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SATISFACTORY RESPONSE OF RITUXIMAB (ANTIBODY ANTI-CD20) ASSOCIATED WITH IMMUNOGLOBULIN AND METHYLPREDNISOLONE PULSE IN TEENAGER WITH REFRACTORY JUVENILE DERMATOMYOSITIS

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BACKGROUND

Juvenile Dermatomyositis (JDM) belongs to a heterogeneous group of

inflammatory myopathies with onset during childhood, characterized by proximal muscle weakness and pathognomonic skin lesions (heliotrope and/or Gottron's papules). The disease's progression bears correlation to the severity of one's vasculopathy. Most patients present a non-progressive and self-limited form, with complete remission of symptoms within a few months. However, some patients manifest a more persistent and chronic variant. High dose of corticosteroids in association with other immunosuppressants increased the survival of these patients. Moreover, the advent of biological drugs has transformed the management of JDM patients.We report a case of refractory JDM with a satisfactory response to rituximabe, a chimeric monoclonal antibody against the protein CD20.

CASE REPORT

LRN, 14-year-old female, presented with proximal muscle weakness at the age of 10, typical skin lesions (heliotrope and Gottron's papule), elevated muscle enzymes, electroneuromyography compatible with JDM. Corticoid therapy in high doses associated with methotrexate achieved satisfactory results initially. Nonetheless, a relapse occurred six months later and cyclophosphamide was added. After the second dose of this srug, the patient's condition degraded into severe septicemia, requiring immediate intervention with immunoglobulin. The following nine months, due to the lack of improvement and in order to discard differential diagnoses (e.g. other myopathies), a muscle biopsy with immunohistochemistry and nailfold capillaroscopy were performed and the diagnosis of JDM confirmed. The immunoglobulin was suspended and azathioprine was introduced instead. After 4 months the patient's condition improved and remained stable for 18 months; despite that, another recurrence clinical (proximal muscle weakness, palpebral ptosis, dysphagia and interstitial pneumonitis) and laboratorial worsening (CPK 3272, LDH 965, aldolase 20, ESR 100 e CRP 52) was observed. Azathioprine was suspended, methotrexate maintained and immunoglobulin was reintroduced at 2g/Kg, along with methylprednisolone at 30 mg/Kg/day pulse therapy over the course of 3 days; on the fourth day, rituximab was given at 375 mg/m². The same therapeutic regimen was repeated after 15 and 180 days. After the second dose, clinical and laboratorial improvement were visible, as follows: CPK 239, LDH 280, aldolase 6, ESR 5 and CRP 3.

CONCLUSION

This case report demonstrates the efficacy of associating immunoglobulin and methylprednisolone pulse with rituximab, in the management of severe refractory JDM. There are a few randomised controlled trials

related the use of rituximab in the treatment of these conditions, but it seems to be a possible future therapy of JDM.