

Image of the Month

Ischemic Ulcerative Colitis in Juvenile Dermatomyositis



FIG. 1. Painful cutaneous vasculitis ulcer over the right elbow.



FIG. 2. Livedo reticularis of the face with heliotrope rash.



FIG. 3. Increased vascular pattern in descending colon with dilated and tortuous blood vessels reflecting the vasculopathy associated with juvenile dermatomyositis.



FIG. 4. Mucosal ulceration (Ø 0.4cm) with a white base in the descending colon with hyperemic, raised edge.

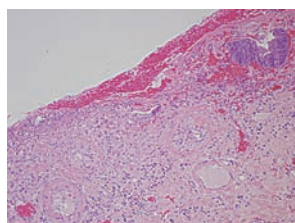


FIG. 5. Hematoxylin and eosin stain; original magnification $\times 100$. Colonic biopsy coinciding with an area of ulceration, lined with fibrin and granulation tissue, and containing a mixed inflammatory infiltrate, mainly composed of granulocytes and scattered lymphocytes. The submucosa appears diffusely edematous and shows a small arteriole with subendothelial edema and swollen endothelia. At the periphery, the mucosa shows hyperplastic-reactive changes. (Image courtesy of Rita Alaggio, MD; Department of Pathology, University of Padua, Italy)

A 6-year-old girl with juvenile dermatomyositis (JDM), diagnosed according to the Bohan and Peter criteria (1), presented with skin ulcerations (Fig. 1) and livedo reticularis (Fig. 2). Three months after diagnosis she developed severe iron-deficiency anemia, abdominal pain, evidence of occult blood in the stool, and occasional hematochezia. Colonoscopy showed a serpiginous vascular pattern and 2 ulcers (Figs. 3 and 4). Histological findings on mucosal biopsies showed granulation tissue and mixed inflammatory infiltrate, mostly composed of granulocytes and scattered lymphocytes, with peripheral mucosal hyperplastic-reactive changes (Fig. 5).

Among the possible complications of JDM (2–9), gastrointestinal (GI) involvement is rare (3) and ulcers are more typical of the juvenile- than of the adult-onset form (2). GI vasculopathy can manifest as ulceration, haemorrhage, pneumatosis intestinalis, or perforation (4,10). These symptoms and signs may arise early in the course of the disease as a result of a noninflammatory endarteropathy or late because of chronic vasculopathy (2,10).

Vasculitides involving the GI tract are part of a systemic process, although the signs and symptoms may initially be vague (9). GI involvement should be considered in every patient with JDM associated with skin ulcerations and abdominal symptoms, to ensure appropriate immunosuppressive treatment and avoid severe complications.

Submitted by:

*Alessandra Meneghel, MD, †Francesco Zulian, MD, †Giorgia Martini, MD, and *Graziella Guariso, MD

*Department of Pediatrics, Gastroenterology-Endoscopy, and †Rheumatology Units, University of Padua, Padua, Italy

Address correspondence and reprint requests to Prof. Graziella Guariso, Dipartimento di Pediatria, Via Giustiniani 3, 35128 Padua, Italy (e-mail: guariso@pediatria.unipd.it).

The authors report no conflicts of interest.

REFERENCES

1. Bohan A, Peter JB. Polymyositis and dermatomyositis (first of two parts). *N Engl J Med* 1975;292:344–7.
2. Mamyrova G, Kleiner DE, James-Newton L, et al. Late-onset gastrointestinal pain in juvenile dermatomyositis as a manifestation of ischemic ulceration from chronic endarteropathy. *Arthritis Rheum* 2007;57:881–4.
3. Cassidy JT, Lindsley CB. Juvenile dermatomyositis. In: Cassidy, Petty, Laxer and Lindsley, eds. *Textbook of Pediatric Rheumatology*. Philadelphia: Elsevier-Saunders; 2005. pp. 407–41.
4. Wang JJ, Hsu WM, Shun CT, et al. Juvenile dermatomyositis complicated with vasculitis and duodenal perforation. *J Formos Med Assoc* 2001;100:844–6.
5. Pahalad S, Bohnsack JF, Maloney CG, et al. Fatal acute fibrinous and organizing pneumonia in a child with juvenile dermatomyositis. *J Pediatr* 2005;146:289–92.
6. Rider LG, Atkinson JC. Images in clinical medicine. Gingival and periungual vasculopathy of juvenile dermatomyositis. *N Engl J Med* 2009;360:e21.
7. Yun SJ, Lee JB, Kim SJ, et al. Calcinosis cutis universalis with joint contractures complicating juvenile dermatomyositis. *Dermatology* 2006;212:401–3.
8. Jat KR, Singh S. Calcinosis in juvenile dermatomyositis. *Indian Pediatr* 2008;45:784.
9. Cucchiara S, Borrelli O. Vasculitides. In: Guandalini S, ed. *Textbook of Pediatric Gastroenterology and Nutrition*. London: Taylor & Francis Group; 2004. pp. 419–34.
10. Feldman BM, Rider LG, Reed AM, et al. Juvenile dermatomyositis and other idiopathic inflammatory myopathies of childhood. *Lancet* 2008;371:2201–12.