Case Report

Inflammatory Duodenal Polyposis Associated with Primary Immunodeficiency Disease: A Novel Case Report

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Agammaglobulinemia is a rare form of B-cell primary immunodeficiency disease characterized by reduced levels of IgG, IgA, or IgM and recurrent bacterial infections. Agammaglobulinemia is most commonly associated with diffuse nodular lymphoid hyperplasia. Duodenal polyps are a rare entity; however, due to wide use of esophagogastroduodenoscopy, incidental diagnosis of duodenal polyps appears to be increasing. Although inflammatory duodenal polyposis has been reported in the literature, its association with common variable immunodeficiency has not been reported till date to the best of our knowledge. We report a case of a 59-year-old male with chronic symptoms of agammaglobulinemia associated with inflammatory duodenal polyposis.

1. Introduction

Duodenal polyps are rare entity; however, due to wide use of esophagogastroduodenoscopy, incidental diagnosis of duodenal polyps seems to be increasing. Similarly, agammaglobulinemia is a form of B-cell primary immunodeficiency disease characterized by reduced levels of IgG, IgA, or IgM and recurrent bacterial infections with normal T-cell immunity in 60% of patients. Being the largest immune organ, gastrointestinal tract is affected by agammaglobulinemia in a wide spectrum of symptoms and signs. Herein, we present a case of inflammatory duodenal polyposis associated with agammaglobulinemia in a male patient admitted for evaluation of chronic diarrhea in our hospital. To our knowledge, there is no such case in literature which depicts inflammatory duodenal polyposis seen in primary immunodeficiency disease. We emphasize the importance of considering agammaglobulinemia as a close differential diagnosis in a patient with duodenal polyposis by presenting this index case in a patient of chronic diarrhea. Prevalence of duodenal polyps is nearly 0.3%–0.5% and 4.6% as revealed by various retrospective [1, 2] and prospective study [3], respectively, on esophagogastroduodenoscopy. Although duodenal polyps may be pedunculated in nature, these polyps are sessile and small, measuring 3 mm–10 mm. Most of polyps occurring in duodenum are nonneoplastic, which include ectopic gastric mucosa, hyperplastic polyps, and Brunner’s gland hyperplasia. Inflammatory polyps contain ectopic gastric mucosa and are frequently present in duodenum [1, 2]. In the duodenal bulb, multiple polyps smaller than 10 mm do not need biopsy or treatment, whereas endoscopic surveillance and biopsy of duodenal polyps are important in patients with familial adenomatous polyposis [3], in which malignant transformation into adenomas or carcinoid tumours can be seen even if size is less than 10 mm and, hence, they need treatment.

2. Case Presentation

A 59-year-old male, Kurdish in origin, came to our hospital with history of recurrent chronic diarrhea for last eighteen years. Each episode of diarrhoea was lasting for more than a month and used to get relieved with antibiotics. The patient had also history of pulmonary tuberculosis and recurrent sinopulmonary infections. We evaluated him thoroughly for his chronic diarrhoea. His stool examination showed...
intestinal malabsorption, nodular lymphoid hyperplasia, and atrophied villi, inflammatory bowel disease, nonspecific chronic-atrophic gastritis, giardiasis, sprue-like disorder with high prevalence of infectious, inflammatory, and malignant infections characterized by a low concentration of antibodies alone [9]. A broad spectrum of histologic changes ranging from marked atrophy of villi and increased lymphocytes in mucosa resembling celiac disease to nodular lymphoid hyperplasia can be seen in patients with agammaglobulinemia [10]. Almost every patient with agammaglobulinemia suffers from acute, recurrent, or chronic infections, especially conjunctivitis, otitis, sinusitis, bronchitis, and pneumonia [2]. However, effects of malabsorption such as deficiency of vitamins and electrolytes may be present in severe cases. Patients with agammaglobulinemia have malabsorption involving carbohydrates, dietary fats, vitamin B12, and folate.

Patients with history of recurrent bacterial infections presenting with gastrointestinal manifestations, specifically chronic diarrhea, should be assessed for the possibility of agammaglobulinemia as any delay in the diagnosis and, hence, treatment can lead to significant morbidity and complications in patients with agammaglobulinemia. Treatment of gastrointestinal disorders in agammaglobulinemia with intravenous immunoglobulins alone may be ineffective in comparison to combination with immunomodulators such as azathioprine and 6-mercaptopurine, because most gastrointestinal manifestations of agammaglobulinemia appear to be due to defects in T-cell mediated immunity [1].

Prevalence of duodenal polyps is nearly 0.3%–0.5% and 4.6% as revealed by various retrospective [1, 2] and prospective studies [3], respectively, on esophagoduodenoscopy. Although duodenal polyps may be pedunculated in nature, these polyps are sessile and small, measuring 3 mm–10 mm. Most of polyps occurring in duodenum are nonneoplastic, which include ectopic gastric mucosa, hyperplastic polyps, and Brunner’s gland hyperplasia. Inflammatory polyps contain ectopic gastric mucosa and are frequently present in duodenum [1, 2]. In the duodenal bulb, multiple polyps smaller than 10 mm do not need biopsy or treatment, whereas endoscopic surveillance and biopsy of duodenal polyps is important in patients with familial polyposis [3]. Malignant transformation into adenomas or carcinoid tumours can be seen in some duodenal polyps having size less than 10 mm and, hence, need treatment. The clinical manifestations of nonneoplastic and neoplastic diseases of duodenum have not been identified so far. Duodenal polyps having hyperplastic (metaplastic) nature have seldom been identified. Data published on these polyps are restricted to case reports or small case reports [6, 8, 10]. They closely resemble hyperplastic polyps of the gastric type instead of the colonic type as they almost always arise in the context of ectopic mucosa of the stomach. Hyperplastic polyps of the duodenum seem to occur most frequently in patients with peptic ulcer disease or other gastric disorders [8] and can be associated with colonisation by *Helicobacter pylori* [7].

### 4. Conclusion

In conclusion, though there is no direct evidence of chronic diarrhea with inflammatory duodenal polypsis, the contributing factors for development of inflammatory polyps of
duodenum with agammaglobulinemia in our patient may be due to recurrent infections of the gastrointestinal tract. We have described a rare case of inflammatory duodenal polyposis coexisting with agammaglobulinemia in a patient with chronic diarrhea, and agammaglobulinemia should be considered in the list of differential diagnoses of inflammatory duodenal polyposis, especially when it is incidentally seen on esophagastroduodenoscopy of a patient with chronic diarrhea such as in our case. The study needs further reports to establish cause and effect relationship between chronic diarrhea and inflammatory duodenal polyposis.

Competing Interests
The authors declare that they have no competing interests.

Authors’ Contributions
Irfan Ali Shera, Sheikh Mudassir Khurshid, and Mohd Shafi Bhat were attending doctors for the patient. Irfan Ali Shera performed the esophagogastroduodenoscopy, colonoscopy, and wireless capsule endoscopy. Sheikh Mudassir Khurshid organized the report and wrote the paper. All the authors were involved in drafting and revising the manuscript, and all the authors read and approved the final manuscript.

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