

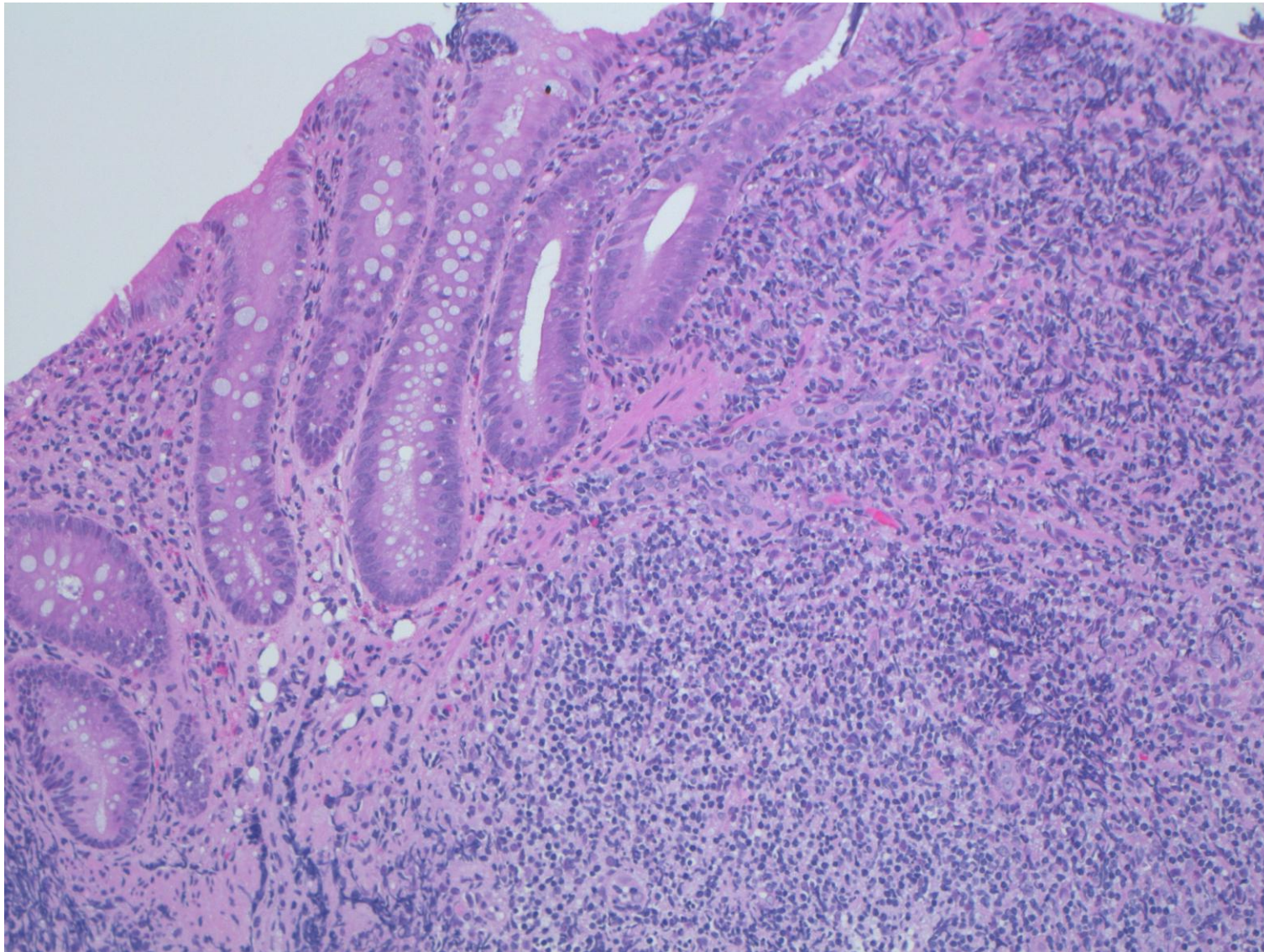
Chronic colitis in common variable immune deficiency (CVID)

Common variable immune deficiency (CVID) is a heterogeneous primary immune deficiency disorder of the adaptive immune system, characterized by a loss of B cell function with a significantly low serum IgG associated with low IgM and/or IgA, and also with poor or absent serological response to vaccines. The loss of plasma cells and the impaired functions of T-lymphocytes are characteristic. The disease is predominantly (90%) sporadic without heredity. Most patients are diagnosed between the ages of 20 and 40, while the diagnosis of CVID is made in children in about 20% of cases. CVID shows an increased susceptibility to bacterial infections, particularly in the sino-pulmonary and digestive tissues. In the GI tract, infection of *Giardia lamblia*, *Campylobacter jejuni*, *Salmonella* species, *Clostridium difficile* or CMV can be complicated. In addition, the risk of non-infectious enterocolitis is also high in CVID cases.

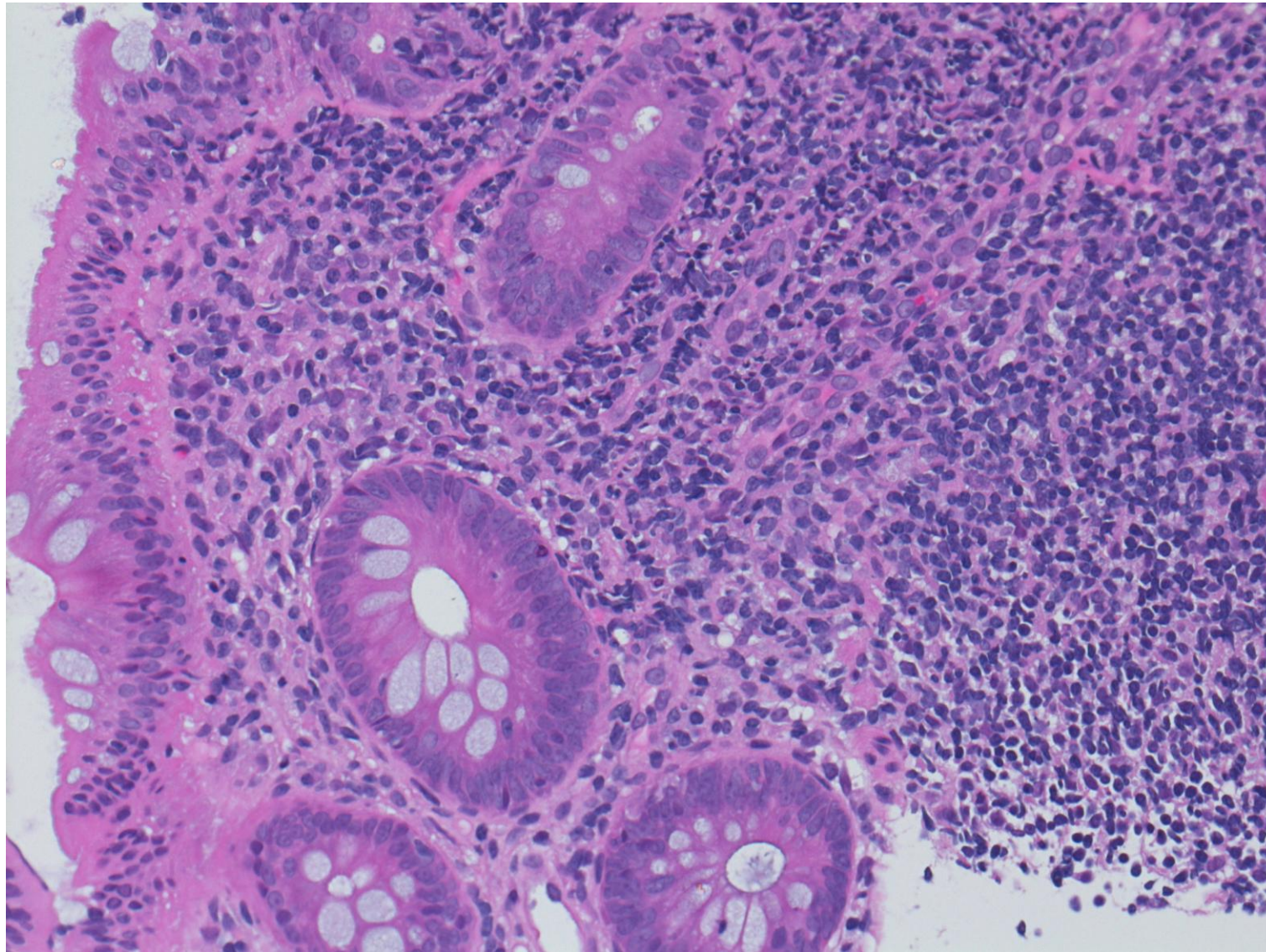
Non-infectious colonic lesions in CVID reveal mild chronic inflammation of the colon with an increase of intraepithelial lymphocytes (IELs), neutrophilic infiltration, follicular lymphoid hyperplasia and the absence of plasma cells. Increased IELs may resemble microscopic features of celiac sprue. The colitis in CVID may also mimic microscopic colitis (mostly lymphocytic colitis and occasionally collagenous colitis), or graft versus host disease. In severe cases with ulceration, the microscopic pattern can be similar to that of ulcerative colitis with crypt abscesses and crypt destruction. Formation of non-caseating granulomas may resemble Crohn's disease.

Ref.-1: Malamut G, et al. The enteropathy associated with common variable immunodeficiency: the delineated frontiers with celiac disease. Am J Gastroenterol 2010; 105(10): 2262-2275. doi: 10.1038/ajg.2010.214

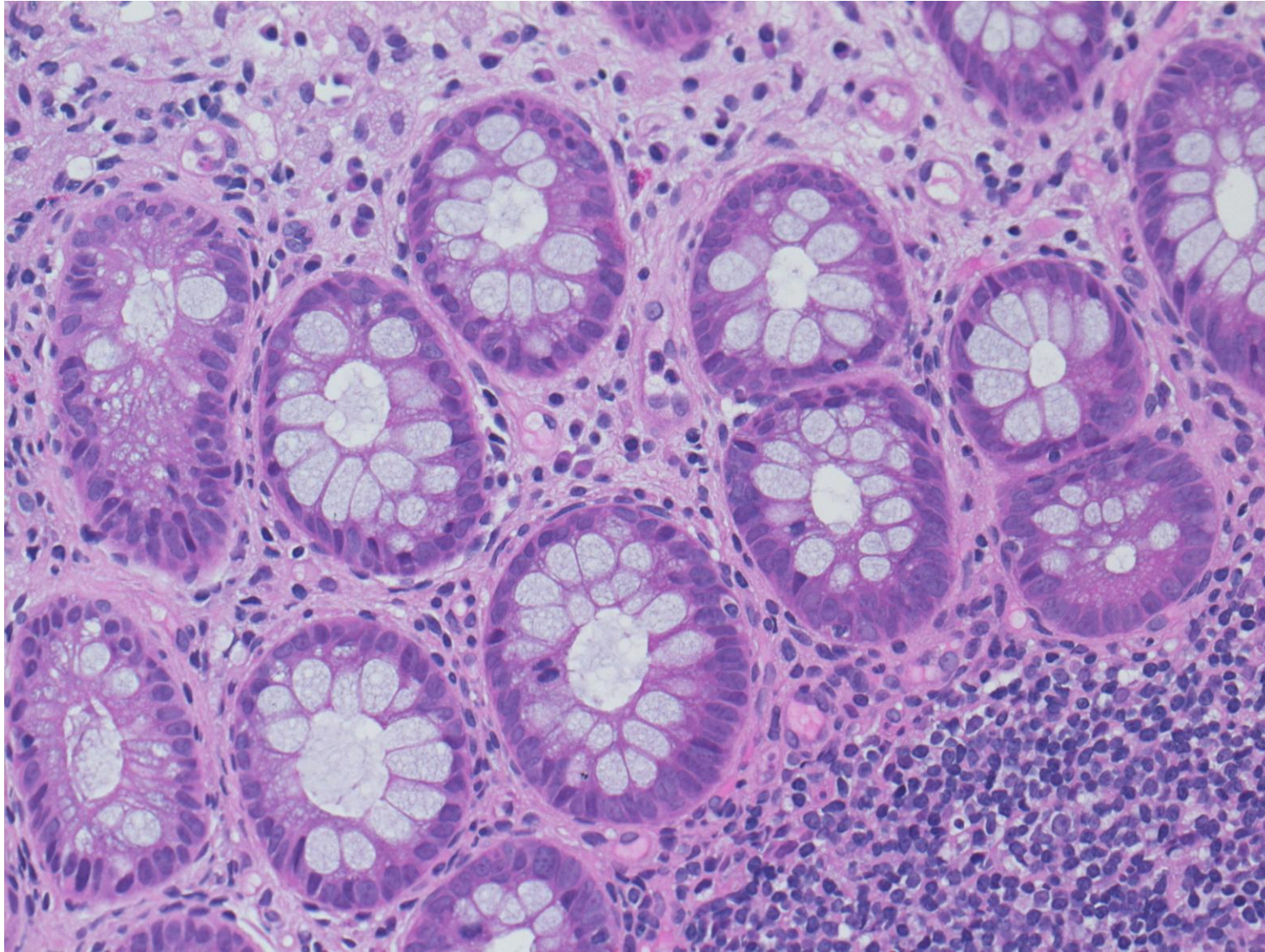
Ref.-2: Uzzan M, et al. Gastrointestinal disorders associated with common variable immune deficiency (CVID) and chronic granulomatous disease (CGD). Curr Gastroenterol Rep 2016; 18(4): 17. doi: 10.1007/s11894-016-0491-3



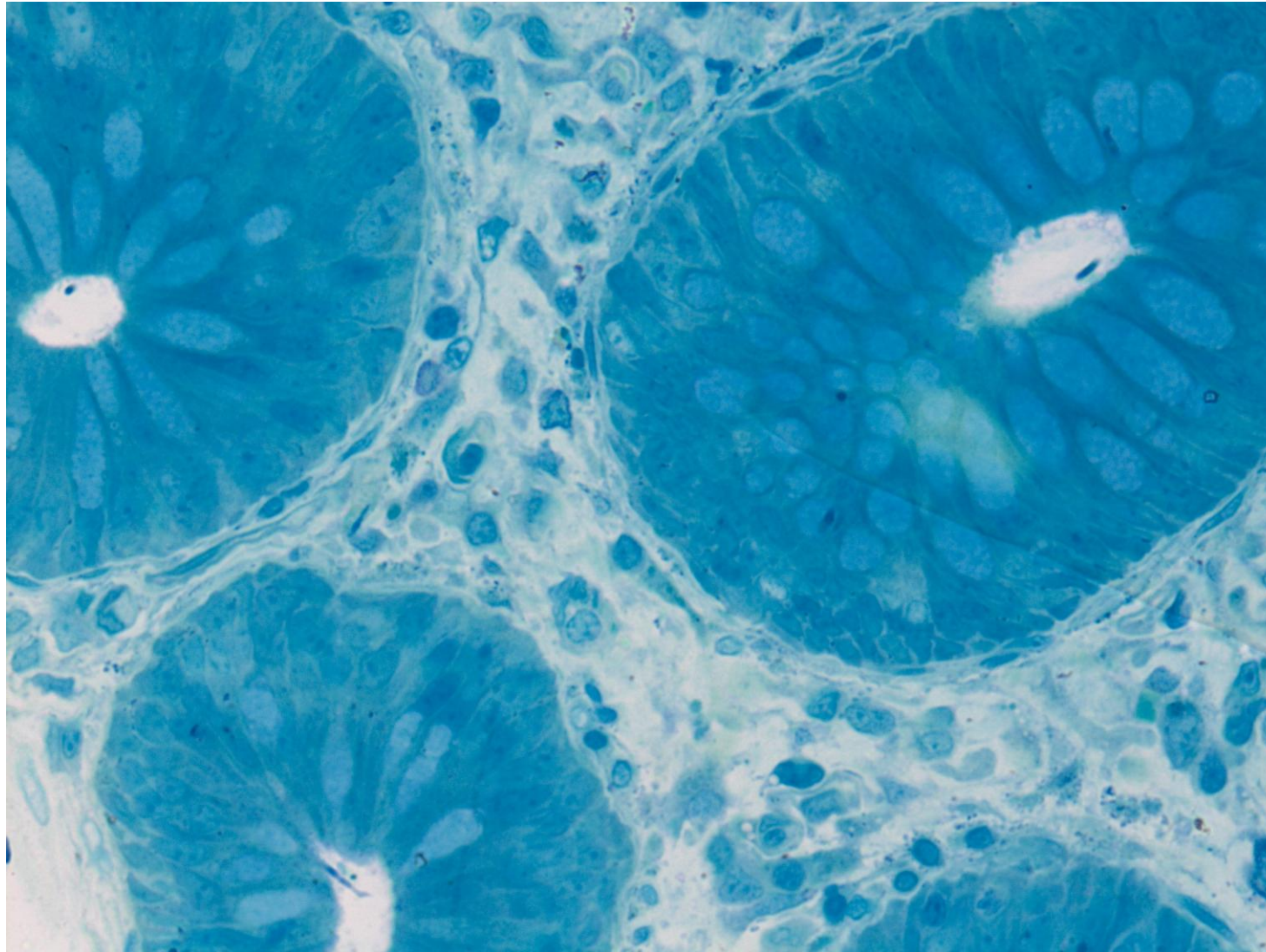
Non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. The colonic mucosa is moderately inflamed with lymphoid follicle formation. Intraepithelial lymphocytes are scattered. H&E-1



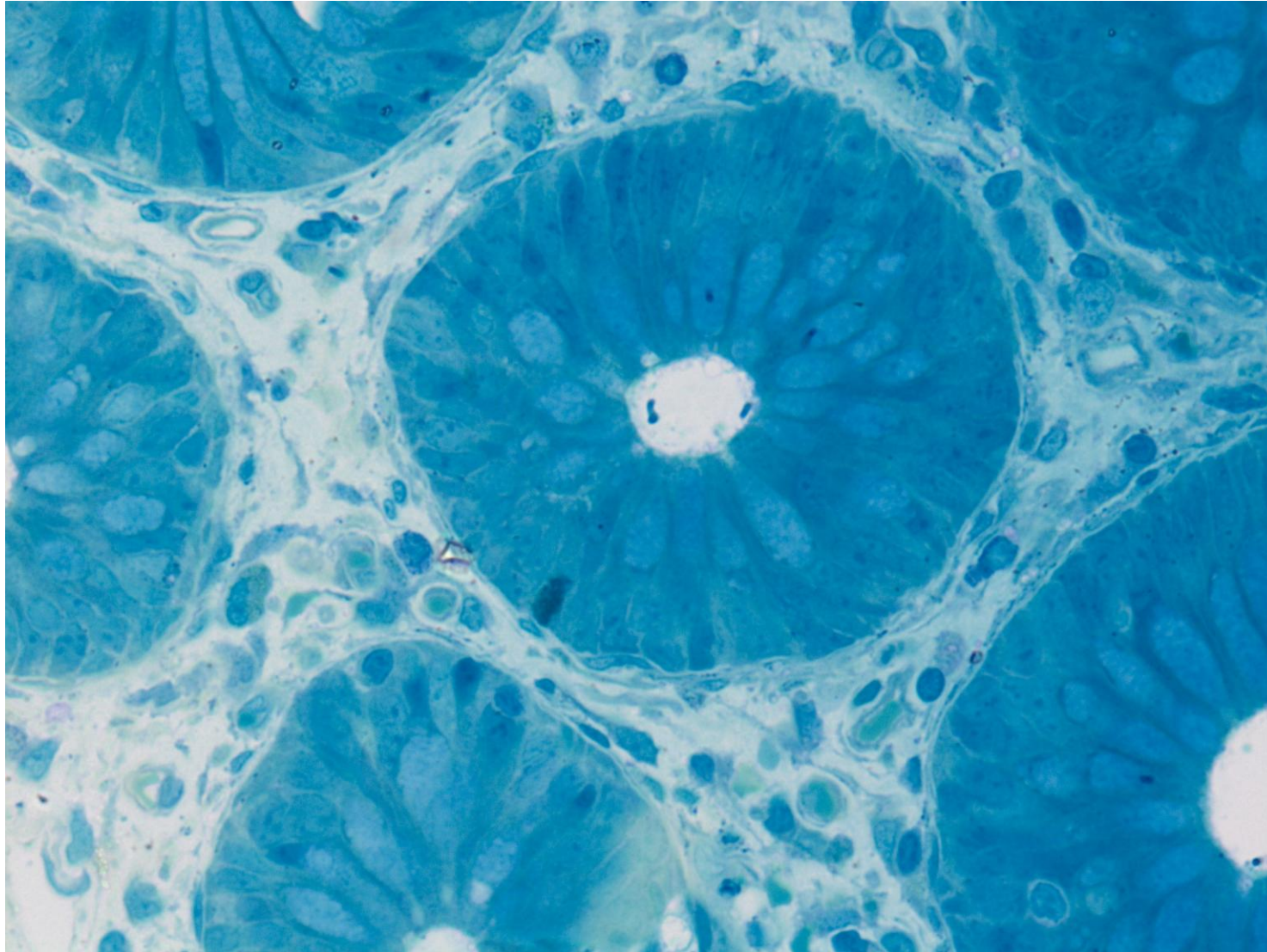
Non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. The colonic mucosa is moderately inflamed with lymphoid follicle formation. Only a small number of plasma cells are seen in the lamina propria mucosae. Intraepithelial lymphocytes are scattered. H&E-2



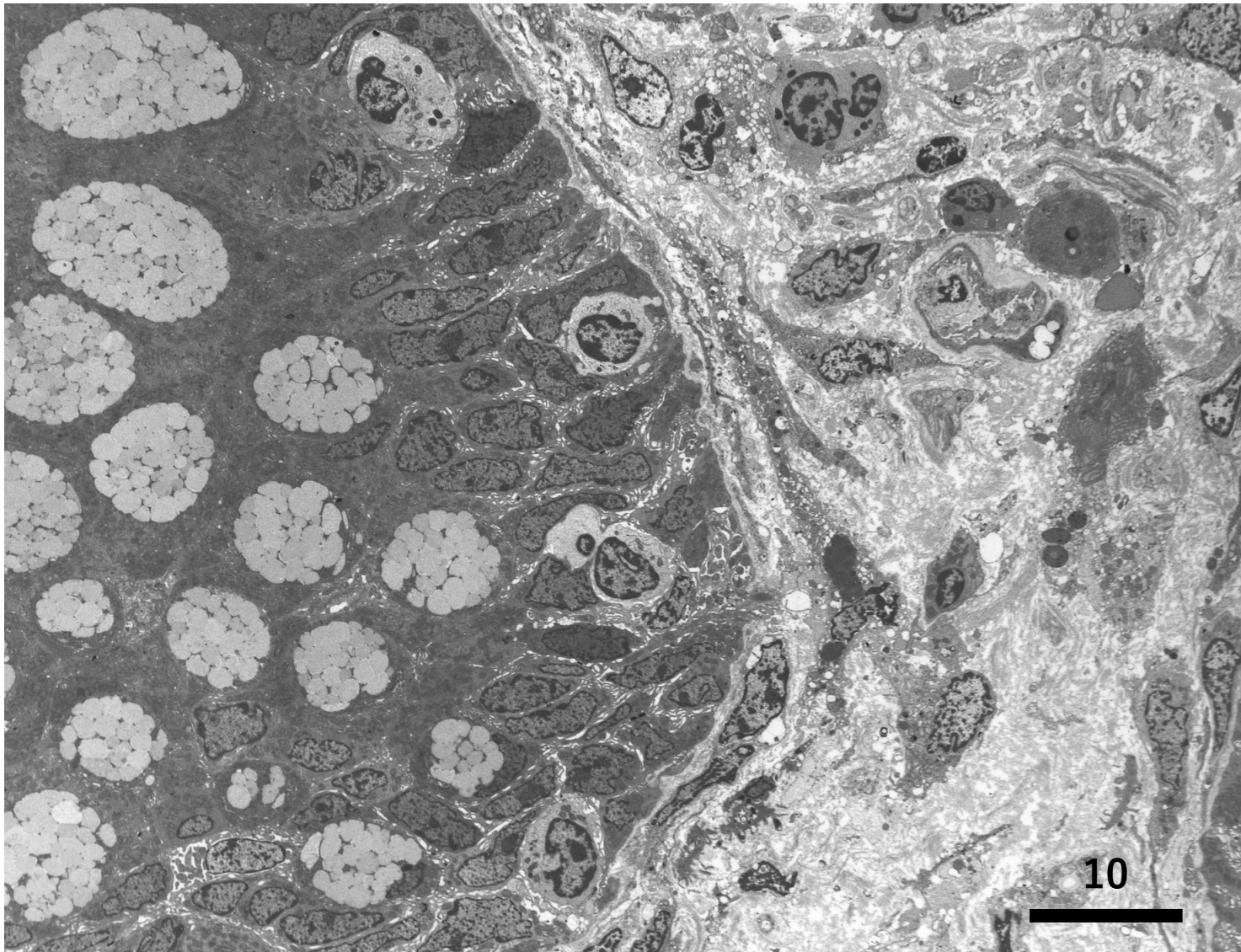
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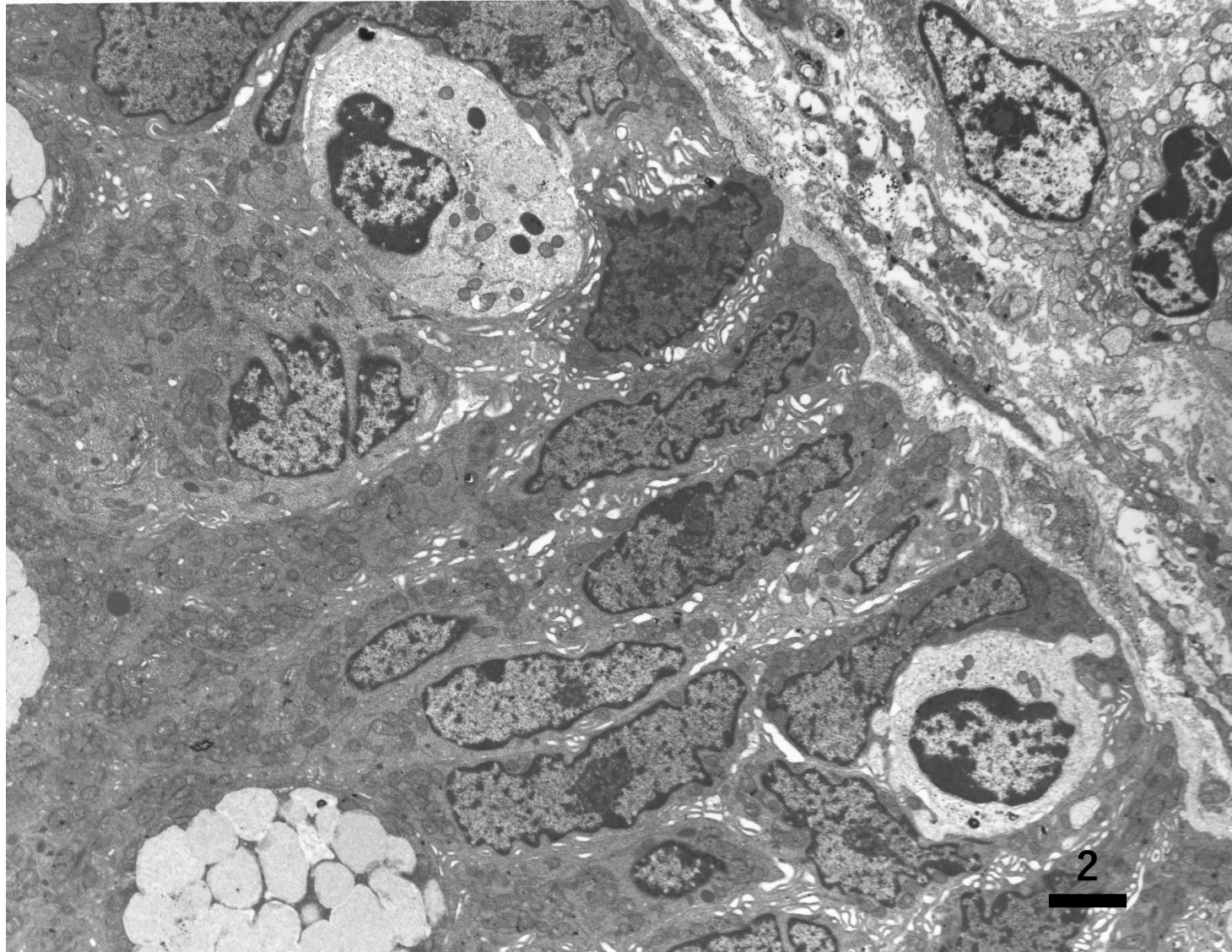
Non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Epon-embedded thick section reveals only a small number of plasma cells in the lamina propria mucosae. Toluidine blue-1



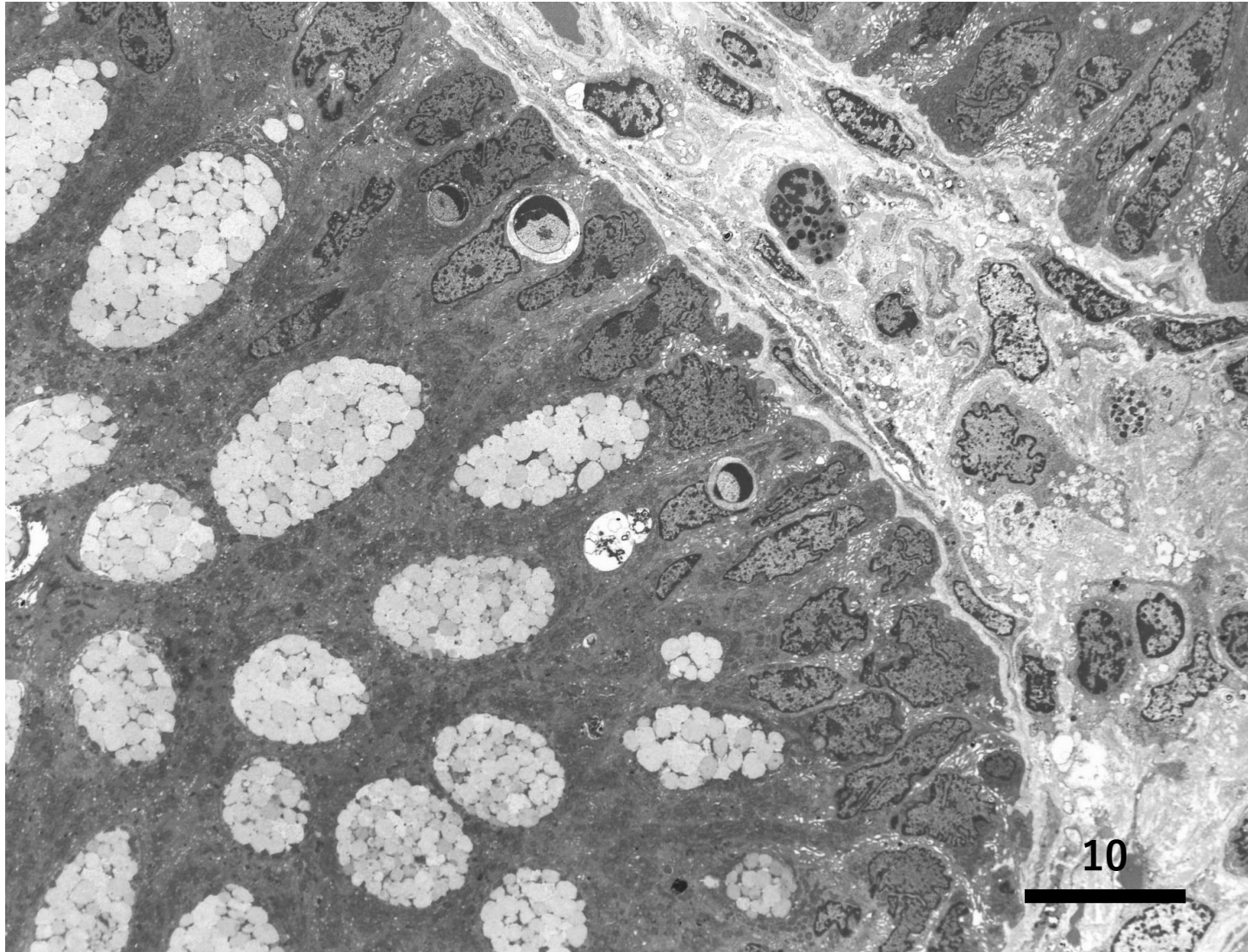
Non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Epon-embedded thick section reveals only a small number of plasma cells in the lamina propria mucosae. Toluidine blue-2



Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Plasma cells are not seen in the lamina propria mucosae. Note the increase of intraepithelial lymphocytes among the columnar epithelial cells. EM-1



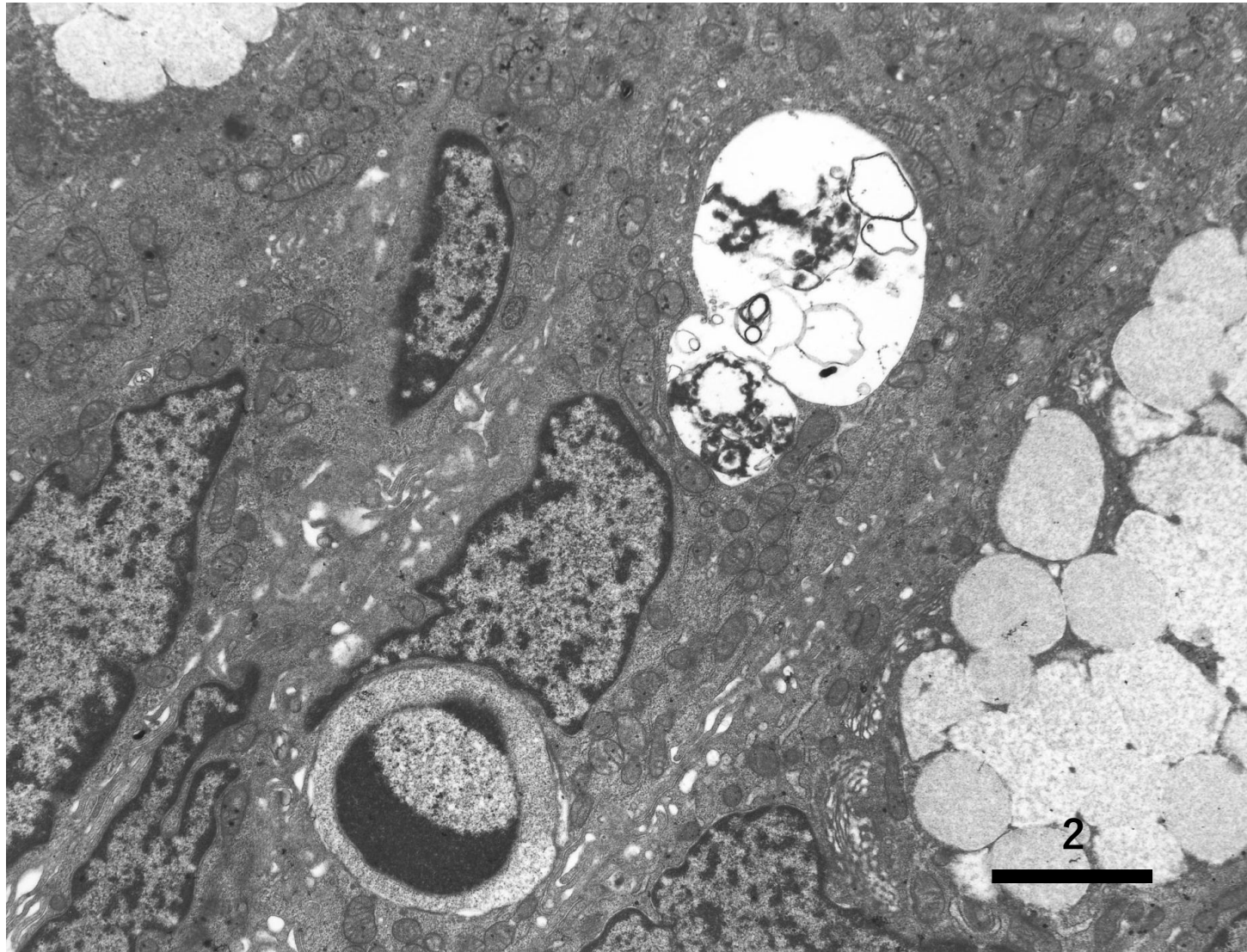
Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Note the increase of intraepithelial lymphocytes among the columnar epithelial cells. EM-2



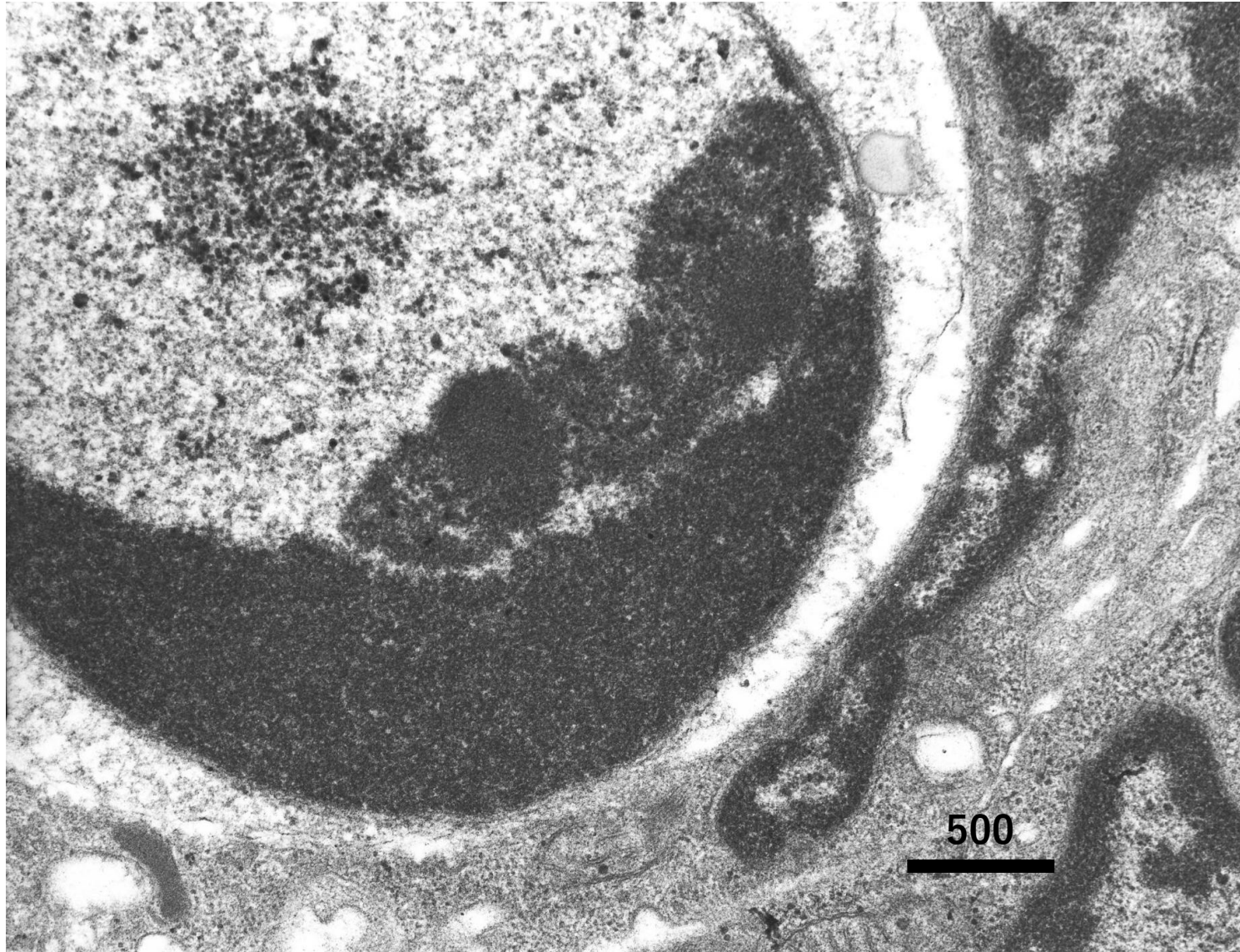
Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Plasma cells are not seen in the lamina propria mucosae. Note the presence of apoptotic intraepithelial lymphocytes with distinct chromatin margination. EM-3



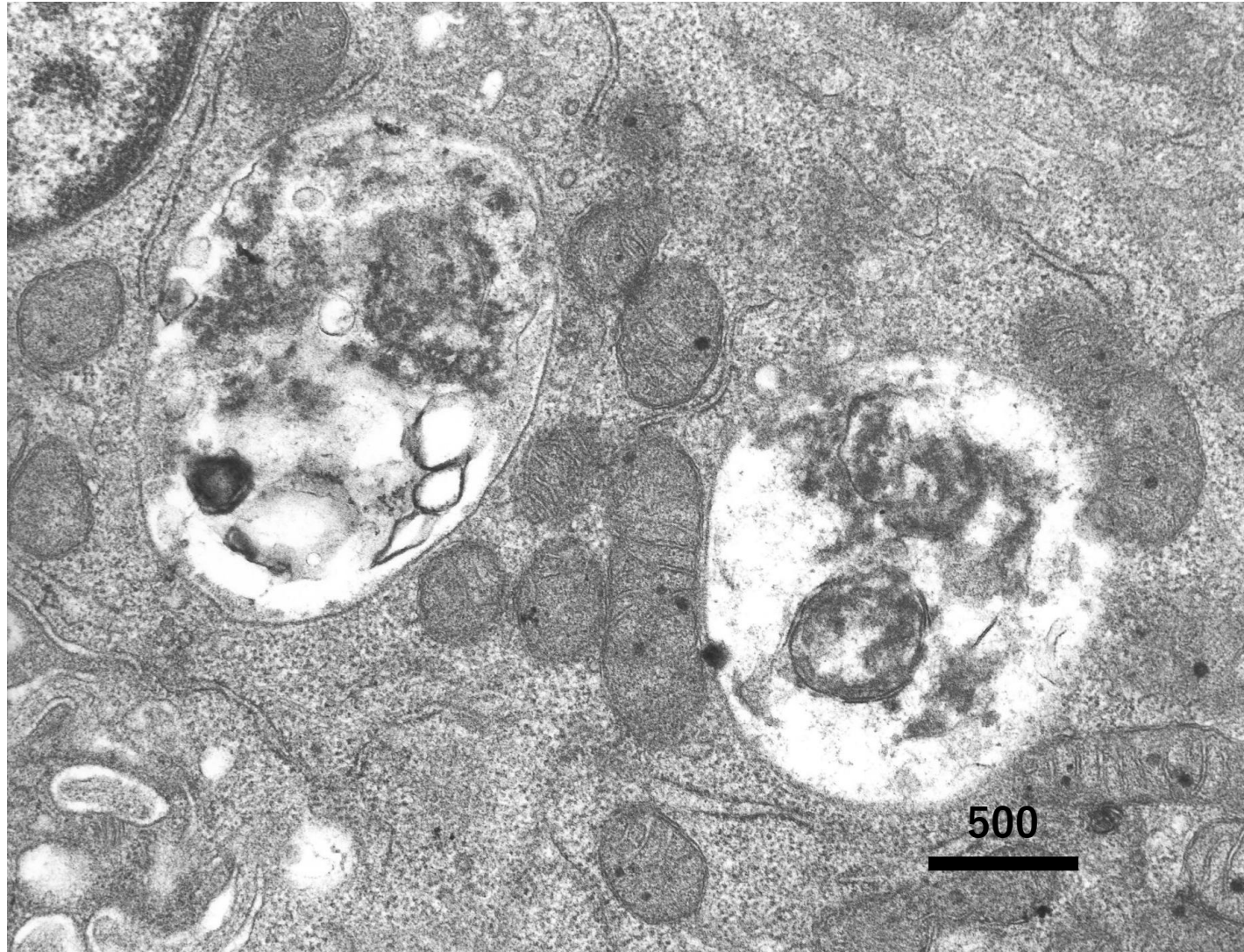
Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Note the presence of apoptotic intraepithelial lymphocytes with distinct chromatin margination. EM-4



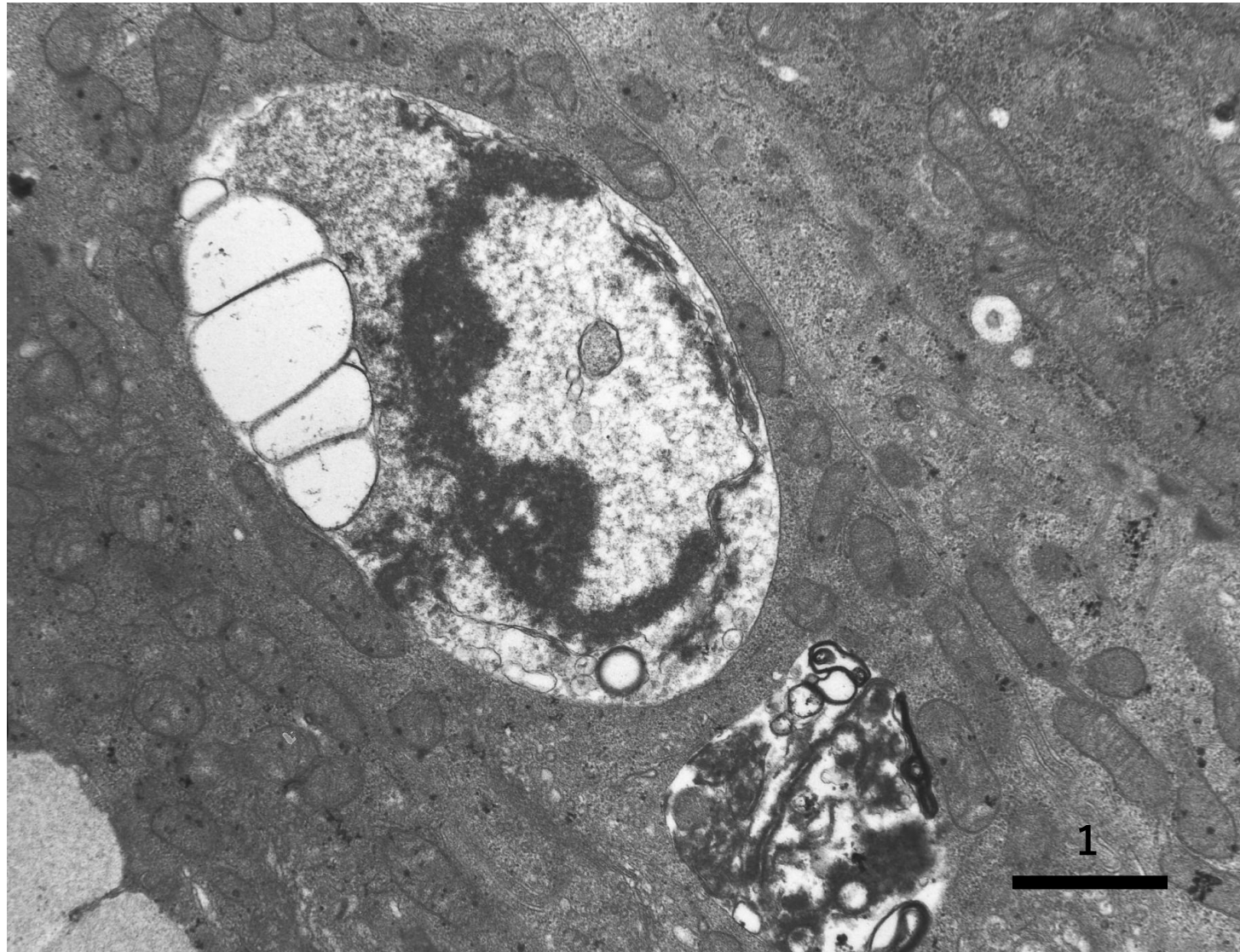
Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Note the presence of apoptotic intraepithelial lymphocytes with distinct chromatin margination. Autophagic vacuoles are observed in the supranuclear cytoplasm. EM-5



Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Note the presence of apoptotic intraepithelial lymphocytes with distinct chromatin margination. EM-6



Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Autophagic vacuoles are observed in the cytoplasm. EM-7



Ultrastructure of non-infectious chronic colitis in common variable immune deficiency (CVID). Colon biopsy was sampled from a 6-year-old boy complaining of intractable diarrhea. Autophagic vacuoles are observed in the cytoplasm. EM-8