

## Advanced gastric cancer, a very rare complication of Common Variable Immunodeficiency: a case report

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### ABSTRACT

**Background:** Common Variable Immunodeficiency (CVID) is associated with a broad spectrum of symptoms related to infections, chronic lung diseases, autoimmune diseases, and tumors; however, solid tumors are rare complications of CVID and gastric cancer, specially, has been reported very rarely.

**Case Presentation:** The present case report describes a 28 years old known case of CVID male, who was referred to us because of chronic diarrhea, weight loss and abdominal pain. He had history of recurrent respiratory infections but first manifestations of his recent problem had appeared six months before the referral as epigastric pain, vomiting, and watery diarrhea.

During his initial workup the patient had undergone partial gastrectomy surgery because of the pyloric stenosis. In early assessment of patient in our center, an ulcerative mass lesion with induration and rigidity in body and antrum of stomach was seen during upper GI endoscopy. Endoscopic ultrasound evaluation revealed thickening in body layers, expansion of tumor in 3<sup>rd</sup> and 4<sup>th</sup> layers, and 2-3 perigastric lymphadenopathy. Eventually, undifferentiated adenocarcinoma of the stomach was reported by histologic examination.

**Keywords:** Common Variable Immunodeficiency, gastric cancer, solid tumor.

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### INTRODUCTION

Common variable immunodeficiency (CVID) is a heterogeneous disorder of B-cells, which is characterized by marked reduction of serum levels of IgG, IgA or IgM, recurrent bacterial infections, impaired antibody response of B cells, and normal or near-normal T-cell immunity; however, the underlying pathophysiology of CVID is not well understood (1, 2). It probably results from a variety of gene defects. Moreover, most cases appear to be sporadic in origin, but at least 10 percent are

familial with an autosomal dominant pattern of inheritance (3, 4).

CVID is associated with a broad spectrum of clinical symptoms related to infections, autoimmune diseases, chronic lung diseases, liver and gastrointestinal disorders, granulomatous infiltrations, lymphoma, and solid tumors (4). Acute, chronic, or recurrent upper and lower respiratory tract infections have been observed practically in all patients with CVID (1). Chronic lung diseases, specifically the development of bronchiectasis, are common problems, often leading to hospitalizations, significant morbidity, and early death (1).

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There is a high prevalence of inflammatory, malignant, and infectious gastrointestinal disorders in patients with CVID. These include nodular lymphoid hyperplasia, inflammatory bowel disease (ulcerative colitis, ulcerative proctitis, or Crohn's disease), sprue-like illness with flat villi, pernicious anemia, giardiasis, and nonspecific malabsorption. Non-Hodgkin's lymphomas (NHL) occur at a markedly increased rate among these patients and CVID females have an increased risk of developing NHL compared to the age-adjusted expected incidence (5,6); however, solid tumors occur rarely in the setting of CVID, and gastric cancer, specifically, has been reported to happen rarely in association with this Immunodeficiency disorder. In fact, two large surveys have reported small clusters (two and three cases) of gastric cancers in CVID patients (1,3).

### CASE PRESENTATION

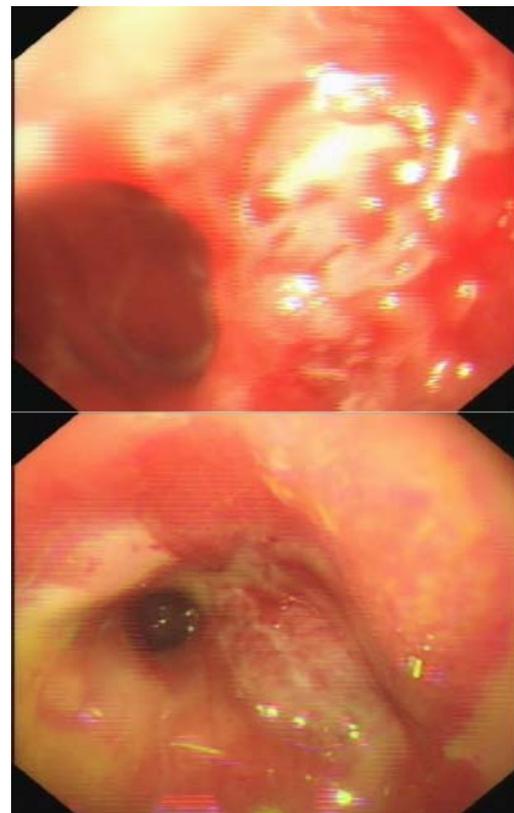
A 28-year old man was referred to our clinic for evaluation and assessment of chronic diarrhea, severe weight loss, and abdominal pain. He was a known case of CVID for 10 years, and was diagnosed primarily during the work-up for his recurrent respiratory infections. His bulky diarrhea had begun 5-6 months before referral with frequency of 3-4 times a day. Moreover, he had history of epi-gastric discomfort with radiating pain to back, early satiety and nausea/vomiting from 4 months preceding the referral, which had lead to diagnosis of pyloric stenosis and gastric outlet obstruction and sequentially, he had undergone partial gastrectomy. He had also lost about 20 kg of weight in this period.

In physical examination, the patient was severely cachectic and wasted away. He had obvious digital clubbing in both his hands and feet (Fig 1). In early assessment of patient with upper gastrointestinal endoscopy, an ulcerative mass lesion with induration in antral part, and rigidity in body and antrum of stomach was seen (Fig 2).

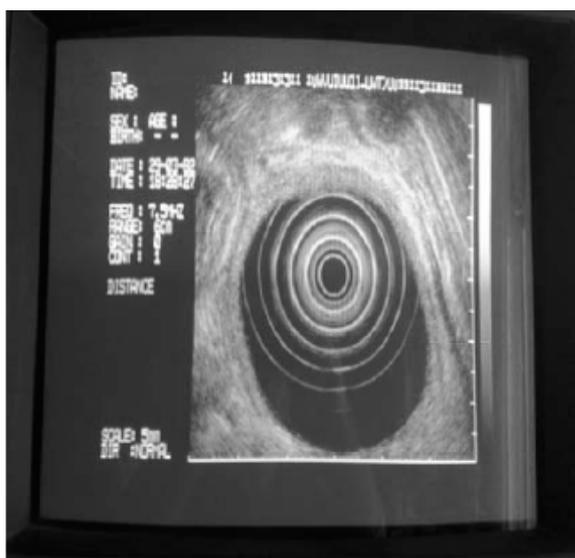
Further evaluation comprised endoscopic ultrasonography which revealed the thickening of gastric wall with expansion of tumor in its 3rd and 4th layers, and three lymph nodes with 5-10 mm size in peri-gastric region (Fig 3). Histological examination revealed undifferentiated intra-mucosal adenocarcinoma of stomach (Fig 4).



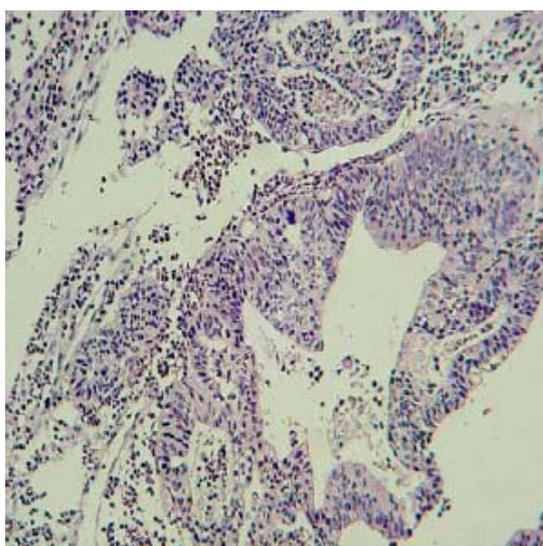
**Figure 1.** Digital clubbing



**Figure 2.** Upper GI Endoscopy



**Figure 3.** Endoscopic Ultrasonography of stomach



**Figure 4.** Histological examination. It revealed undifferentiated intra-mucosal adenocarcinoma of stomach

## DISCUSSION

As mentioned before, the solid tumors are rare complications of CVID, and, specifically, there is rare evidence of CVID related gastric cancers. In one, records of patients with CVID have been reviewed for 20 years and high incidence of lymphoma and gastric carcinoma is highlighted in CVID (3). In another study an enhanced risk of malignancy, granulomatous disease, and arthritis is noted (5). In the latter study, just one patient out of 98 patients who were followed from 1 to 13 years developed gastric cancer, while the incidence of other cancers especially NHL was much higher.

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