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CASE REPORT | STOMACH

# A Patient With Good Syndrome Complicated With Phlegmonous Gastritis

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## ABSTRACT

Phlegmonous gastritis (PG) is a rare and severe infection, with less than 500 cases reported. Similarly, Good syndrome represents a rare adult-onset immunodeficiency with a prevalence of 1 in 500,000 people. We present the first case of a patient with Good syndrome complicated with PG. Given that up to 50% of patients with PG do not have an identified risk factor, underlying immunodeficiencies should be conscientiously investigated.

## INTRODUCTION

Phlegmonous gastritis (PG) constitutes a rare but severe infection of the gastric submucosa and muscularis propria. It presents acutely with epigastralgia, nausea, emesis, and fever. Its mortality oscillates between 50% and 70%.[1–3] Good syndrome represents a rare adult-onset immunodeficiency associated with a thymoma. Patients have humoral and cellular immunodeficiency, which increases susceptibility to invasive bacterial infections.[4,5]

## CASE REPORT

A 37-year-old woman, with diagnoses of myasthenia gravis, thymoma (resected in June 2018), and primary hypothyroidism, presented in October 2018 with epigastralgia, nausea, and melena. An upper endoscopy revealed a friable, erythematous mucosa with active oozing bleeding and purulent discharge (Figure 1). Biopsies were obtained, which showed chronic and acute gastritis with abundant purulent material. *Streptococcus oralis* was isolated from gastric biopsies, which supported the diagnosis of PG (Figure 2). A thoracoabdominal computed tomography showed thickening of the gastric wall with mucosal enhancement and jejunojejunal intussusception (Figure 3). Considering the recently resected thymoma, immunoglobulin (Ig) levels were measured, which revealed normal IgA and IgM levels and low IgG levels (400 mg/dL, reference range [RR]: 635–1741 mg/dL). IgG subclasses levels showed low IgG1 (354 mg/dL, RR: 405–1,011 mg/dL), low IgG2 (89 mg/dL, RR: 169–786 mg/dL), low IgG3 (10 mg/dL, RR: 11–85 mg/dL), and normal IgG4 (10 mg/dL, RR: 3–201 mg/dL). A low CD4+ T cell count (404 cells/μL) further supported a diagnosis of Good syndrome. Seven weeks of antibiotics (ceftriaxone) led to partial symptomatic improvement without improvement on follow-up endoscopic studies, which prompted a total gastrectomy in December 2018. Specimen obtained from gastrectomy showed thickening of the gastric wall (Figure 4). Five months after surgery, the patient has had a favorable follow-up as an outpatient reporting mild epigastralgia occasionally, without any further episodes of gastrointestinal bleeding or infections.

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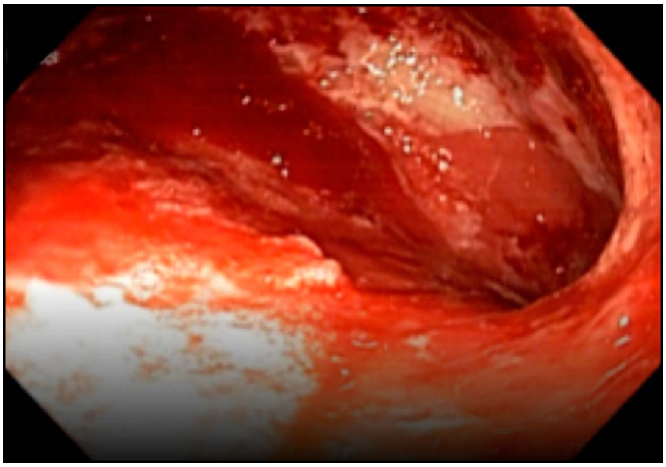

**Figure 1.** Upper endoscopy revealing a friable, erythematous mucosa with purulent discharge.

## DISCUSSION

Less than 500 cases of PG have been reported in the literature. Similarly, less than 200 cases of Good syndrome have been reported with an estimated prevalence of 1 in 500,000 people.[1,5] PG typically presents with acute epigastric pain and malaise, which occurred in our patient, although with a subacute presentation. Upper endoscopy, the diagnostic gold standard, classically revealed a thickened and edematous gastric mucosa and, occasionally, purulent discharge which is pathognomonic. Primary and secondary forms have been described. Primary PG occurs after injury to the gastric mucosa, either following an endoscopic intervention or in the presence of invasive carcinoma.[6] Secondary forms occur through adjacent/local (eg, pancreas) or hematogenous dissemination.[1] Alternatively, PG can be classified by the extent of involvement (localized or diffuse). Diffuse PG has higher mortality rates (10 vs 54%).[3,7] This case was classified as primary and diffuse.

*Streptococcus pyogenes* is isolated in approximately 70% of the cases, which contrasts with our case, where *S. oralis* was isolated.[1] This is the first case of PG where this microorganism has been isolated. Early antibiotic therapy with appropriate stewardship is essential, as demonstrated in this report.[1] Indication and timing of surgery represent a controversial therapeutic aspect of PG. Surgery has been associated with a reduced mortality rate (20% vs 50% in patients undergoing medical treatment).[3] Thus, it should always be considered in patients with a lack of response and/or worsening despite conservative treatment.[8] Our patient's stable clinical condition motivated conservative treatment but because of a suboptimal response, gastrectomy was indicated, leading to a favorable outcome.

Risk factors for PG include a history of gastrointestinal surgery, endoscopic procedures, hematologic malignant diseases (eg, leukemia), acquired and primary immunodeficiency disorders, alcoholism, and/or uncontrolled diabetes mellitus.[1–3] Still, up to

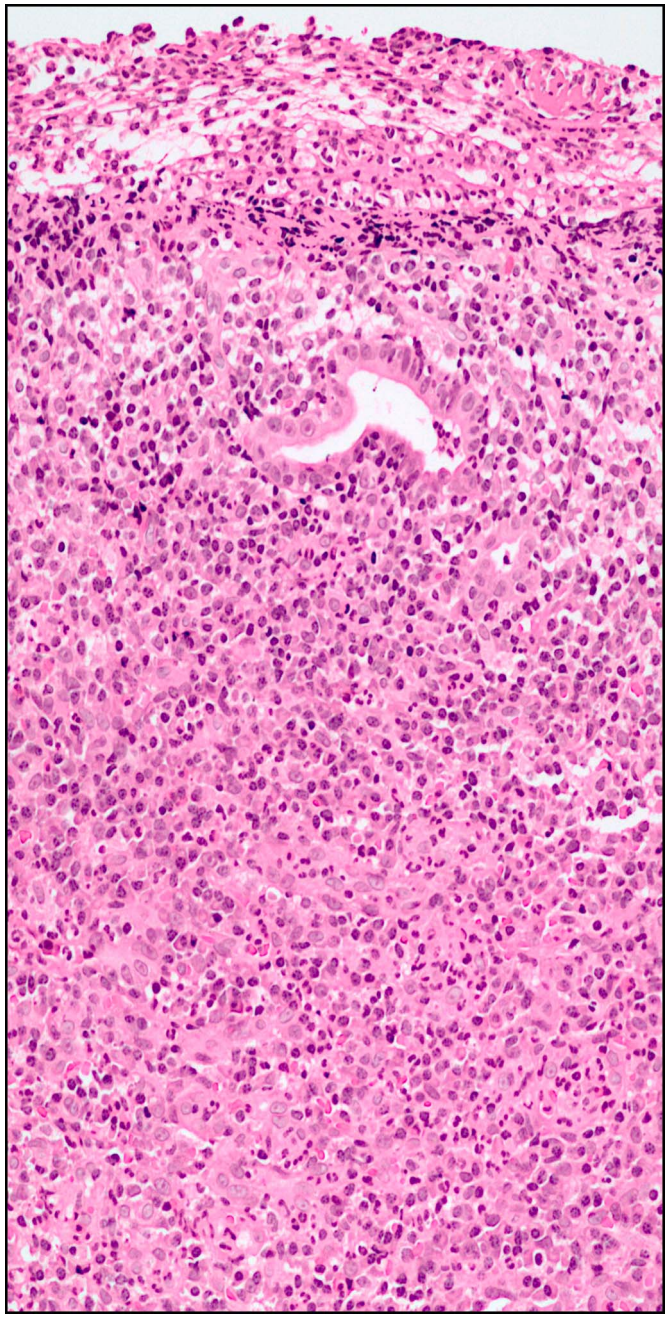

**Figure 2.** Gastric biopsy demonstrating chronic and acute inflammation (hematoxylin and eosin).

50% of patients do not have a clearly identified risk factor.[2,3] Good syndrome, the risk factor for PG in our patient, occurs in patients in their 50s and 60s, equally affecting both genders. Its pathogenesis remains unknown, although evidence suggests both primary bone marrow and/or T cell defect. Clinical presentation varies, and symptoms related to the thymoma, which may precede (up to 18 years) the diagnosis of Good syndrome in 42.4% of the cases, may be the first manifestations.[9] Alternatively, patients may present with recurrent invasive and/or opportunistic infections, respiratory tract infections being the most common.[4,5] In one case series, 90.5% of the patients

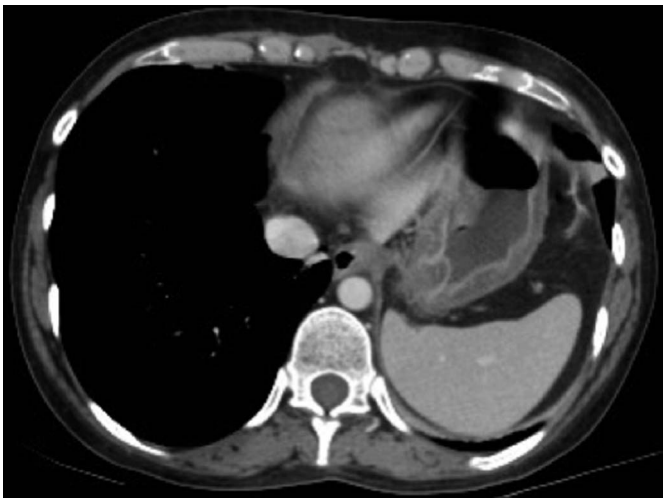

**Figure 3.** Abdominal computed tomography showing thickening of the gastric wall with mucosal enhancement.

debuted with invasive bacterial infections and 86% developed pneumonia; the most common bacteria were *Streptococcus pneumoniae* (33%) and *Haemophilus influenzae* (29%). Interestingly, concomitant autoimmune disease is present in 26%–76% of the patients, the most common being pure red cell aplasia, hypothyroidism, inflammatory arthritis, and myasthenia gravis, which is compatible with our case.[5,10]

According to case series and case reports, the laboratory findings in Good syndrome include hypogammaglobulinemia (100%), decreased B cells (87% of the cases), decreased CD4+ T cells (73.2%), and inverted CD4/CD8 ratio (76.1%), among others.[9] Our patient presented with decreased levels of every IgG subclass, except IgG4, and low CD4+ T count. Good syndrome should be considered in patients older than 40 years with atypical, opportunistic, and/or recurrent infections, particularly those with a diagnosed thymoma. Importantly, thymectomy (and its extent), although an important indicator of long-term prognosis, does not reverse the immunological abnormalities. Thus, acknowledging the quality of the evidence, we recommend that every patient with a previous or current diagnosis of thymoma should have immunoglobulin levels measured.[4,5]

Treatment includes long-term immunoglobulin replacement therapy, which may decrease the number of recurrent infections. Patients with Good syndrome have an overall survival rate of 70% at 5 years, whereas other primary immunodeficiencies, such as common variable immunodeficiency or X-linked agammaglobulinemia, have survival rates of almost 100% at 5 years.[4]

In conclusion, we report the first case of a patient with Good syndrome complicated with PG. Early and aggressive treatment of patients with PG should be the norm, always considering surgery in severe cases and/or in those failing conservative

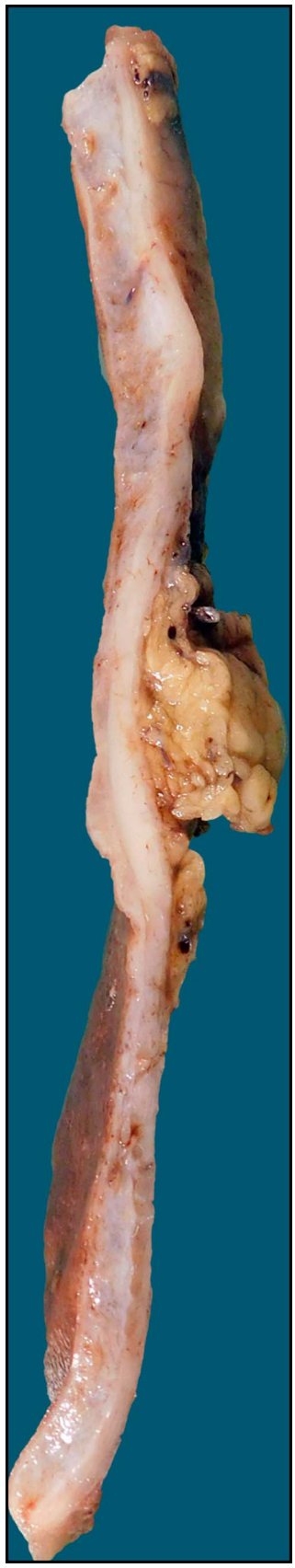

**Figure 4.** Specimen obtained from gastrectomy showing thickening of the gastric wall.

treatment. Risk factors, such as underlying immunodeficiencies, should be conscientiously investigated.

## DISCLOSURES

Author contributions: All authors contributed equally to this manuscript. A. Campos-Murguía is the article guarantor.

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