Juvenile dermatomyositis (JDMS) is an idiopathic inflammatory myopathy manifested by progressive symmetrical proximal weakness, increased plasma muscle enzymes, an abnormal electromyogram, heliotrope rash, Gottrön’s papules, and polyarthralgia [1]. Gastrointestinal tract involvement in JDMS mainly presents as bowel vasculitis with mucosa thickening and lymphocyte infiltration, a decrease in esophageal motility, and bowel stasis secondary to motility disturbance. Focal vasculitis and ischemia with consequent bowel ulceration and necrosis may also occur in patients with JDMS [2]. The deposition of immune complexes and terminal complement components with release of von Willebrand factor antigen from damaged endothelial and anti-endothelial cell antibodies leads to occlusion of capillaries, infarction, ischemia, and focal vasculitis [3]. Duodenal perforation has been reported in patients with Helicobacter infection or those taking steroids and non-steroidal anti-inflammatory drugs (NSAIDs) [4]. However, its association with JDMS is extremely rare. To our knowledge, only seven such cases have been reported in the literature [5–8]. Making a differential diagnosis in a patient with JDMS presenting with isolated duodenal vasculitis or duodenal ulcer and perforation is difficult. Herein, we report a rare case of JDMS with duodenal perforation and discuss its diagnosis and treatment.

**Case Report**

A 4-year-old boy with JDMS presented with fever, rhinorrhea, cough, sore throat, maculopapular rashes symmetrically distributed over the extensor sides of the joints of all extremities (ie, heliotrope sign and Gottrön’s papules), and muscle weakness. Elevated plasma creatinine kinase 2,360 U/L (normal, < 190 U/L), lactate dehydrogenase 1,344 U/L (normal range, 230–460U/L), erythrocyte sedimentation rate 34 mm at 1 hour and 46 mm at 2 hours, and positive Gower’s sign were also noted. Muscle biopsy revealed positive dystrophin stain and several foci of degeneration in myofibers, with increased mononuclear infiltration over both intramyofascial and perivascular regions, compatible with the findings in polymyositis. Disease activity was controlled by prednisolone (2.5 mg·kg⁻¹·d⁻¹ for 7 days, 2 mg·kg⁻¹·d⁻¹ for 14 days, a cumulative dose of 546 mg), which was gradually replaced by azathioprine and meloxicam. The patient sought traditional Chinese therapy. However, intermittent abdominal pain, bile-stained vomiting, and fever developed 2 months after the onset of the disease. JDMS-associated vasculitis was suspected and steroid pulse therapy with methylprednisolone...
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30 mg·kg\(^{-1}\)·d\(^{-1}\) for 3 days was given. His general condition worsened and the abdominal pain became persistent, even radiating to the back and the right leg. A hemogram revealed leukocytosis, while plain abdominal roentgenogram disclosed a distended bowel loop with a thickened bowel wall. Abdominal computerized tomography showed widespread retroperitoneal abscesses from the level of the hepatic hilum to that of the vesico-rectal pouch (Fig. 1). Emergency laparotomy revealed a retroperitoneal abscess extending from the subhepatic area to the pelvis and a 1 x 0.5-cm duodenal perforation at the junction of the third and fourth portions. Debridement of necrotic tissues with primary repair of the duodenal perforation and multiple peritoneal drainage was performed. Histologic examination of the surgical specimens showed a perforation of the duodenum with ischemic changes in the adjacent mucosa (Fig. 2). No rod-like Helicobacter bacteria were seen in either duodenal or antral biopsied tissues. The pus culture yielded methicillin-resistant Staphylococcus aureus, methicillin-resistant Staphylococcus epidermidis, Candida albicans, and Serratia Spp. Leakage from the anastomosis site occurred 3 days later and a reanastomosis was successful. The patient’s clinical condition improved thereafter and he could tolerate full oral feeding 24 days after the second operation. Antibiotics were used for 6 weeks while steroids were tapered. He was discharged 50 days after hospitalization.

Discussion

Perforated duodenal ulcer is an extremely rare complication of JDMS [5–8]. Administration of glucocorticoid or NSAIDs, Helicobacter pylori infection [4], or intestinal vasculitis could possibly have resulted in this rare complication in our patient. Conn and Blitzer reviewed 42 randomized studies and suggested that steroids increased the risk of duodenal ulcer only if the patients took the medication for more than 1 month or in a cumulative dosage exceeding the equivalent of 1000 mg of prednisolone [9]. Our patient took prednisolone with a cumulative dose of 546 mg, which did not fulfill the criteria of Conn and Blitzer [9]. Steroid-, NSAID-, or H. pylori-induced ulcers are often located at the antrum or proximal duodenum [10]. In contrast, duodenal vasculitis with ulcer and perforation in patients with JDMS is usually located in the posterior distal descending portion of the duodenum and the ulcer is larger [6, 8]. This was seen in our patient. The duodenal perforation could be attributed to JDMS-related intestinal vasculitis.

Early diagnosis is the only strategy to prevent extensive retroperitoneal abscess, sepsis, and death in a patient with JDMS and duodenal ulcer and perforation. However, it is difficult to differentiate intestinal vasculitis from a complicated duodenal perforation. The pain associated with JDMS-related intestinal vasculitis is intermittent because of vascular spams and intestinal cramping. Its severity is frequently relevant to the underlying disease activity characterized by elevated muscle enzyme levels and degree of muscle weakness [11]. By contrast, the pain associated with duodenal perforation is persistent due to peritoneal irritation, may radiate to the back, and is not relieved by body position changes [12]. If the abdominal pain does not improve after steroid pulse therapy, duodenal perforation should be highly suspected.

Once the diagnosis of duodenal perforation is made, emergency surgery is recommended. To treat JDMS-associated duodenal perforation, segmental resection with end-to-end anastomosis may be a safe procedure since the extent of vasculitis in the surrounding tissues is difficult to assess [6, 8]. This particular patient had the complication of anastomotic leakage, which could be due to extensive vascular inflammation.

In summary, duodenal ulcer with perforation should be considered in a JDMS patient with abdominal pain that results from lesions other than gastroduodenitis or vasculitis, especially when the abdominal pain is persistent. Early detection and immediate surgical repair of a duodenal perforation should provide a better result.
References


