INTESTINAL VASCULITIS, MESENTERITIS, MICROANGIOPATHIC ANEMIA AND ALVEOLAR HEMORRHAGE: A SEVERE PRESENTATION OF DERMATOMYOSITIS

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BACKGROUND

Intestinal vasculitis is an extramuscular manifestation of inflammatory myositis, reported mainly on young-age patients with juvenile dermatomyositis (JDM). We hereby report a case of an adolescent with severe dermatomyositis (DM) evolving to small intestinal perforation.

CASE REPORT

A 19-year-old male patient was diagnosed with dermatomyositis presenting with heliotrope rash, Gottron sign, proximal muscle weakness and severe dysphagia, requiring nasogastric tube. There was elevated inflammatory markers, high skeletal-muscle enzymes and positive ANA 1/80 with nuclear fine speckled pattern. Anti-Jo-1, anti-Mi-2 and anti-SRP were negative. The disease was refractory to 3,000 mg of methylprednisolone (MP) and 1,000 mg of cyclophosphamide. A progressive improvement was achieved after 2,000 mg/kg of intravenous immunoglobulin (IVIg). Methotrexate was initiated as maintenance therapy with slow tapering of glucocorticoid. In outpatient follow-up appointments, he had progressive improvement, being able to attend the gym after a few months. Six months after discharge, the patient presented with fever, microangiopathic anemia and acute abdomen preceded by 2 months of epigastric pain and weight loss. Abdominal computed tomography (CT) revealed mesenteritis and small intestine distention. Thorax CT showed a small area of alveolar hemorrhage, with no clinical repercussion (Figure 2b). Due to his poor nutritional status, the surgery consultant chose a conservative approach and he received 200 mg of MP, IVIg and empirical antibiotics even though cultures were negative. Microangiopathic anemia and lung alterations responded to immunosuppression, nonetheless the patient evolved with pneumoretroperitoneum and hematemesis. Laparoscopy revealed a large retroperitoneal duodenum perforation. Anatomopathological of intestinal mucosa revealed hemorrhage and necrosis. Immunohistochemical analysis of a reactive lymph node discarded the hypothesis of associated lymphoma. Immunosuppression was dose-limited due to nosocomial infections and a long-term antibiotic was maintained. A few weeks later hematemesis recurred. A new laparoscopy evidenced an increase in size of previous lesions besides a new jejunal perforation. A 13-cm jejunum resection and an omentopexy with bovine pericardium on duodenum were performed. The patient was treated with 100 mg of MP, monthly 2,000 mg/kg of IVIg, long-term antibiotics and exclusive parenteral nutrition. He had no further surgical complications after a 3-month follow-up period and he is gaining weight aiming at a reconstruction surgery in the future.

CONCLUSION

To our knowledge, this is the first case of DM presenting microangiopathic anemia, alveolar hemorrhage, mesenteritis and mesenteric vasculitis. Small bowel perforation is associated with high morbimortality, rising awareness for its early recognition.

KEYWORDS

Inflammatory myopathy, Dermatomyositis, Refractory myopathy, Duodenal vasculitis, Mesenteric vasculitis.