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The patient was an 8-year-old boy known case of hyper IgM (HIGM) syndrome since the age of 2 years with vomiting, abdominal pain, and chronic diarrhea for two months. He underwent upper gastrointestinal endoscopy after unsuccessful empirical therapy. Histologic examination of duodenum revealed variable villus atrophy with round basophilic microorganisms on the surface measuring 2 to 5 micrometers in diameter (Figure 1). Laboratory tests revealed elevated IgM (506 mg/dL, normal range = 31–208) and decreased IgG (302 mg/dL, normal range = 572–1474), IgA (2 mg/dL, normal range = 27–195), and IgE (<0.1 IU/mL, normal range = up to 90) in keeping with the previous diagnosis of HIGM as well as mild leukocytosis (white blood cells = 12200/μL) and elevated alkaline phosphatase (556 U/L, normal range = 142–335). Therefore, the patient underwent magnetic resonance cholangiopancreatography (MRCP) which revealed thickening of the distal common bile duct (CBD) with mild dilation of CBD, pancreatic duct and intrahepatic ducts (IHD) with irregularity of the IHD along with their focal narrowing (Figure 2). Earlier MRCP findings were in favor of a recurrent cholangitis. Treatment with intravenous immunoglobulin (IVIG) and paromomycin (250 mg every 12 hours) was started for him. Nitazoxanide was not available in the Iranian pharmaceutical market.

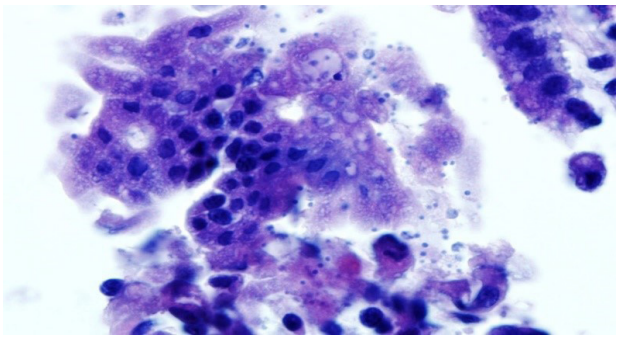

Figure 1. Duodenal biopsy showed small spherical microorganisms of variable size on the microvillus surface (Hematoxylin & Eosin, 1000x).

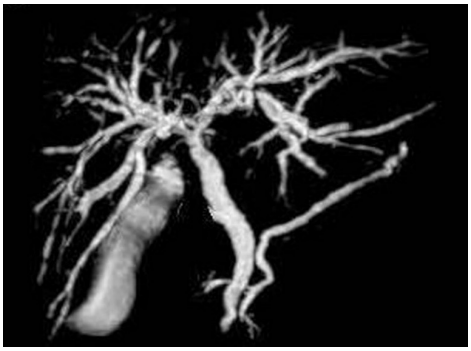

Figure 2. Magnetic resonance cholangiopancreatography (MRCP) showed common bile duct (CBD) thickening and dilation along with intrahepatic ducts (IHD) dilation, irregularity and focal narrowing.

What is your diagnosis?
See the next page for your diagnosis.

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Photoclinic Diagnosis

Cryptosporidiosis in Hyper IgM Syndrome

Cryptosporidium is a member of gastrointestinal apicomplexan protozoa which can cause self-limited watery diarrhea in immunocompetent patients and chronic or severe life-threatening diarrhea in immunocompromised individuals.[1] The two most common *Cryptosporidium* species are *Cryptosporidium hominis* and *Cryptosporidium parvum* which infect humans and animals like the cattle.[2] In children, especially in developing countries, cryptosporidiosis may cause malnutrition and failure to thrive (FTT).[1] In developed countries, *Cryptosporidium* is an important public health issue due to outbreaks. *Cryptosporidium* species can also result in traveler's diarrhea.[3]

HIGM is an immune deficiency condition which is defined as normal or increased IgM and decreased IgG, IgA, and IgE concentrations due to defective immunoglobulin class switch recombination. HIGM is rare with an incidence of 1–2: 1 000 000. *Cryptosporidium parvum* infection is common in HIGM. Chronic or prolonged diarrhea due to Cryptosporidium infection happens in approximately one-third of patients with HIGM and may cause FTT. It is also associated with biliary tract disease which may present as recurrent and/or sclerosing cholangitis.[4]

Antimotility drugs play an important role in the management of immunocompetent and immunocompromised patients with cryptosporidiosis, while the efficacy of antiparasitic drugs in cryptosporidiosis remains debatable, particularly in immunocompromised patients. A meta-analysis of trials of antiparasitic drugs in cryptosporidiosis showed nitazoxanide as an efficient antiparasitic drug in non-AIDS patients but revealed no significant efficacy for the other antiparasitic drugs in *Cryptosporidium* infection or for nitazoxanide in patients with AIDS.[5] A meta-analysis of human studies of the effect of paromomycin on treatment of cryptosporidiosis showed a response rate of 67% and long-term success rate of only 33% due to common relapses. Cryptosporidiosis treatment is a challenge in HIGM patients. Nitazoxanide and azithromycin might be beneficial in patients in the active phase of the disease but will rarely help its eradication. The best preventive measures for *Cryptosporidium* infection are avoidance of baths and swimming in non-chlorinated pools, lakes, or rivers.[4]

Authors' Contribution

MS and NZ involved in pathology interpretation, image preparation, and drafting the manuscript. PR involved in patient clinical management, and drafting the manuscript.

Conflict of Interest Disclosures

The authors have no conflicts of interest.

Ethical Statement

Informed consent was obtained from the patient's parents.

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