

Crohn's disease-like colonic lesions in chronic granulomatous disease

Chronic granulomatous disease (CGD) is an X-linked recessive immunodeficiency affecting phagocytes/neutrophils, with defective NADPH oxidase activity. Neutrophils are unable to enhance neutrophil extracellular traps (NETs) for entangling pathogens. The patients are quite susceptible to catalase-producing bacteria such as *Enterobacteriae*, *Staphylococcus*, *Burkholderia cepacia*, *Serratia marcescens*, *Nocardia* and *Aspergillus*. Catalase-negative bacteria such as *Streptococcus*, *Enterococcus* and *Lactobacillus* can be inactivated and killed. Gastrointestinal involvement is quite common in CGD, and mimics inflammatory bowel disease, particularly Crohn's disease. Crohn-like longitudinal ulcers can be observed. The colon is the most commonly site of involvement. Microscopic features include non-caseous epithelioid granulomas, pigmented macrophages, tissue eosinophilia and chronic inflammation infiltrates. Neutrophilic reaction is minimal. Infectious etiology is scarcely proven in the lesion.

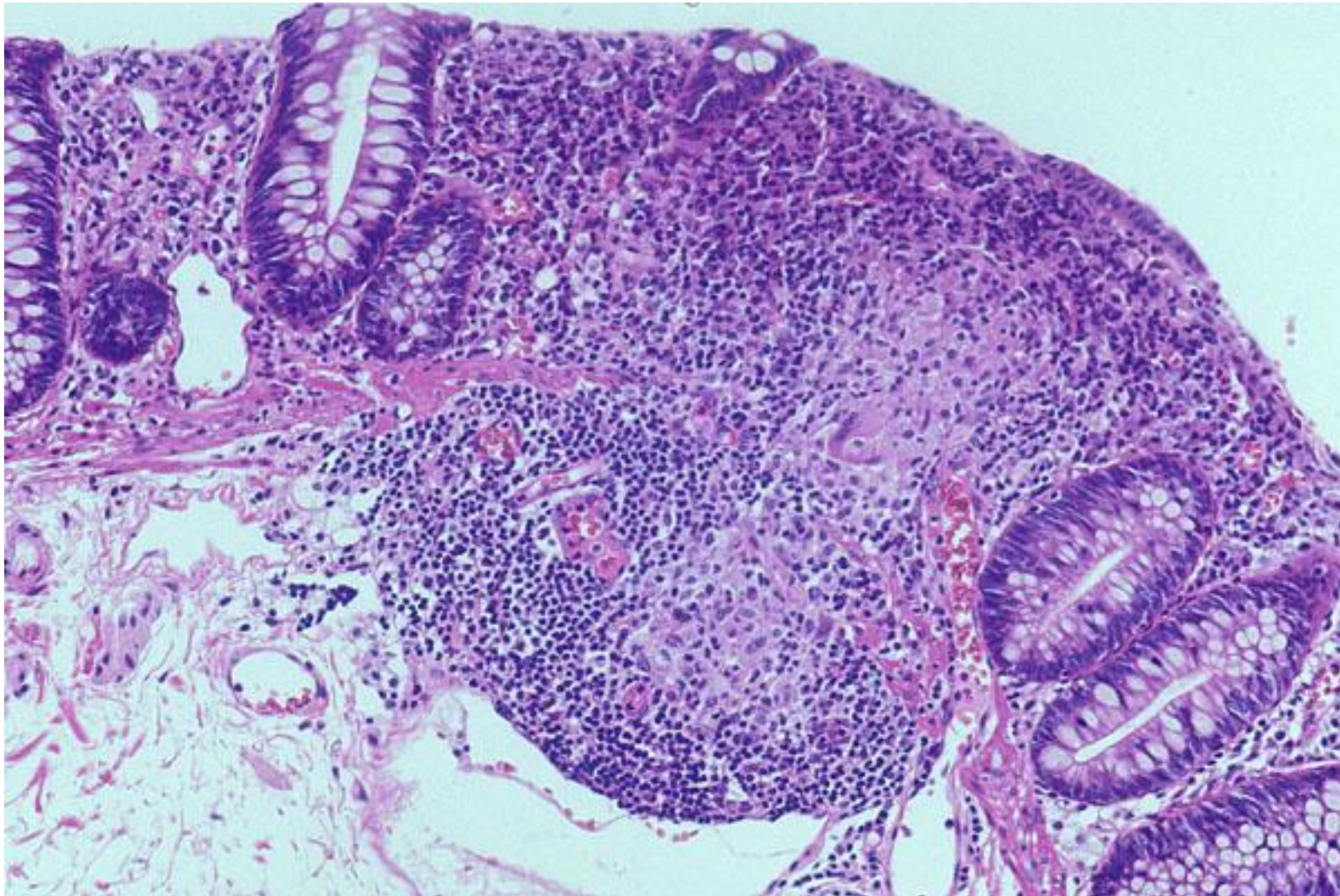
Ref.-1: Tsutsumi Y. Crohn-like longitudinal colonic ulcer in chronic granulomatous disease. 2003, <https://pathos223.com/en/case/case035.htm>

Ref.-2: Alimchandani M, et al. Gastrointestinal histopathology in chronic granulomatous disease: a study of 87 patients. Am J Surg Pathol 2013; 37(9): 1365-1372. doi: 10.1097/PAS.0b013e318297427d

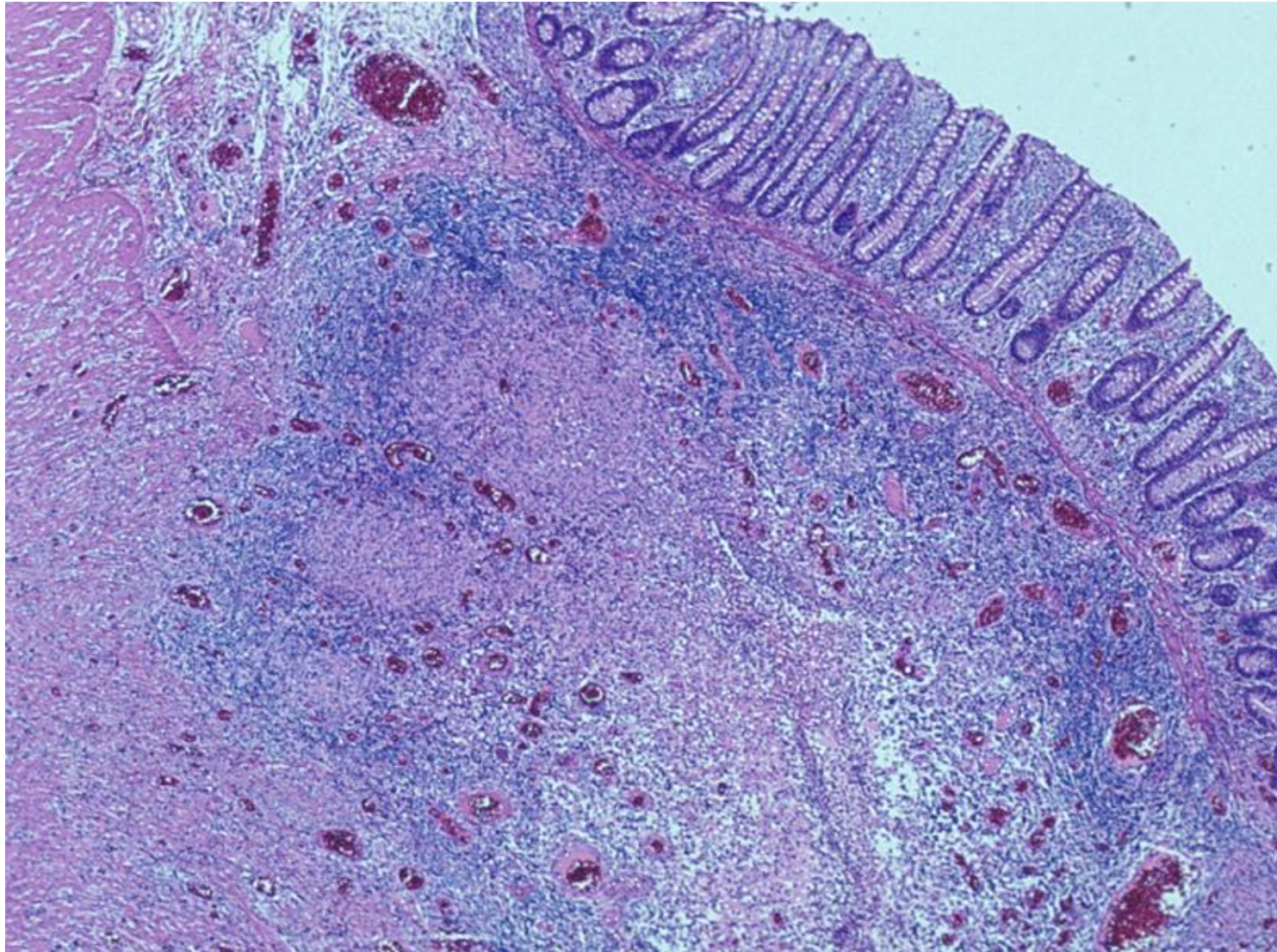
Ref.-3: Zerbe CS, Holland SM. Functional neutrophil disorders: chronic granulomatous disease and beyond. Immunol Review 2024; 322(1): 71-80. doi: 10.1111/imr.13308



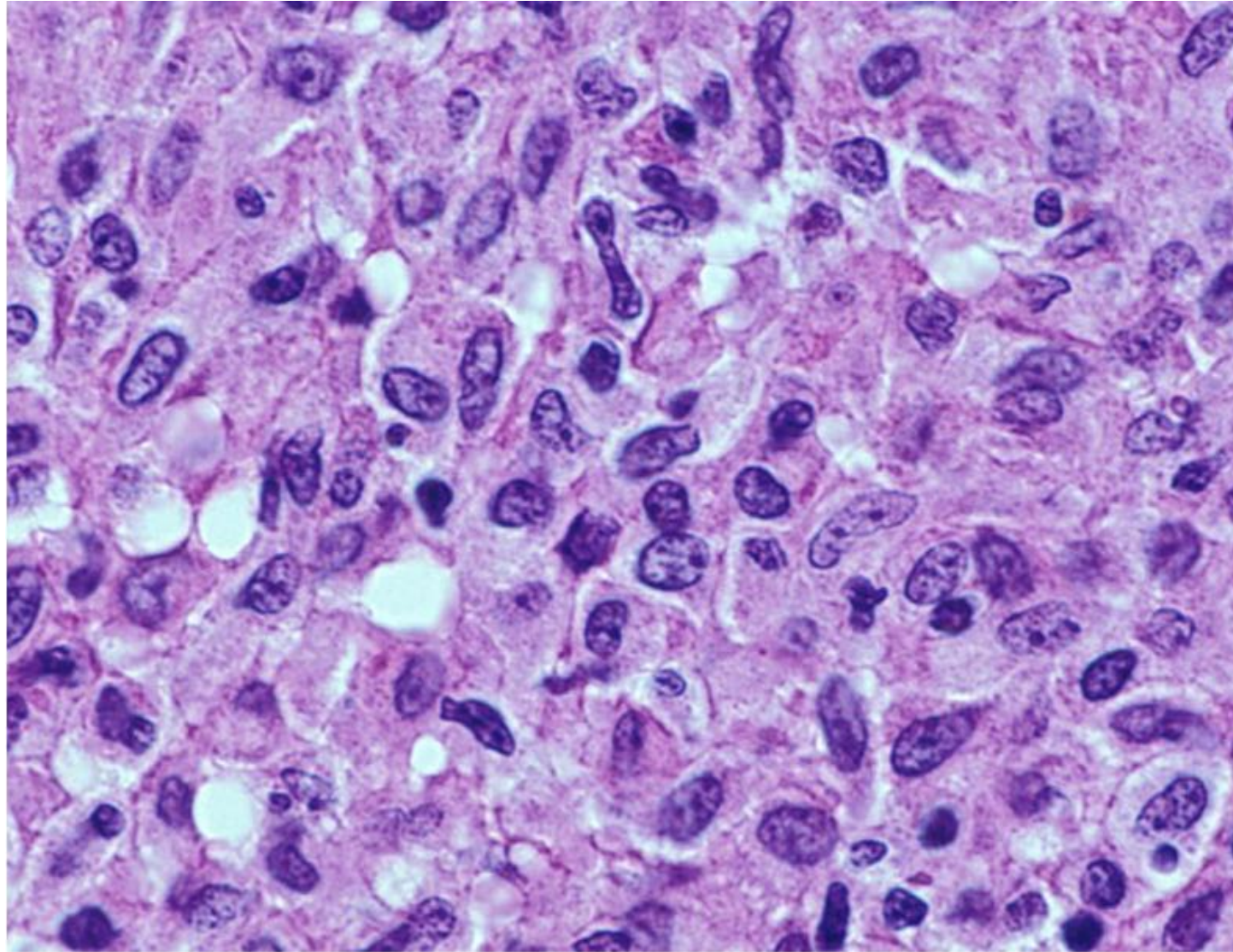
Crohn-like longitudinal colonic ulcer in chronic granulomatous disease (CGD) in an 8-year-old boy). Colon surgically resected for intractable ulceration associated with CGD. CGD is characterized by hereditary neutrophilic dysfunction. The defective neutrophils cannot digest catalase-producing microbes, such as *Staphylococcus*, *E. coli*, tuberculous bacilli and fungi.



The colon lesion in chronic granulomatous disease in an 8-year-old boy. Non-caseous granulomas are formed in the lamina propria mucosae through the submucosa of the colon. H&E-1



The colon lesion in chronic granulomatous disease in an 8-year-old boy. Non-caseous granulomas are formed in the lamina propria mucosae through the submucosa of the colon. H&E-2



The colon lesion in chronic granulomatous disease in an 8-year-old boy. High-powered view of non-caseous granuloma. Immunostaining for *E. coli* antigen is negative in the present case. H&E-3