Generalized Pyoderma Gangrenosum Associated with Unrecognized Ulcerative Colitis

Dear Editor,

We present a case of a 36-year-old female patient who was initially admitted to the Department of Infectious Diseases for evaluation of diarrhea. Two years prior to admission, she had been treated in another hospital for erythema nodosum on her calves. There was no further diagnosis at that time. The patient presented with frequent loose stool for three weeks before admission to our Department, at the time without any bloody tinge present. Four days before admittance, the patient's temperature rose to 39°C, with bloody loose stools and a C-reactive protein (CRP) level of 42.8 mg/L. On the fourth day of hospitalization, laboratory tests showed anemia and erythrocyte sedimentation rate (ESR) of 62 mm/3.6 ks. The next day, the patient developed generalized pyogenous pustules with necrotizing borders on her skin. Serologic testing was negative for VDRL, HIV, ANA, HBsAg, and HCV antibodies, as well as and RA factor. Skin ulcer smear was sterile, and chest X-ray and abdominal ultrasound did not reveal any abnormality. Pathohistological analysis of the skin biopsy specimen was compatible with pyoderma gangrenosum (PG). The dermatologist established a diagnosis of severe generalized PG. Stool samples tested negative for infectious agents. Laboratory tests showed progressed anemia, higher leukocyte levels, ESR of 86 mm/3.6 ks, and CRP level of 151 mg/L. On the fourteenth day of hospitalization, a colonoscopy was performed and showed that the mucosa of the descending colon and rectum was completely covered with ulcerations and pseudopolypous lesions (Figure 1). The patient was transferred to our Department. Pathohistological specimens taken during colonoscopy were confirmed to be positive for ulcerative colitis (UC). Within a few days of the commencement of appropriate treatment, the patient's general condition improved considerably, stool frequency diminished, her laboratory parameters (ESR, CRP) regressed, and her skin lesions showed significant signs of healing (Figure 2).



Figure 1. Colonoscopy image of the patient, performed on day 14.

PG is a sterile inflammatory neutrophilic dermatosis characterized by recurrent cutaneous ulcerations with mucopurulent or hemorrhagic exudate (1). Esti-



Figure 2. Generalized skin lesions ten days after beginning of ulcerative collitis treatment.

mated incidence of PG is 0.63/100000/year (2). About 50% of PG cases are associated with an underlying disorder (1). Inflammatory bowel diseases (IBDs) are found in 20% of patients (2). UC is a chronic IBD, causing uninterrupted mucosal inflammation of the large intestine without granulomas on biopsy (3). UC has an incidence of 1-20/100000/year and a prevalence of 6-246/100 000 (4,5), but less than 3% of patients with UC develop PG (2). Diagnosis of PG relies primarily on clinical signs, and is supported by histopathological biopsy (1). It is most often diagnosed by exclusion of other skin disorders (6). After the diagnosis of PG is established, it is must be assumed that an underlying disease is present, since this is the case in 50% of the patients. Feliciani et al. (7) emphasize the importance of connecting physical findings with the patient's history, and Suárez-Pérez et al. (8) report that patients with PG are often referred to dermatologists by other specialists after a varying period of time has elapsed without achieving an accurate diagnosis. In our case, the patient had been treated for erythema nodosum two years prior to this episode of generalized PG. She was not followed-up after resolution of the condition and, since she had only mild abdominal symptoms, she had not been further investigated. If our patient had been diagnosed with UC after her first skin manifestations two years ago, it is possible that the severe PG and UC that she presented with could have been prevented. Therefore, in patients with skin disorders such as PG, routine colonoscopy should be considered in order to discover an underlying disease such as IBDs, so that adequate treatment can be administered on time, and possible severe extraintestinal manifestations can be prevented.

References

- 1. Wollina U. Pyoderma gangrenosum a review. Orphanet J Rare Dis 2007;15:2-19.
- Langan SM, Groves RW, Card TR, Gulliford MC. Incidence, mortality, and disease associations of pyoderma gangrenosum in the United Kingdom: a retrospective cohort study. J Invest Dermatol 2012;132:2166-70.
- Dignass A, Eliakim R, Magro F, Maaser C, Chowers Y, Geboes K, et al. Second European evidence-based Consensus on the diagnosis and management of ulcerative colitis Part 1: Definitions and diagnosis. J Crohns Colitis 2012; 6:965-90.
- Loftus EV Jr. Clinical epidemiology of inflammatory bowel disease: incidence, prevalence and environmental influences. Gastroenterology 2004;126:1504-17.
- 5. Mijandrušić Sinčić B, Vucelić B, Peršić M, Brnčić

N, Jurišić Eržen D, Radaković B, et al. Incidence of inflammatory bowel disease in Primorsko-goranska county, Croatia, 2000-2004: a prospective population-based study. Scand J Gastroenterol 2006;41:437-44.

- Santos M, Talhari C, Rabelo R, Schettini APM, Chirano C, Talhari S. Pyoderma gangrenosum – a clinical manifestation of difficult diagnosis. An Bras Dermatol 2011;86:153-6.
- 7. Feliciani C, De Simone C, Amerio P. Dermatological signs during inflammatory bowel diseases. Eur Rev Med Pharmacol Sci 2003;13 Suppl 1:15-21.
- Suárez-Pérez JA, Herrera-Acosta E, López-Navarro N, Vilchez-Márquez F, Prieto JD, Bosch RJ, et al. Pyoderma gangrenosum: a report of 15 cases and review of the literature. Actas Dermosifiliogr 2012;103:120-6.

Vanja Giljača¹, Davorka Lulić¹, Milan Ličina¹, Davor Štimac¹, Leo Čabrijan², Brankica Mijandrušić Sinčić¹

¹Department of Internal Medicine, Division of Gastroenerology, ²Department of Dermatovenereology, Clinical Hospital Center Rijeka, Rijeka, Croatia

Corresponding author:

Vanja Giljača, MD Department of Internal Medicine, Division of Gastroenterology Clinical Hospital Center Rijeka Krešimirova 42, 51000 Rijeka Croatia *vanja.giljaca@gmail.com*

> Received: March 19, 2013 Accepted: October 20, 2013