yet unknown, and in sporadic reports it has been associated with bacterial or viral infections; its treatment still is a major challenge for clinicians, rheumatologists and dermatologists.

CASE REPORT

A 55 year-old woman reported the appearance of Raynaud's phenomenon, livedo reticularis, asthenia and arthralgia in lower limb five years ago. In the past four years, erythematous and tender nodules appeared on the lower limbs. After seeking medical care, p-Anca and FAN were measured and tested negative. The biopsy of the nodules revealed vasculitis and vascular necrosis, suggesting polyarteritis. She did not meet enough criteria of the American College of Rheumatology for the diagnosis of systemic polyarteritis nodosa. Serologic tests for hepatitis were negative. Complete blood count, urine analysis and blood chemistry were normal. The remaining physical examination was normal.

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From that time on, she has been treated with: prednisone, colchicine, thalidomide, oral cyclophosphamide, azathioprine and chloroquine, methotrexate and chloroquine and prednisone, and sulphasalazine, as determined by the rheumatology division; however, no remission of the lesions occurred, which caused the patient to be unhappy.

For the past year, the nodular lesions in the lower limbs have become ulcerated, tender and a new biopsy was done, showing neutrophilic septal panniculitis. At the same time, she presented distal sensitive symmetric neuropathy (Figures 1 and 2). The diagnosis for the case reported was cutaneous polyarteritis nodosa.

At this time, treatment with intravenous cyclophosphamide and long term antibiotics –ciprofloxacin and clindamycin- was prescribed with ulcer healing. Since then, no new lesions have appeared (Figures 3 and 4).

DISCUSSION

Polyarteritis nodosa (PAN), the first vasculitis described, is a classical example of vasculitis of medium-size vessels, sparing arterioles, venules and capillary vessels, and not associated with glomerulonephritis.[1]

The pathophysiological alterations are limited to the arterial bed, and histology depicts vascular destruction with fibrinoid necrosis, and neutrophilic and polymorphonuclear infiltrates. The lesions are segmental and preferably occur in vessel bifurcation sites.[1]

Usually, the systemic illness begins with fatigue, asthenia, fever, myalgia and arthralgia, followed by involvement of specific organs, mainly the skin, peripheral nerves, digestive tract and kidneys (medium-size vessels).

Anemia, thrombocytosis, increased erythrocyte sedimentation rate (ESR) and hematuria without glomerulonephritis are some common findings. For the diagnosis of systemic PAN, the patient must meet at least three of the following American College of Rheumatology criteria, at the same time: weight loss greater than 4 kg, livedo, testicular pain, myalgia, mononeuropathy, diastolic blood pressure greater than 90mmHg, increased serum urea or creatinine, hepatitis B, abnormal arteriography or compatible histological changes.[1-3]

Cutaneous polyarteritis nodosa seems to be a skin limited form of PAN. Clinically, the cutaneous form lesions may present as erythematous nodules, which may ulcerate, and livedo reticularis, especially in the lower limbs. Usually, up to 50% of patients have different symptoms, and the most common are low grade fever, arthargia, myalgia, fatigue and asthenia. Systemic involvement is usually absent, and the illness is chronic. This description very much resembles what this patient presented.[2-4]

In patients with ulcerated cutaneous polyarteritis nodosa, a more chronic course of the disorder and with neurological involvement is common. Up to 59% of patients with ulcers present nodules concomitantly. This has occurred in the patient here reported.[2]

Differently from the systemic disease, cutaneous polyarteritis nodosa does not show immunologic abnormalities that could be detected by FAN, ANCA, rheumatoid factor, cryoglobulin and diminished complement levels. However, these tests must be done to rule out other more severe forms of vasculitis or rheumatologic disorders.[3]

Histology of the cutaneous disorder shows vasculitis of medium-sized arteries and arterioles. The

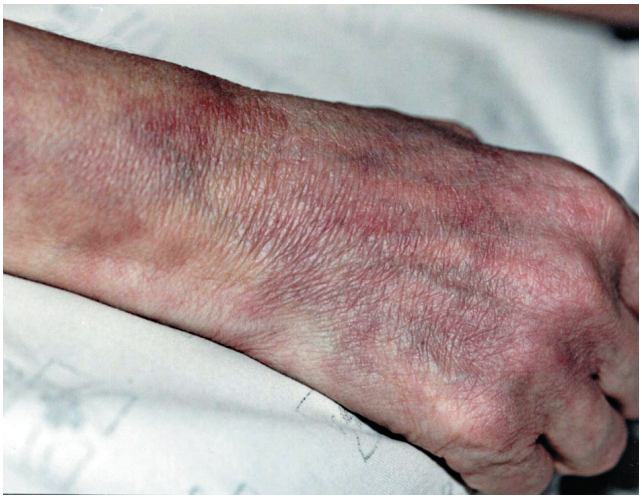

Figure 1: Intense livedo reticularis

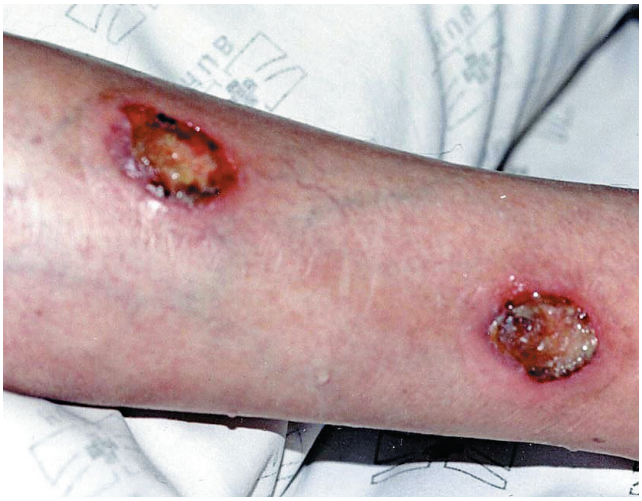

Figure 2: Ulcers in lower limbs

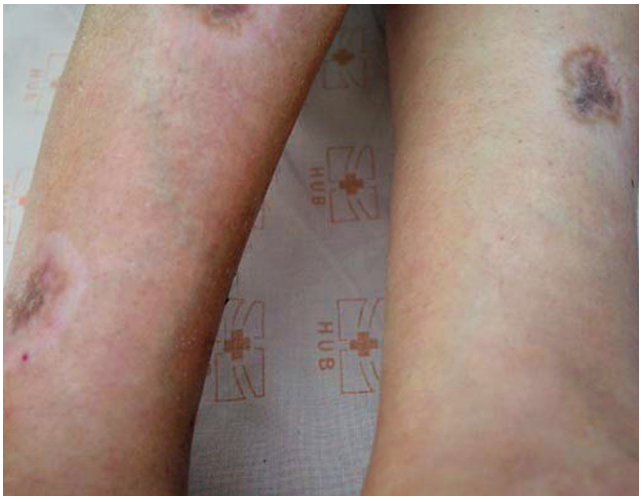

Figure 3: Hand after treatment with antibiotic therapy and cyclophosphamide

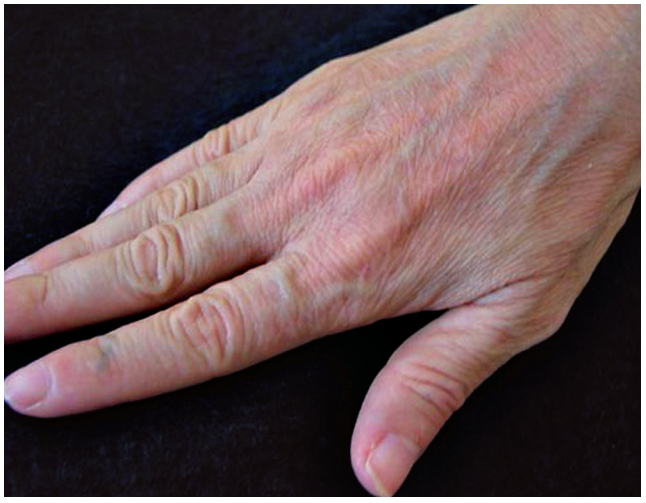

Figure 4: Aspect of atrophic scars after treatment

affected vessels show thickened walls with neutrophilic infiltration. A characteristic finding is an eosinophilic ring of fibrinoid necrosis in the arterial intima, resembling a target.

In most cases of cutaneous polyarteritis nodosa, there are no intraluminal thrombi, although narrowing of the vessels is common. The lesions are usually segmental, but in contrast to the systemic disorder, it has no preference for bifurcations.[3,5,6]

Direct immunofluorescence of cutaneous polyarteritis nodosa reveals deposits of IgM and complement in the affected vessels.[6,7]

The treatment of different forms of polyarteritis is still a problem for the physician. The systemic forms of the disease may evolve well when treated with steroids and cyclophosphamide, which require frequent monitoring of patients. The course of the cutaneous form is fluctuating, rendering the assessment of different treatments, difficult. In the mild forms the use of prednisone in small doses (20mg/day) associated with anti-inflammatory drugs can be attempted. In patients with chronic disease, long term antibiotics may be useful. In patients with severe forms or in those extremely unhappy with the disorder, several immunosuppressants have been tried with controversial results. The assessment of the disease is also difficult due to its course, and many reported cases still have symptoms 20 years later.[1-3,7]

The follow-up of patients with cutaneous polyarteritis nodosa is important because in some cases reported, it progressed to the systemic form.

Cutaneous polyarteritis nodosa is a distinct and localized process of vasculitis, involving medium- and small-sized vessels in the deep dermis and panniculus. Ulceration may indicate a more intense process, often associated with neuropathy. Differently from the systemic form, there is no evidence supporting a relationship with the hepatitis viruses. Moderate increase of ERS and mild anemia are the laboratory changes that may be found, with negative immunological tests. Its diagnosis is clinical and histopathological, it has fluctuating course, and variable and difficult response to treatments.[4,8-12] ❑

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