



Mikołaj Kamiński , Paweł Hrycaj

Department of Rheumatology, District Hospital in Koscian, Kościan, Poland

Myositis with creatinine phosphokinase activity over 13,500 U/L in the course of granulomatosis with polyangiitis

ABSTRACT

This study presents a detailed account of an exceptional case of severe myositis, characterized by an initial creatine phosphokinase (CPK) level of 13,575 U/L, in a 66-year-old female patient with a seven-year history of granulomatosis with polyangiitis (GPA). The patient's condition was marked by significant deterioration in overall health, characterized by muscle weakness in the upper and lower extremities that resulted in her becoming bedridden. Treatment with methylprednisolone was initiated, lead-

ing to a significant decrease in CPK levels, yet the patient's health saw only minor improvement. The patient did not have autoantibodies usually associated with idiopathic inflammatory myositis. The patient ultimately succumbed to complications arising from pneumonia and sepsis a month after admission. This case provides valuable insights into the diagnostic challenges and treatment complexities of severe myositis in patients with GPA.

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KEY WORDS: granulomatosis with polyangiitis; myositis; creatine phosphokinase; vasculitis

INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's Granulomatosis, is a rare systemic vasculitis characterized by inflammation of small- and medium-sized blood vessels, affecting various organs, including the lungs, kidneys, and upper respiratory tract. Here, we present an unusual case of GPA with severe myositis.

CASE PRESENTATION

A 66-year-old female suffering from GPA for seven years presented with a significant decline in overall well-being and muscle weakness in the upper and lower extremities over the past two weeks, eventually becoming bedridden. Recent outpatient laboratory tests revealed significantly elevated serum liver transaminase levels. The patient's rheumatologist

suspected myositis and urgently referred her to the hospital.

The patient was diagnosed with GPA lately, three years after the first symptoms involving paranasal sinusitis. Further, the patient developed left-side deafness and saddle nose deformity until she was referred to a rheumatologist. For two years before described admission, the patient had been receiving rituximab treatment under the national health insurance program. Before initiation of rituximab, the patient was treated with cyclophosphamide, resulting in only minor improvements (the total cumulative dose of the drug is 7,200 mg). Other medical conditions included: arterial hypertension, osteoporosis with compression fractures of the Th11, Th12, and L1 vertebrae, history of two functional endoscopic sinus surgeries (FESS) for the paranasal sinuses, steroid-induced diabetes, glaucoma, cataracts, and a history of trabeculectomy in the left eye.

Address for correspondence:

Mikołaj Kamiński, District Hospital in Koscian, Department of Rheumatology, Szpitalna 7, 64-000 Kościan, Poland; e-mail: mikolaj.w.kaminski@gmail.com

On admission, the patient's creatine phosphokinase (CPK) activity levels were remarkably high at 13,575 U/L, as well as activity levels of lactate dehydrogenase (LDH) reaching 1,003 U/L, aspartate aminotransferase (AST) 730 U/L and alanine aminotransferase (ALT) 571 U/L. White blood count (WBC), C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR) were also elevated at 15,1 G/L, 95.9 mg/L, and 84 mm/h, respectively. Other parameters, e.g., hemoglobin (Hb) levels (13.3 g/dL) and platelets counts (439 G/L), were in the normal range or slightly increased, e.g., creatinine levels (1.4 mg/dL). The patient had increased activity of CA-125 (92.5 U/mL) and serum concentration of carcinoembryonic antigen (CEA, 6.1 ng/mL). However, gynecological examination with vaginal ultrasound, chest X-ray, abdominal ultrasound, and computed tomography of the abdomen and pelvis with contrast found no malignancy.

The patient received methylprednisolone [500 mg intravenously (*i.v.*) per 3 consecutive days), resulting in the quickly diminished activity of CK (two days after admission: 6,710 U/L, five days after admission: 435 U/L, three weeks later on discharge: 19 U/L]. The dose of oral steroids was further reduced to 24 mg of methylprednisolone on the day of discharge. However, pharmacological treatment and rehabilitation provided only a minor improvement, and the patient remained bedridden. Antinuclear antibody (ANA) titer, ANA-5, and ANA-myositis (16 antibodies: anti-Jo-1/EJ/OJ/PL-7/PL-12/SRP/TIF1 γ /Mi-2 α /Mi-2 β /MDA5/NXP2/SAE1/Ku/Ro52/PM-Scl75/PM-Scl100) profiles of the patient were negative. Due to immunosuppression caused by rituximab and steroids as well as poor condition of the patient, we restrained from a muscle biopsy. We did not perform muscles MRI nor electromyography (EMG).

One month later, patients died due to pneumonia complicated by sepsis.

DISCUSSION

We performed a systematic search using the PubMed search engine by using the following query: (“granulomatosis with polyangiitis” OR “Wegener”) AND (“myositis” OR “muscle” OR “myopathy”) NOT “eosinophilic”. The search was performed on 20th April 2023. We retrieved 282 records. Myositis in GPA usually has a form of local inflammation of

extraocular muscles [1]. Conticini et al. performed a systematic review of muscle involvement in systemic vasculitis proved by EMG, magnetic resonance imaging (MRI), or muscle biopsy [2]. In cases analyzed by Conticini et al., none of the patients with GPA and muscle involvement (all concerned lower limbs) had elevated CPK activity [3–5]. Other cases also reported myositis in GPA with normal CPK [6,7]. Hervier et al. Reported one case of biopsy proved muscular vasculitis in the course of GPA with increased CPK activity, but they did not provide an exact value [8]. Figueiredo reported one case of severe pediatric GPA with myositis and CPK was equal to 1,423 U/L [9]. To date, none described a similar case of an adult with GPA and severe myositis with such high CPK activity.

Our case is unusual because the myositis 1) occurred in a patient with a severe, long-standing GPA, and 2) the course of myositis was notably rapid, accompanied by exceptionally high CPK activity upon admission. Therefore we speculate that the patients might have had severe muscle vasculitis or developed idiopathic inflammatory myositis (IIM), e.g., polymyositis. The activity of CPK > 10,000 U/L, limb muscle involvement, and response to methylprednisolone pulses might suggest IIM. Autoantibodies are detected in approximately 60% of individuals with IIM [10], but our patient did not have the characteristic antibodies. Moreover, the lack of histopathological investigation of the inflamed muscle does not allow us to solve this diagnostic dilemma.

AUTHOR CONTRIBUTIONS

Concept: M.K., P.B.; Data collection: M.K., P.H.; Original manuscript preparation: M.K.; Critical review and final manuscript approval: M.K., P.B.

CONFLICT OF INTEREST

Authors declare no conflict of interest.

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ETHICS STATEMENT

This is a retrospective medical record review, and the study did not require Ethical Committee approval.

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