GENERALIZED MYALGIAS WITH DECREASED PROXIMAL STRENGTH - A CASE OF DERMATOMYOSITIS

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ABSTRACT None

KEYWORDS Dermatomyositis, myositis, myalgias

Case report

64-year-old male, previous history of IgA nephropathy since the age of 40, with stage 4 chronic kidney disease, with regular follow-up in nephrology consultation and proposed for peritoneal hemodialysis. History of low-grade papillary carcinoma of the bladder at the age of 52, treated with 6 sessions of chemotherapy without evidence of disease recurrence. Medicated with furosemide 80 mg plus 40 mg, amlodipine 5 mg and alfazosin 10 mg.

Observed in the emergency department for generalized myalgias with 3 months of evolution with decreased proximal strength in the upper limbs and 15 days with decreased proximal strength in the lower limbs and worsening of myalgias, becoming disabling with limitation in gait and in carrying out activities of daily living. He has also reported bilateral palpebral erythema for 1 week and describes symptoms suggestive of Raynaud’s phenomenon. On objective examination summary neurological examination with proximal strength in the upper limbs G2 and proximal strength in the lower limbs G3. From the study performed hemoglobin 11g/dL, AST 915 U/L, ALT 480 U/L, Creatine Kinase (Ck) 21615 U/L, myoglobin >12000ng/mL, urea 228 mg/dL, creatinine 3.66mg/dL (basal 2,8mg/dL), urine II proteinuria. Arterial blood gases without changes. Abdominal and reno-vesical ultrasound without liver changes and pyelocaliceal dilatation. Internment for study.

On the third day of hospitalization, a heliotrope rash and gottron’s papules were documented (Figure 1 and 2), so the study was directed to dermatomyositis.

Autoimmune study with ANA 1/1000, positive anti-Mi2 antibody, negative anti-ENA antibody (anti Jo1; RNP; SCL70; SM; SSb; SSA) and negative antiKu. Search for HIV, HCV, HBV negative, negative serology for borrelia, parovirus B19, rickettsia, TPPA/TP, thig, CMV, leptospira. Normal thyroid function. Search for negative occult neoplasia with upper digestive and lower digestive endoscopy and cervico-thoracic-abdominopelvic computerized tomography without suspicious lesion.

Electromyography is concordant with myositis and muscle biopsy with evidence of nonspecific severe inflammatory myopathy, characteristic of dermatomyositis or necrotizing myopathy.

Started methylprednisolone 1mg/kg/day, gastric protector and opportunistic infection prophylaxis. Progressive improvement in renal function and rhabdomyolysis, after 10 days, he was discharged and guided for consultation. After a week, he maintained a favourable evolution in the first reassessment consultation. In the reassessment at the 4th week with clear clinical worsening with disabling myalgias, generalized decrease in strength and again with dysphagia for solids, he was hospitalized again for a pulse of methylprednisolone and onset of azathioprine. He repeated an occult neoplasm study that remained negative. Pneumococcal 13-valent conjugate vaccine and hepatitis B vaccine were administered; Interferon Gamma Release Assay (IGRA) was inconclusive. Due to severe pancytopenia, suspend azathioprine and it was not possible to start methotrexate due to hepatic and renal involvement, or mycophenolate mofetil due to neutropenia, and he took 2 doses of human immunoglobulin 1g/kg with clinical improvement. However, after 5 days, he initiated hypoxemic and hypercapnic respiratory failure. Due to the inconclusive IGRA result, with no increase in inflammatory parameters, it was delayed to start rituximab. Unfortunately, the patient died due to respiratory and cardiac involvement of the myopathy at the end 2 months of dermatomyositis diagnosis.

Figure 1: Heliotrope rash. Figure 2: Gottron’s papules more evident in the left hand.

Discussion/Conclusion

Dermatomyositis is paraneoplastic in 80% of cases, and about 50% of cases appear in adults over 65 years of age. The diagnosis of dermato-
myositis increases the risk of neoplasia by 3-6 times. The neoplasm is present at diagnosis or in the 1st year after diagnosis in 68% of patients, and if not present at diagnosis, it usually occurs within the first 3 years after diagnosis [1,2]. The most frequent neoplasms are breast, lung, pancreas, stomach, colon, ovary and Hodgkin lymphoma. Cancer screening should be performed every 3 years before suggestive symptoms [1]. The presence of anti-Jo and anti-Mi2 antibodies is not usually associated with neoplasia [3]. In the case presented, despite the anti-Mi2 antibody, which provides a good prognosis, the outcome was fatal. We attribute this outcome to the involvement of cardiac and pulmonary muscles due to lack of disease control, the limitation in the use of immunomodulators due to renal and hepatic involvement, and neutropenia secondary to azathioprine. Therefore, it was most likely a case of paraneoplastic dermatomyositis; however, in the extensive study carried out, it was impossible to efficiently detect and treat the neoplasm, which would be essential for controlling the control dermatomyositis.

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**Conflict of interest**

There are no conflicts of interest to declare by any of the authors of this study.

**References**

