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Case Report

Dermatomyositis-related intestinal dysmotility

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ABSTRACT

Dermatomyositis (DM) is an inflammatory myopathy (IIM) characterized by proximal muscle weakness and pathognomonic skin lesions. A 69-year-old woman with a recent diagnosis of DM 1 month prior, treated with corticosteroids and immunomodulators, presented to our inpatient rehabilitation with worsening dysphagia and constipation. At the time of our evaluation, physical examination was notable for erythematous papules over the metacarpophalangeal joints, proximal interphalangeal joints, elbows, and knees as well as a violaceous rash on the face. Muscle strength was diminished bilaterally with proximal distribution being affected greater than distal. Laboratory studies were notable for the creatine kinase (CK) level of 31 IU/l, antinuclear antibodies (ANA) by immunofluorescence of 1:80, and aldolase 4 u/l. The 11-antibody myositis panel was negative showed partially treated acquired IIM with perifascicular atrophy. During hospitalisation, she was found to have pulmonary embolism. She received enoxaparin 1 mg/kg subcutaneous BID. Soon after, she developed rectal bleeding. Colonoscopy showed a stercoral ulcer caused by chronic constipation. While dysphagia is common, being present in 25–50% of patients with DM, lower gastrointestinal problems involving the small and large intestine are rare and typically present as a late manifestation of the disease. Decreased peristalsis in the large colon can lead to constipation, impaction, and subsequent mucosal ulceration, and pressure necrosis induced by faecaloma formation. Although rare, our case highlights the importance of recognising gastrointestinal complications that DM can cause and the effects that those complications have on morbidity and mortality.

KEYWORDS: Dermatomyositis; intestinal dysmotility; myopathy-related constipation; gastrointestinal bleeding

Introduction

Dermatomyositis (DM) is an idiopathic inflammatory myopathy (IIM) that is caused by immune-mediated muscle inflammation characterised by proximal muscle weakness and skin involvement typically affecting the extensor surfaces of the hands and face. Given its protean clinical manifestations and recently recognised serological profiles, different subtypes have been identified for the classification of DM. DM usually appears in the adolescent period and between 40 and 60 years of age, with a female predominant pattern [2, 10]. The aetiology of DM has been thought to be idiopathic or associated to an underlying neoplastic process [2]. Other organ manifestations have also been described, including the blood vessels, lungs, oesophagus, joints [3], and rarely the lower gastrointestinal tract, affecting the motility of the colon [4]. We present a case of DM associated with slow bowel transit inducing constipation and subsequent gastrointestinal bleeding in an otherwise healthy woman.

Case report

A 69-year-old woman presented to her primary care physician due to inability to comb her hair secondary to left upper extremity weakness and a new onset rash to the extensor surfaces of metacarpophalangeal joints (MCPs), proximal interphalangeal joints (PIPs), elbows, knees, and her face. The patient underwent laboratory tests, which showed an ANA titre 1:640 by immunofluorescence, negative anti-Jo-1 and anti-Mi2 antibodies. Given her clinical presentation, the patient was diagnosed with inflammatory myositis. The patient was started on pulse methylprednisolone at 1 mg/day for 3 days. Following completion of the pulse therapy, the patient was sent home to continue prednisone 50 mg orally daily along with azathioprine 50 mg orally twice daily. The patient did have an improvement in her rash and some of her proximal muscle weakness; however, about 1 month later the patient was taken to an outside hospital due to worsening dysphagia and severe constipation. The patient underwent laboratory tests, which showed normal levels of creatine kinase (CK) of 34 IU/l, the sedimentation rate of 32 mm/h, aldolase of 2.7 u/l, negative anti-smith, anti-double stranded-DNA, anti-RNP, anti-SS-A, anti-SS-B, and anti-neutrophilic cytoplasmic antibodies. At that time, the patient was found to have severe fecal impaction (Figures 8–9) and colonoscopy was attempted three times due to poor bowel preparation and the patient had to be manually disimpacted. The patient eventually underwent a successful colonoscopy with polypectomy of three polyps. Pathology showed that all three polyps were benign adenomas. Due to continued dysphagia, the patient underwent an esophagogastroduodenoscopy, which showed an oesophageal ring. The patient

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subsequently underwent jejunostomy tube placement. The patient was transferred to our inpatient rehabilitation centre for physical therapy 1 month after her initial presentation, for which our rheumatology team was consulted. Upon our questioning, the patient noted that she was well up until about one and a half months prior to current admission. Prior to that, the patient did endorse an unintentional 30-lb weight loss but did not have any night sweats, cough, haemoptysis, haematochezia, vaginal bleeding, melena, early satiety, or any other worrisome symptoms. She reported that all her age-appropriate cancer screenings were performed in Mexico and that everything was 'normal'. Our physical examination was notable for erythematous papules over the MCPs, PIPs, elbows, and knees as well as a violaceous rash on the face not sparing the nasolabial folds (Figures 10–11). Her muscle strength was diminished bilaterally in a proximal and distal distribution; however, proximal was more affected being 3/5 strength compared to 4/5 strength, respectively. Laboratory studies were notable for a TSH of 22 uIU/ml, the CK level of 31 IU/l, the sedimentation rate of 34 mm/h, ANA by immunofluorescence of 1:80, negative anti-Scl-70 antibody, and aldolase 4 u/l. The 11-antibody myositis panel showed negative anti-EJ, anti-Jo-1, anti-MDA-5, anti-Mi-2 Alpha, anti-Mi-2 Beta, anti-NXP-2, anti-OJ, anti-PL-12, anti-Pl-7, anti-SRP and anti-TIF-1 gamma antibodies. Skeletal muscle biopsy of the left thigh was performed, which showed partially treated acquired IIM with perifascicular atrophy (Figures 1–5). The patient had a calculated myositis diagnostic score based on the European League Against Rheumatism (EULAR)/American College of Rheumatology (ACR) Classification Criteria for Idiopathic Inflammatory Myopathies of 18.8 out of 19 with a 100% probability of IIM. Due to continued weakness, the patient was started on methylprednisolone 40 mg IV every 24 h along with azathioprine 50 mg BID. During the hospital stay, the patient began to complain of shortness of breath and was noted to be tachycardic. Venous Doppler of bilateral lower extremities was performed along with chest angiogram, which showed bilateral lower extremity deep venous thrombi and multifocal pulmonary emboli. The patient was started on full dose anticoagulation with enoxaparin at 1 mg/kg subcutaneous BID. Four days after starting treatment, the patient was transferred from inpatient rehabilitation to our emergency department due to

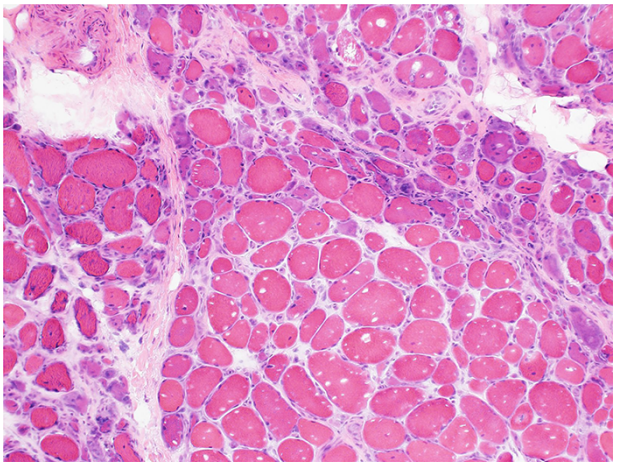

Figure 1. H&E stain showing a mild-to-moderate increase in perimysial connective tissue, with variation in the fibre diameter due to the presence of frequently angulated and atrophic muscle fibres, frequent regenerating muscle fibres, and occasional necrotic muscle fibres.

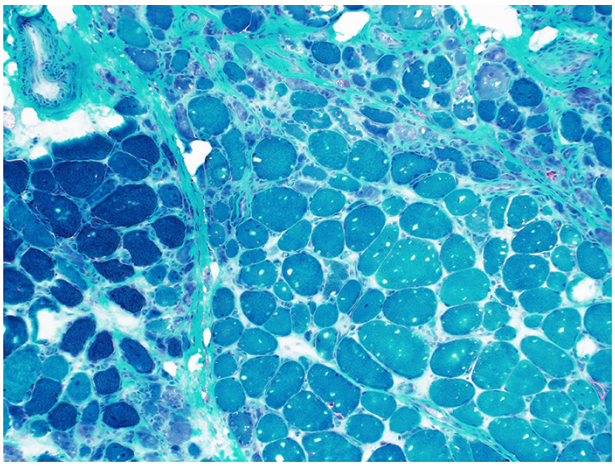

Figure 2. No abnormal inclusions or rimmed vacuole-type structures noted on modified gomori trichome preparation.

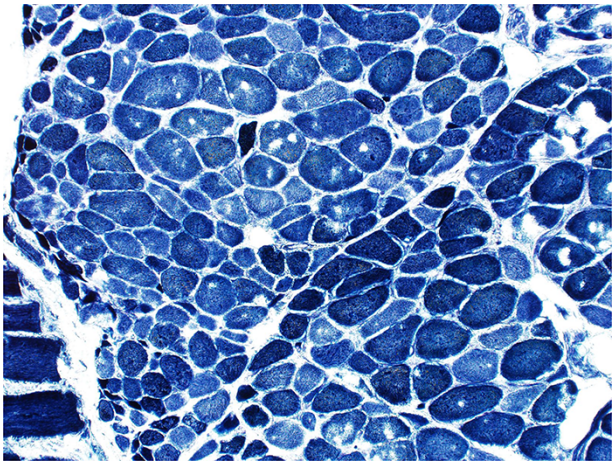

Figure 3. NADH-TR stain showing occasional targetoid muscle fibres.

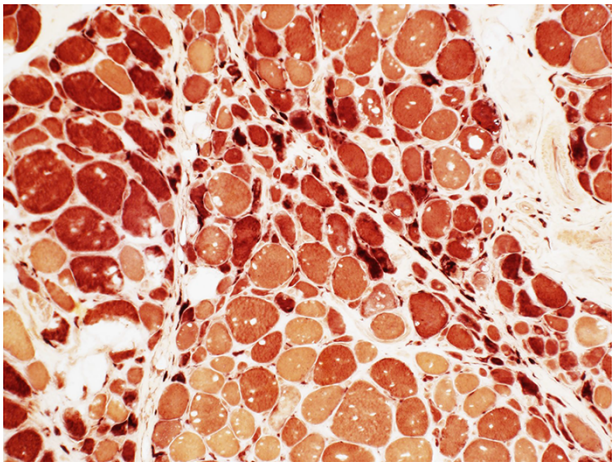

Figure 4. Esterase stain showing atrophic muscle fibres and highlights and occasional macrophages within the perimysium.

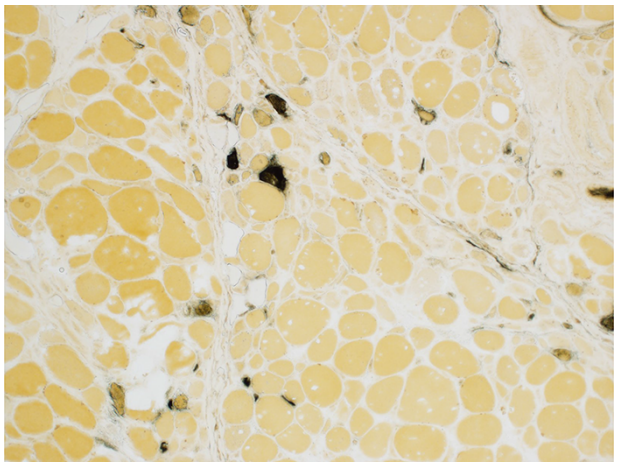

Figure 5. Alkaline phosphatase preparation demonstrates a patchy mild increase in capillary staining and highlights regenerating necrotic muscle fibres located in the perifascilar distribution.

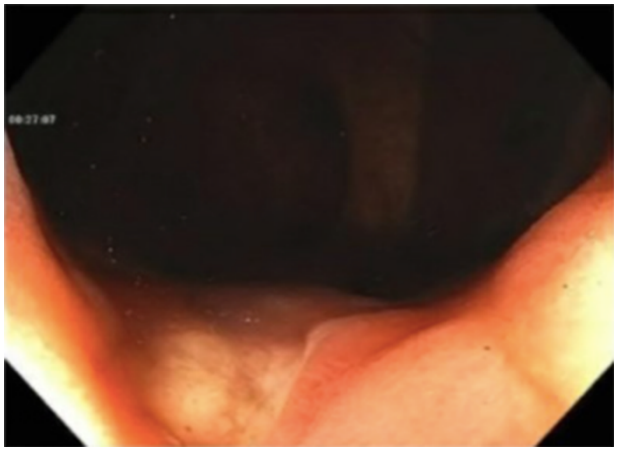

Figure 6. Colonoscopy showing rectal ulcer.

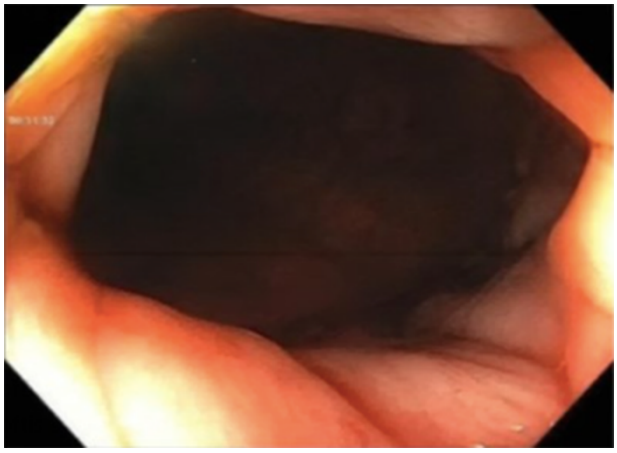

Figure 7. Colonoscopy showing rectal ulcer.

haematochezia. The patient was found to have a decrease in her haemoglobin from 11 to 8.8 g/dl. She underwent a repeat colonoscopy and was found to have a 1-cm rectal ulcer (Figures 6–7) with hallmark features consistent with a

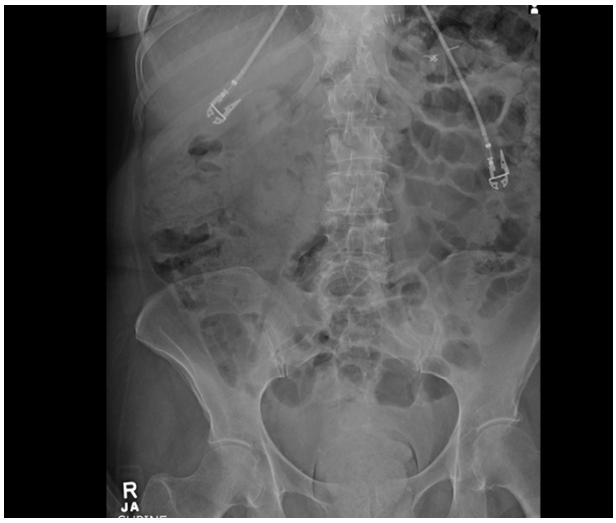

Figure 8. Abdominal XR showing evidence of constipation.

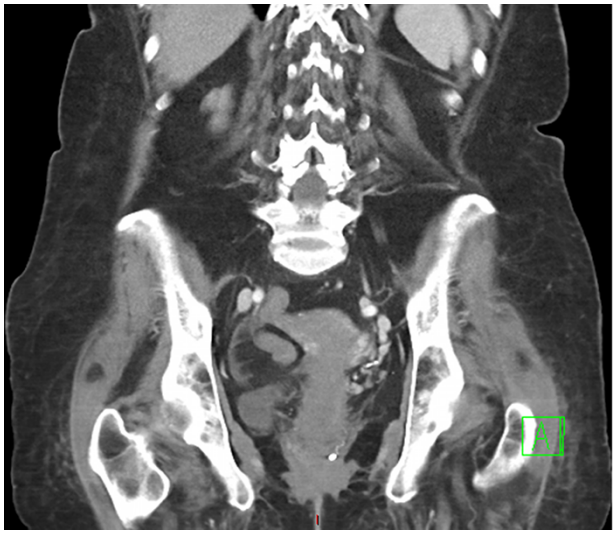

Figure 9. CT abdomen/Pelvis showing large stool burden with fecal impaction.

stercoral ulcer suspected to be caused by chronic constipation secondary to impaired peristalsis in the setting of DM. Anticoagulation was discontinued and the patient underwent the placement of an inferior vena cava (IVC) filter. Computed tomography scan results of abdomen and chest were negative for masses or lymphadenopathies. The patient continues treatment with methylprednisolone 40 mg IV twice daily via jejunostomy, methotrexate 7.5 mg weekly, and azathioprine 50 mg twice daily. She has not had recurrence of rectal bleeding; however, she continues to have difficulty with constipation and dysphagia. Currently, she can swallow thick liquids such as pudding. Her limb involvement has significantly improved, and she is now able to ambulate without assistance. Even though there was no identifiable malignancy found, the presence of venous thrombus emboli increases suspicion for malignancy that has not yet been identified. The malignancy can precede, is concurrent with, or follows the diagnosis of DM.

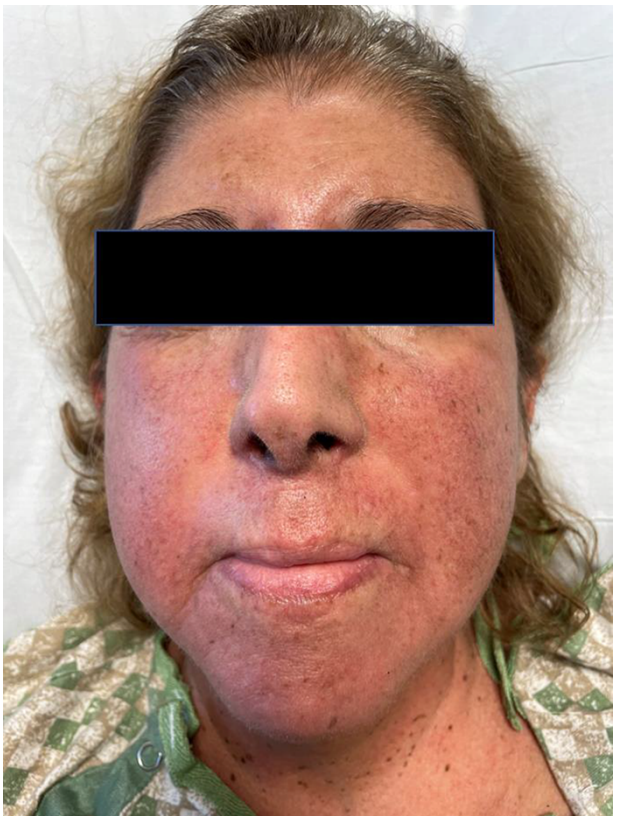

Figure 10. Violaceous rash not sparing nasolabial folds.

Figure 11. Erythematous papules over PIPs and MCPs.

Discussion

DM is an inflammatory muscle disease that typically affects skeletal muscles and presents proximal weakness. The characteristic skin manifestations include rash particularly in the dorsa of the hands, face, V of the neck, lateral aspect of the thighs, and periungual erythema or possible ulcerated papules. Patients can also manifest occasional fever, weight loss, arthralgia, and gastrointestinal abnormalities. The latter usually refers to motility issues that arise from the striate muscle in the oesophagus and typically presents as dysphagia. While dysphagia is common being present in 25–50% of patients with DM, lower gastrointestinal problems involving the small and large intestine are rare and typically present as a late manifestation of the disease. DM-related dysphagia is caused by inflammatory involvement of the muscles involved in swallowing. According to the literature, DM-related dysphagia can be classified as proximal versus distal based on the type of muscle involved. Proximal dysphagia occurs due to the involvement of striated muscles of the pharynx or upper one-third oesophagus whereas distal dysphagia involves smooth muscle and is typically associated with other connective tissue diseases such as systemic sclerosis [5]. The involvement of the upper oesophagus and cricopharyngeal muscle leads to dysphonia and difficulty swallowing. In contrast, distal involvement presents with delayed gastric emptying and dysmotility, which can cause recurrent abdominal pain and bloody diarrhoea. Radiologically these can manifest as dilated atonic oesophagus, thickening of the small bowel wall, or 'stacked coin' appearance. There are four degrees of dysphagia in DM based on the patient's symptoms and clinical findings ranging from nonobvious complaints up to severe dysphagia requiring feeding tubes. Our patient's dysphagia would be classified as degree 4 as she required the placement of jejunostomy tube for nutrition. It is important to note that dysphagia not only decreases the quality of life in patients diagnosed with DM, but it can also increase mortality due to complications such as aspiration pneumonia and malnutrition [5]. Immunosuppressive therapy with corticosteroids and intravenous immune globulin (IVIG) significantly improves proximal dysphagia; however, distal dysphagia treatment is more challenging and further research needs to be conducted to determine the best treatment approach [1, 9].

As mentioned above, lower gastrointestinal involvement in DM is rare; however, there have been a few cases published in the literature. Gadiparthi and associates described an association between inflammatory myopathies and intestinal pseudo-obstruction, which clinically presented with recurrent obstructive symptoms but lacked evidence of a mechanical obstruction [6]. Levesque *et al.* described a case of a young patient with DM who presented with dysphagia and melena and was found to have multiple ulcerations in the oesophagus and the duodenum. Unlike our patient, histopathology of the ulcerations showed evidence of vasculitis [7]. Several other cases of lower gastrointestinal tract involvement have been reported including the study by Tweezer-Zaks *et al.* who performed a retrospective review on the clinical course of 48 patients diagnosed with polymyositis and DM and identified patients with severe gastrointestinal manifestations. Of the 48 patients within the study population, only

three patients presented with lower gastrointestinal manifestation, which included findings of the oedematous hyperaemic bowel wall with multiple erosions and ulcerous lesions on endoscopy. Histologically, these lesions consisted of diffuse mucosal inflammation and multiple vascular ectasias without evidence of vasculitis [8]. Similarly, our patient's colonoscopy showed a stercoral ulcer at the rectum, which resulted in lower gastrointestinal bleeding. Muscular inflammation in DM can impair peristalsis throughout the gastrointestinal tract. Decreased peristalsis in the large colon can lead to constipation, faecal impaction, and subsequent mucosal ulceration, and pressure necrosis induced by faecaloma formation. Our patient was being treated with full dose anticoagulation for bilateral lower extremity deep venous and pulmonary emboli. This along with severe faecal impaction was the underlying mechanism that resulted in haematochezia in our patient.

Conclusion

Although rare, our case highlights the importance of recognising gastrointestinal complications that DM can cause and the effects that these complications have on morbidity and mortality. Patients admitted in the inpatient setting with DM should have an aggressive bowel regimen to help decrease complications from intestinal dysmotility. It is also important that these patients work with physical therapy and occupational therapy to decrease the risk of functional debility. The literature is limited regarding DM gastrointestinal complications; thus, it is important to report similar cases to determine the best treatment strategies to mitigate complications such as the one presented here.

Conflict of interest

None declared.

Funding

The study was performed while employed by the University of Texas Rio Grande Valley School of Medicine.

Patient consent

Patient has given signed consent, using the Doctors Hospital at Renaissance Health Institute for Research and Development, to participate in this case report. Date consent obtained 04/05/21.

Ethical approval

Not applicable.

References

[1] Dourmishev LA, Dourmishev AL. *Dermatomyositis Advances in Recognition, Understanding and Management*. 1st edn. Berlin, Heidelberg: Springer, 2009.

[2] Mainetti C, Terziroli Beretta-Piccoli B, Selmi C. Cutaneous manifestations of dermatomyositis: a comprehensive review. *Clinic Rev Allergy Immunol* 2017;**53**:337–56.

[3] Bogdanov I, Kazandjieva J, Darlenski R *et al.* Dermatomyositis: current concepts. *Clin Dermatol* 2018;**36**:450–8.

[4] Yamada C, Sato S, Sasaki N *et al.* A case of dermatomyositis and anti-ej autoantibody with chronic intestinal pseudoobstruction successfully treated with octreotide. *Case Rep Rheumatol* 2016;**2016**:9510316.

[5] Labeit B, Pawlitzki M, Ruck T *et al.* The impact of dysphagia in myositis: a systematic review and meta-analysis. *J Clin Med* 2020;**9**:2150.

[6] Gadiparthi C, Hans A, Potts K *et al.* Gastrointestinal and hepatic disease in the inflammatory myopathies. *Rheum Dis Clin North Am* 2018;**44**:113–29.

[7] Marie I, Levesque H, Cailleux N *et al.* Intravenous immunoglobulin for treatment of gastro- intestinal haemorrhage in dermatomyositis. *Ann Rheum Dis* 2001;**60**:723–4.

[8] Tweezer-Zaks N, Ben-Horin S, Schiby G *et al.* Severe gastrointestinal inflammation in adult dermatomyositis: characterization of a novel clinical association. *Am J Med Sci* 2006;**332**:308–13.

[9] Fukunaga E, Kunishigea M, Mitsuia T *et al.* Severe dermatomyositis with rhabdomyolysis and paralytic ileus: a case successfully treated with plasmapheresis and intravenous immunoglobulin. *EFNS Eur J Neurol* 2002;**9**:687–702.

[10] Senécal J, Raynauld J, Troyanov Y. Editorial: a new classification of adult autoimmune myositis. *Arth Rheumatol (Hoboken, NJ)* 2017;**69**:878–84.