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Novel treatment (new drug/intervention; established drug/procedure in new situation)

Refractory calcinosis in a patient with dermatomyositis: response to intravenous immune globulin

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Summary

Dermatomyositis is an inflammatory myopathy and commonly presents with progressive, symmetric proximal muscle weakness and cutaneous findings. Calcinosis is a severe manifestation that can be debilitating. The cutaneous manifestations of dermatomyositis may also develop in the absence of detectable muscle disease, and can persist after the successful treatment of myositis. The author reports a 30-year-old woman with biopsy-proven dermatomyositis who had failed previous trials of azathioprine and methotrexate. Her muscle weakness was controlled with mycophenolate mofetil and prednisone; however, she had recurrent attacks of painful calcinosis. The patient responded to intravenous immune globulin (IVIG) along with intravenous methylprednisone, followed by IVIG for 2 consecutive days each month. This regimen has been effective in preventing recurrence of her calcinosis. This case illustrates the cutaneous manifestation of dermatomyositis, which is often more refractory to treat as compared to the muscle involvement and require additional approaches such as IVIG.

BACKGROUND

Dermatomyositis is an idiopathic inflammatory myopathy and commonly presents with progressive, symmetric proximal muscle weakness and a group of characteristic cutaneous findings. The cutaneous findings seen typically with dermatomyositis include the Gottron's sign, shawl sign, helitrope rash and generalised erythroderma. Calcinosis is a severe manifestation of the disease which can be debilitating in many patients. The cutaneous manifestations of DM may also develop in the absence of detectable muscle disease, and can persist after the successful treatment of DM-associated myositis.[1]

CASE PRESENTATION

The author reports a 30-year-old woman who presents to her rheumatologist for new developing tender nodules. The painful nodules were present in the pannus of her abdominal folds and her medial thighs bilaterally, on her right and left arms and above her sacral area. These lesions developed over the past 3 days and represented the previous episodes of calcinosis. The patient had been diagnosed with dermatomyositis a year ago by a muscle biopsy during a work-up of muscle pain and growing skin lesion. The biopsy had demonstrated pattern of fibre atrophy that was predominantly perifascicular. Additionally, there are small foci of endomysial lymphocytic infiltrates. The patient has had good control of her muscle weakness related to her dermatomyositis over the past year with mycophenolate mofetil 1 g twice a day and prednisone 10 mg daily. She had failed previous trials of a combination of azathioprine and methotrexate and had recurrent muscle weakness around the proximal muscles of the hips and the shoulder girdle. The other cutaneous signs for dermatomyositis in this patient were the heliotrope rash on the upper eyelids and the diffuse, flat erythematous lesion occurring over the chest and shoulders, also known as the shawl sign. She did not have any other comorbid medical conditions and was not on any other medication that could contribute to her dermatological manifestation. She had a 10-pack-year history of smoking with occasional alcohol use; however she denied any illicit drug use.

DIFFERENTIAL DIAGNOSIS

However, even with the control of the muscle weakness, which is classic for dermatomyositis, she did develop these calcinosis recurrently which were painful. They were differentiated from an infectious process such as cellulitis by a skin biopsy, which showed benign reactive skin and granulation tissue with fat necrosis, and dystrophic calcifications.

TREATMENT

Considering previous failure to the first-line agents for skin manifestation of dermatomyositis, the patient was tried on intravenous immune globulin (IVIG) after discussing the off-label use for this condition. The patient was given IVIG 60 g (1 g/kg per day) for 2 days along with intravenous methylprednisone 40 mg. The patient responded well to this regimen and had significant improvement in her calcinosis.

OUTCOME AND FOLLOW-UP

To prevent recurrence the patient has been continued on the same dose of IVIG for 2 consecutive days in a month, for the past 6 months and this has been effective in preventing any recurrence of her cutaneous manifestation. The patient is also continued on her regimen of mofetil 1 g twice a day and prednisone 10 mg daily.

DISCUSSION

Management of cutaneous disease in DM can be challenging, and cutaneous manifestations are often more resistant to therapy than concomitant muscle involvement. The efficacy of IVIG was illustrated in a double-blind, placebo-controlled trial of 15 patients with DM who had resistant disease; however, the mechanism of action is not known.[2] Patients were given IVIG (2 g/kg) or placebo once monthly for three consecutive months. Marked improvement in skin disease occurred in 8 out of 12 patients treated with IVIG. Improvements in cutaneous disease preceded or coincided with muscle improvement, which was evident approximately 15 days after the first infusion of IVIG and peaked between the second and third infusions. A case report of a similar patient responding to IVIG (2 g/kg/month) at a dose of 0.4 g/day for 5 consecutive days with complete resolution of the cutaneous symptoms adds to our background knowledge.[3] The patient described in the earlier case report received maintenance treatment with a yearly course of IVIG and has not had recurrence in her disease during a 5-year follow-up. Our patient has been receiving IVIG monthly for the past 6 months without any recurrence of her disease. The above case illustrates the cutaneous manifestation of dermatomyositis, which is often more refractory to treat as compared with the muscle involvement and requires additional approaches such as IVIG.

Learning points

- Dermatomyositis is an idiopathic inflammatory myopathy and commonly presents with progressive, symmetric proximal muscle weakness and a group of characteristic cutaneous findings.
- Calcinosis is a severe manifestation of the disease which can be debilitating in many patients.
- The cutaneous manifestations of DM may also develop in the absence of detectable muscle disease, and can persist after the successful treatment of DM-associated myositis.
- Management of cutaneous disease in DM can be challenging, and may require additional approaches such as intravenous immune globulin.

Competing interests None.

Patient consent Obtained.

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