Case Report

**Polymyositis associated with nephrotic syndrome**

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RESUMO

Polymyositis (PM) is a systemic disease of the idiopathic inflammatory myopathy group, clinically characterized by symmetric and proximal muscle weakness. There are reports in literature of PM associated with malignancies, autoimmune diseases, and viral infections. However, the association between PM and nephropathy is not common. We describe a case report of a patient with polymyositis who developed nephrotic syndrome due to mesangial glomerulonephritis.

**Keywords**: polymyositis, nephrotic syndrome, myositis.

INTRODUCTION

Polymyositis (PM) is defined as an idiopathic myopathy with evolution period of weeks to months and having as its main feature the development of nonsuppurative inflammation in skeletal muscles, which is clinically manifested by symmetric and proximal muscle weakness. Its incidence is approximately one case per 100,000 inhabitants, predominantly in women.¹,²

Progressive muscle weakness is usually the first symptom presented and its subacute character is associated with delays in seeking medical attention by patients; more rarely, the disease may manifest acutely, followed by rhabdomyolysis and myoglobinuria.³,⁴

Affected patients report difficulty in daily tasks requiring the use of proximal muscles, such as climbing stairs, climb the sidewalk curb, lifting objects, or combing their hair. The skeletal muscles of the posterior pharyngeal wall and proximal third of the esophagus are often compromised, leading to dysphagia and choking. Constitutional symptoms may be present and include fatigue, appetite loss, weight loss, arthralgia, or arthritis affecting small and medium joints.⁵,⁶

PM can occur alone or be associated with systemic autoimmune diseases and viral infections, such as lupus, rheumatoid arthritis, Crohn’s disease, HIV infection, and HTLV.⁶ Howewer, renal involvement in PM is not common.⁷⁸ We report the case of a 37-year-old patient who developed anasarca and renal injury, as detected by biopsy.

CASE REPORT

A male patient, 37 years old, was admitted with signs of anasarca, progressive muscle weakness, dysphagia to solids, and development of asthenia for 45 days. The patient reported a weight gain of 15 kg during this period. On physical examination, he was afebrile, hydrated, ruddy, and anicteric; respiratory and cardiac auscultation presented no changes; abdomen was soft, tender, and sensitive to palpation in epigastrium, with edema of the wall, without visceromegaly; oral cavity and skin presented no lesions; the patient had important edema in both lower and upper limbs (Figure 1). Neurological examination showed no change in cranial nerves; symmetric reflexes; superficial and deep sensitivity preserved; and muscle strength grade II, with proximal muscular weakness. Initial tests revealed CPK 15,969 U/L (normal up to 232 U/L), GOT 1,356 U/L (normal up to 37 U/L), GPT 519 U/L (normal up to 65 U/L),...
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albumin 1.2 g/dL (normal up to 3.4 to 5.0 g/dL), and LDH 1.551 U/L (normal up to 190 U/L). Total cholesterol was 143 mg/dL (0-200 mg/dL), with LDL 91.4 mg/dL (0-100 mg/dL), HDL 18 mg/dL (35-60 mg/dL), and triglycerides 168 mg/dL (30-150 mg/dL). Blood count at admission showed mild normochromic normocytic anemia (hemoglobin 11.8 g/dL) and leukocytosis of 18,900 cells/mm³, 85% segmented. Erythrocyte sedimentation rate was 38 mm³ (normal up to 15), positive PCR (qualitative determination), ANA (HEp-2) was 1/160 (normal up to 1/40), with fibrillar cytoplasmic pattern. Deltoid muscle biopsy revealed chronic interstitial and endomysial lymphomononuclear inflammatory reaction (Figure 2A), with areas of fibrosis, presence of necrotic muscle fibers, and muscle fibers with frequent regenerative changes (Figure 2B); histological picture compatible with inflammatory myopathy. Considering the clinical and laboratory findings, the diagnosis of polymyositis was made and treatment initiated with prednisone (1 mg/kg/day). During hospitalization, the patient underwent pulse therapy with methylprednisolone (1 g/day) for three days.

Urine sediment analysis showed proteins and red blood cells (++); 24-hour proteinuria was 3,771 mg (20-150 mg). The presence of these findings on urinalysis associated with the problem of anasarca and hypoalbuminemia led to renal biopsy, which revealed diffuse mesangial expansion (mesangial proliferative glomerulonephritis) and tubular atrophy with focal mild interstitial fibrosis. After 55 days of hospitalization, the patient was discharged, his general condition was good, although he still had difficulty walking, had proximal muscle weakness but without dysphagia, and significant improvement of the anasarca. CPK was markedly reduced – the result at discharge was 663 U/L. He continues as an outpatient at the Department of Rheumatology, Hospital Universitário da Universidade Federal de Sergipe, taking prednisone and participating in a program of monthly pulse therapy with cyclophosphamide, due to the persistence of myopathic and renal involvement.

DISCUSSION

The association of polymyositis with renal disease has been reported in literature both as acute renal failure, due to rhabdomyolysis and myoglobinuria, and associated with glomerulonephritis.4-8-12 This relationship between idiopathic inflammatory myopathies and nephropathy is not frequent, although Yen et al.4 have found some degree of hematuria and proteinuria in 14 of 65 patients (21.5%) with polymyositis and dermatomyositis (DM).

The glomerular lesion most commonly associated with PM is mesangial proliferative glomerulonephritis,9,11 which is consistent with our report and the study by Valenzuela et al.10 However, there are reports on patients with PM who developed rapidly progressive glomerulonephritis.12 Nephrotic syndrome associated with PM is rare, with few cases reported in literature. In the study by Pasquali et al.,8 of the eight patients with polymyositis, only one presented with nephrotic syndrome. On the other hand, there are reports on patients with idiopathic inflammatory myopathies (DM, PM) who developed anasarca not related to nephrotic syndrome.13,14 Gorelik et al.,15 for example, found seven cases of DM and PM related to anasarca in adult males, as described in the episode, and they also reported dysphagia in four of these patients. However, none of these patients progressed to nephrotic syndrome.

The interesting aspect of this case is the association between polymyositis, anasarca, and nephrotic syndrome, unlike other reports in literature. The clinical and laboratory findings of generalized edema, major muscle weakness, and very high
elevations of CPK (15,969 U/L) suggested the presence of muscle mass destruction. However, it is noteworthy that the patient had no renal function deterioration at any time. Despite the good initial response to therapy, the clinical course of this patient is uncertain, since there are few cases in literature.

REFERENCES