CASE REPORT

Colonic vasculopathy and perforation in the initial presentation of adult dermatomyositis in a patient with improving muscle weakness

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SUMMARY

A 63-year-old woman with diabetes presented with 8 weeks of proximal muscle weakness and change in bowel habits. Muscle biopsy confirmed myositis, and serological studies were consistent with dermatomyositis (DM), without evidence of overlapping connective tissue disease or malignancy. On day 12 of prednisone therapy and after receiving one dose of IVIG with improvement in muscle strength, the patient developed abdominal pain and was diagnosed with a gastrointestinal (GI) perforation and peritonitis requiring emergent colectomy. The pathology revealed diffuse mucosal ulceration, prominent lymphoplasmacytic infiltration, venous occlusion and arterial hyperplasia. Although GI manifestations due to GI vasculopathy are rare in adult DM and are often a delayed complication, in this patient, it was one of the initial manifestations of this condition. In addition to being a fatal complication, clinicians should be aware of these complications, as immunosuppression used to control the muscular and cutaneous inflammation may not control the GI vasculopathy.

We report a case of adult DM in a patient with type 1 diabetes who presented with subacute proximal muscle weakness and change in bowel habits. While on IVIG and steroids, the patient developed a GI perforation and peritonitis and was found to have diffuse vasculopathy.

CASE PRESENTATION

A 63-year-old Caucasian woman presented with bilious, non-bloody emesis for a few days and profound upper extremity weakness and oedema. Her history was remarkable for imperfectly controlled type 1 diabetes, hypertension, hypothyroidism, obesity and constipation. Two months prior to admission, she developed a rough, red, scaly plaque, measuring 10×20 cm, on the dorsal aspect of her left arm, which then spread to her posterior neck. The rash resolved within a few days but she began noting bilateral proximal upper extremity weakness, and within weeks, she could not raise her arms to put her glasses on or comb her hair. At the same time, she noted a change in consistency of her bowel movements, from well-formed to loose.

BACKGROUND

Gastrointestinal (GI) inflammation has long been recognised in patients with juvenile dermatomyositis (JDM), but rarely in adult DM.^{1 2} The most common GI manifestation in JDM is dysphagia, present in 25–50% of patients. Unlike distal dysphagia, which is treatment-resistant and mostly occurs in overlap syndromes, proximal dysphagia generally responds to steroids. Less commonly, ulceration and perforation of the upper GI tract³ and pneumatosis intestinalis⁴ can occur secondary to vasculitis and thrombosis of upper GI blood vessels.

When present in adults, inflammation of the small arteries of the GI tract is generally mild, secondary to damage to smooth muscle of the myenteric system, leading to ineffective peristalsis and motility disorders.⁵ This can occur years after initial presentation and presents with non-specific symptoms such as abdominal pain, distension, change in bowel habits and dysphagia.² Some of these manifestations resolve with time⁶ or respond to continued immunosuppression.⁷ However, rarely, they lead to severe complications, usually in patients with overlap syndrome. Very little attention has been given to those cases of severe GI complications in patients with DM without evidence of overlap.⁵

INVESTIGATIONS

On examination, she had mild facial erythema with periorbital violaceous patches (figure 1A). On her left dorsal forearm was an ill-defined thin, pink plaque with overlying thin scale (figure 1B). She had 3/5 strength in deltoids and biceps, 4/5 in triceps, forearms and wrists, and 4/5 strength in the lower extremities. She had significant pitting oedema in both upper extremities, and chronic pitting oedema in the lower extremities. Breast and lymph node examinations were negative. The patient had undergone a screening colonoscopy 10 years prior and a Pap smear 5 years prior, which were both negative. Labs were significant for creatine kinase (CK) 2480 U/L, erythrocyte sedimentation rate 58 mm/h, C reactive protein 82.7 mg/dL, aldolase 12.9 U/L, albumin 2.7 g/dL and thyroid-stimulating hormone 2.81 U/mL.

An inflammatory myositis was suspected, and the patient was treated initially with 40 mg intravenous methylprednisolone and one treatment of 2 mg/kg IVIG. MRI of the upper extremities with contrast revealed myositis of the bilateral shoulder girdles (figure 2A). Biopsy showed diffuse lymphocytic infiltration with slight perivascular predominance (figure 2B). On immunohistochemistry, a mixed infiltrate of macrophages (figure 2C) and T-cells (CD3+) was present (figure 2D), but there were

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Figure 1 (A) On presentation, the patient had mild facial erythema with periorbital violaceous patches. (B) Careful skin examination revealed an 8×3 cm thin, pink plaque on the left dorsal forearm, with overlying thin scale and ill-defined borders.

Figure 2 (A) MRI of the upper extremities with contrast revealed myositis of the bilateral shoulder girdles including the deltoid, subscapularis, infraspinatus and supraspinatus, pectoralis major, trapezius, greater than the triceps, brachialis and latissimus dorsi. (B) Biopsy of the left deltoid showed diffuse lymphocytic infiltration with slight perivascular predominance and mild interstitial fibrosis, with groups of atrophic muscle fibres and without any frank necrosis. (C–E) On immunohistochemistry, the mixed infiltrate was composed of macrophages (CD68+) (C), T cells (CD3+) (D) and B cells (CD20+) (E). This immunohistochemical profile does not allow for clear differentiation between polymyositis and dermatomyositis, but the relative numbers of B cells in conjunction with focal atrophic fibres definitely include dermatomyositis as a possibility.

significant numbers of B cells (CD20+) (figure 2E), consistent with either DM or polymyositis. Myositis panel was positive for anti-Mi2 38.6 U, negative for antiextractable nuclear antibody Jo1, antisignal recognition particle, Sjogren's syndrome-A (Ro) and Sjogren's syndrome-B (La) and antinuclear antibodies, suggestive of DM.⁸

As part of a malignancy screen, a CT scan of the chest, abdomen and pelvis was carried out on day 3, which showed no neoplasm or interstitial lung pathology but noted a proximal partial small bowel obstruction (SBO) and diffuse pan-colonic thickening with loss of haustral pattern, suggestive of colitis. Clinically, the patient appeared well, and with conservative management, her SBO initially resolved. By day 10, the patient's weakness improved to the point where she was able to feed herself and lift her arms above her head. For that reason, she was switched to 30 mg PO prednisone.

OUTCOME AND FOLLOW-UP

On day 14, the patient became febrile to 39.2°C. Abdominal examination was unchanged. Repeat abdominal X-ray demonstrated intraperitoneal air. Broad-spectrum Gram negative and anaerobic coverage was started. The patient underwent

emergent laparotomy, which revealed perforation of the transverse colon with multiple sites of mucosal ulceration (figure 3A), and a subtotal colectomy with end ileostomy was performed (with future plans for re-anastomosis). Colonic pathology revealed vasculitis, characterised by prominent lymphoplasmacytic infiltrate around and within the vessel wall (figure 3B), with multifocal mucosal ulceration of varying depth with multiple sites of transmural necrosis leading to perforation and serositis involving the colon (figure 3C). Additionally, submucosal veins demonstrated various degrees of occlusion and arteries showed marked eccentric intimal hyperplasia with focal collections of histiocytes (figure 3D). Such changes are consistent with GI vasculopathy in DM and JDM.^{5 9}

Postoperatively, the patient's methylprednisolone dose was increased from 25 to 80 mg (1 mg/kg). She recovered well, and was ultimately discharged to a rehabilitation facility, with a normal CK of 18 U/L while on 60 mg prednisone and 50 mg azathioprine daily. By 2 months, she had received two additional treatments of 2 mg/kg IVIG, and a prolonged prednisone taper was initiated.

DISCUSSION

In their paper, Tweezer-Zaks and colleagues retrospectively analysed the course of 48 patients with DM or polymyositis, none of whom had any evidence of overlap syndrome.⁵ Three patients had severe lower GI manifestations weeks-months after their myositis diagnosis was made. The predominant findings on upper and lower endoscopy in all three patients were oedematous, hyperaemic bowel wall with multiple erosions and ulcerations, and diffuse mucosal involvement with multiple vascular ectasias but no frank vasculitis. Although all three patients were placed on aggressive immunosuppression, two of the three patients had perforations and significant GI bleeding, and one expired. In another case report, a patient with polymyositis (and no evidence of overlap syndrome) developed nausea and abdominal distension.¹⁰ Despite being on dual

immunosuppression therapy, her symptoms progressively worsened to malabsorption and severe weight loss. Ultimately, this patient had to begin total parenteral nutrition to maintain her weight. Eshragi *et al*¹¹ reported a case of a 24-year-old patient whose GI abscesses ultimately led to duodenal perforation while the patient was on prednisone and methotrexate.

The patient reported here had a change in bowel habits that occurred almost simultaneously with her presentation with proximal muscle weakness. In the reported cases of mild GI manifestation in DM, symptoms usually resolve or improve with immunosuppression. However, symptoms may worsen, as occurred in our patient, despite dual immunosuppressive therapy. The reason for this may be the extent of advanced vasculopathy at the time of treatment. The patient had significant weakness and GI symptoms for 8 weeks, before treatment was initiated. The significant vascular damage revealed on GI biopsy led to chronic narrowing and thrombosis of the small arteries, caused decreased blood flow limiting the efficacy of immunosuppression, and, ultimately, led to ischaemia and perforation. This hypothesis is consistent with the observation that GI complications are usually a long-term complication of JDM, often when the muscle disease is in good control. The biopsies in these paediatric cases^{8 12} often reveals a non-inflammatory vasculopathy, which is likely the down-stream damage caused by earlier inflammation. This model of early vasculitis leading to damage and late ischaemic non-inflammatory vasculopathy is seen in lupus (SLE). Patients with SLE are known to have an increase in cardiovascular disease and strokes as late complications, often when their inflammatory disease is well-controlled.¹³

Although GI complications are classically delayed, this case demonstrates that GI manifestations should be considered as part of the initial presentation of DM. We know that patients with DM can have progression of lung disease when their muscle and skin disease is well-controlled.¹⁴ This case is a reminder that GI manifestations can also progress in patients

Figure 3 (A) On resection, multiple ulcers were noted along the transverse colon. (B) A prominent lymphoplasmacytic infiltrate around and within the vessel wall, with multifocal mucosal ulceration of varying depth. (C) In some sites of ulceration, transmural necrosis was noted resulting in perforation and serositis. (D) Submucosal veins demonstrated various degrees of occlusion and arteries showed marked eccentric intimal hyperplasia with focal collections of histiocytes.

who appear to have stable or improving disease, and that clinicians should be vigilant because of the significant increase in mortality and morbidity associated with GI vasculopathy.

Learning points

- ▶ Although gastrointestinal (GI) complications of dermatomyositis are classically delayed, this case demonstrates that GI vasculopathy should be considered as part of the initial presentation of adult dermatomyositis, especially in patients who are symptomatic.
- ▶ While skeletal muscle myositis responds to conventional immunosuppression, GI disease may be more difficult to control.
- ▶ Pathophysiologically, early inflammation of GI vessels results in chronic ischaemia and thrombosis of arteries, ultimately leading to perforation.

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