Intravenous human immunoglobulin for the treatment of recurrent focal myositis

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Recurrent focal myositis (RFM) is a rare inflammatory myopathy (IM) presenting with localized pain, tenderness and oedema, predominantly in lower extremities[1,2] In general, RFM improves spontaneously and its treatment is mainly symptomatic with analgesics and non-steroidal anti-inflammatory drugs (NSAIDs)[1,3].

We present a clinical case of refractory RFM despite treatment with corticosteroids and immunosuppressive therapies, in which treatment with intravenous human immunoglobulin (IVIG) was performed.

Teixeira *et al.* has previously described the clinical case of a patient with RFM followed in our department[2]. This 42-year-old woman was diagnosed with RFM 8 years ago. Initial complaints were controlled with NSAIDs. However, in the following 5 years, she had recurrent episodes of local pain and swelling involving different muscle groups, with increased sedimentation rate (ESR) and C-reactive protein (CRP). By this time, the patient was treated with prednisolone and methotrexate up to 25mg/week without benefit. Posteriorly, azathioprine was tried up to 2mg/Kg/day with transient response that lasted less than a year. Due to clinical worsening (left forearm) and lack of response to corticosteroids and immunosuppressants, the patient was treated with IVIG, taking into account data available for polymyositis (PM) and dermatomyositis (DM)[4–8]. Treatment with IVIG 1g/kg/day for 2 days was started and repeated monthly for 6 months. The patient showed clinical and laboratorial improvement soon after the first cycle. Azathioprine was tapered to 100mg/day and prednisolone was reduced until discontinuation and the disease remained in remission. One year after the end of IVIG treatment, there was a new recurrence with involvement of left peroneal, soleus and anterior tibial muscles. It was decided to restart IVIG 1g/Kg per day for 2 days at fixed intervals of 12 weeks that were enough to restore remission which was sustained after 1 year of treatment.

This patient corresponded to a typical case of focal myositis at initial clinical presentation. However, further involvement of several muscles in upper and lower limb in different occasions is infrequent. Some authors describe that focal lesions may progress to a more generalized PM mainly when there is more than one muscle involved, early rise in ESR, CPR or an early recurrence after the first episode[1,2,9]. However, in this patient there was no evidence of PM or other systemic disease during follow-up.

In RFM, recurrent disease that does not respond to analgesics and NSAIDs should be treated with corticosteroids and immunosuppressants, such as azathioprine and methotrexate. The use of IVIG has been a therapeutic option in severe, refractory and rapidly progressive cases in DM and PM[6,7,10]. To the best of our knowledge, this is the first report of IVIG use in RFM. This clinical case reveals a good response to treatment with IVIG, as described for PM and DM[6,8].

At the moment, the IVIG's mode of action is not fully clarified and no definitive guidelines are available regarding dose, total days of administration or timing of subsequent administration for IM. Several studies suggested IVIG at a dose of 2g/kg over a 2-day or 5-day period, followed by monthly doses for 3 to 6 months[4,6–8,10].

The use of IVIG has been proven effective in the treatment of adult patients with PM/DM especially those with refractory, flare-up, rapidly progressive or severe disease. Difficulty in establishing conclusive results seems to be due to the rarity and heterogeneity of these diseases.

This case reports that the use of IVIG can be effec-

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tive in RFM and this treatment may be considered among other treatment options in refractory IM that are unresponsive to other immunosuppressive therapies, namely in RFM. However, further studies are needed to confirm the effectiveness of the drug and to evaluate long-term administration effects.

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