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Nodular Lymphoid Hyperplasia in a Patient Initially Believed to Have Familial Adenomatous Polyposis

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Abstract

A 50-year-old male was initially thought to have familial adenomatous polyposis (FAP) after innumerable small nodules in the upper GI tract were discovered upon endoscopic retrograde cholangiopancreatography for common bile duct stone extraction. ERCP was unsuccessful due to inability to find the major papilla amongst the nodules found in the duodenum. Biopsy of the nodules was consistent with nodular lymphoid hyperplasia. The patient was later found to have common variable immunodeficiency.

Introduction

Common variable immunodeficiency (CVID) is the second most prevalent primary immunodeficiency disorder after selective immunoglobulin A deficiency. Nodular lymphoid hyperplasia (NLH) of the gastrointestinal tract is a rare disorder associated with immunodeficiency syndromes, and characterized by numerous visible mucosal nodules. We report a patient with CVID and NLH initially mistaken for familial adenomatous polyposis (FAP).

Case Report

A 50-year-old male with past medical history of hypertension, polio infection, and cholecystectomy was referred for evaluation of persistent abdominal pain for the past 5 weeks. The patient complained of gastrointestinal reflux and sharp pain localized to the right upper quadrant exacerbated by fatty meals. Laboratory tests revealed an alkaline phosphatase (ALP) of 461 U/L, aspartate aminotransferase (AST) of 297 U/L, alanine aminotransferase (ALT) of 599 U/L, and a total bilirubin level of 0.6 mg/dL. Abdominal CT showed a common bile duct diameter of 1 cm. Initial attempt at endoscopic retrograde cholangiopancreatography (ERCP) was unsuccessful due to the inability to find the major papilla amongst the innumerable, 5–8-mm nodules found within the upper GI tract (Figure 1).

With familial adenomatous polyposis (FAP) as the working diagnosis, repeat ERCP resulted in successful biliary cannulation and extraction of CBD stones with resolution of the abdominal pain and normalization of the liver chemistry abnormalities. Biopsy of the nodules showed prominent hyperplastic lymphoid follicles with a reduced number of plasma cells in the lamina propria, consistent with nodular lymphoid hyperplasia. Further lab tests showed low levels of IgM, IgA, and IgG (11 mg/dL [34–214], <6 mg/dL [83–407], and 246 mg/dL [680–1,445], respectively), and the patient was referred to an immunologist. The patient admitted to annual episodes of sinusitis and bronchitis, and had persistently low levels of IgA and IgG. He was diagnosed with common variable immune deficiency (CVID) and treatment with intravenous immunoglobulin was initiated.

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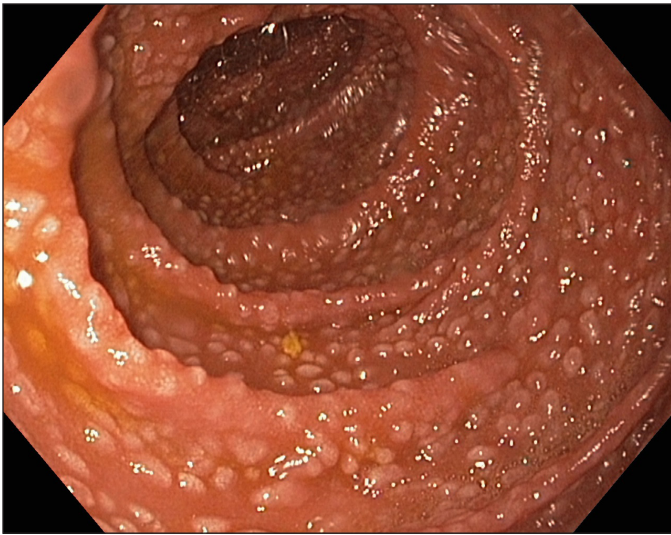


Figure 1. Endoscopic view of the second segment of the duodenum.

Discussion

Nodular lymphoid hyperplasia (NLH) is a lymphoproliferative disease that has been reported in patients with CVID, a disorder characterized by impaired B cell differentiation with defective immunoglobulin production. It is the most prevalent form of severe antibody deficiency affecting both children and adults. The clinical manifestations include recurrent sinopulmonary infections, autoimmune disorders, gastrointestinal disease, increased susceptibility to lymphoma, and a decreased immunogenicity to protein and polysaccharide vaccines. Histologically, there is enlargement of the mucosal B cell follicles, a normal-appearing mantle zone, and hyperplastic lymphoid follicles with large germinal centers within the lamina propria and superficial submucosa. Mahsa et al reported a case of NLH mimicking FAP wherein they attributed the misdiagnosis to similarities in the polyp-like appearance on endoscopy and large lymphoid follicles in the lamina propria on histology.¹ Studies have shown that infection with *Giardia lamblia* may create a nodular mucosal pattern.²

There have been suggestions that NLH is a risk factor for intestinal and extra-intestinal lymphoma.³ Abolhassani et al emphasized the need for an endoscopic surveillance protocol for gastric cancer in addition to immunoglobulin replacement therapy. They suggest looking for risk factors in CVID patients, such as *Helicobacter pylori* infection, pernicious anemia, or dyspepsia, prior to initiating screening esophago-

gogastroduodenoscopy (EGD). Frequency of follow-up EGD would be based on site and extent of gastric lesions.³

Disclosures

Author contributions: S. Altafi contributed to acquisition of data, drafting and revising of case report, and is the author guarantor. A. Volfson contributed to analysis of data and revising of case report. M. McKinley contributed to conception, acquisition, and analysis of data, and revising of case report.

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Informed consent was obtained for this case report.

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