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Clinical Pictures: A Rare Case of IgG4-Related Disease Manifesting as an Ulcerated Gastric Cancer Complicated with Gastric Mucormycosis

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Case Series

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ABSTRACT

Background: IgG4-Related Disease (IgG4-RD) is a novel clinical entity that mimics many malignant, infectious and inflammatory disorders. Many organs or tissues arLe now known to be involved in IgG4-RD; However, there has been no report about IgG4-RD in the stomach with fungal infection.

Case presentation: We reported a 56-year-old male with IgG4-RD manifesting as ulcerated gastric cancer complicated with gastric mucormycosis. Both clinical and imaging manifestations were atypical, in clinical, it was highly suspected that the disease is a malignant tumor as ulcerated gastric cancer. However, no pathological evidence of malignancy could be found in repeated biopsies and surgically excised gastric specimens. Histological examination showed ulcer and inflammatory granulation with lymphocytes and plasma cells in sheets, obliterative phlebitis, formation of new fibrous tissue and inflammatory necrosis with hyphae and spores. The plasma cells had immunohistochemically strong staining for CD38, CD138, kappa, lambda, IgG and IgG4. And notably, IgG4/IgG positive plasma cell ratio was more than 40%, kappa/lambda positive staining ratio was approximately 1:1. Gastric mucormycosis was diagnosed with hyphae and spores confirmed by Gomori's methenamine silver staining and immunofluorescence and confocal laser-scanning microscopy. IgG4-RD manifesting as ulcerated gastric cancer complicated with gastric mucormycosis was diagnosed by multidisciplinary team discussion from the gastroenterology, radiology and pathology department. The patient vomited blood three times for the following month. An emergent exploratory laparotomy was conducted and a mass with 10 × 9 × 8 cm adhered closely to adjacent organs could be touched in the lesser curvature

of the gastric antrum. Anastomotic leakage occurred after the surgery and symptomatic treatments (negative pressure drainage and antibiotics) were applied without steroids and anti-fungus drugs. The patient gradually recovered and was advised to consult the gastroenterology department for further steroids and anti-fungus drug treatment.

Conclusion: To our knowledge, it is the first report about IgG4-related disease manifesting a gastric ulcer with mucormycosis infection.

Keywords: IgG4-related disease; Gastric ulcer; Mucormycosis; Kappa

Abbreviations: IgG4-RD: IgG4-Related Disease; CT: Computer Tomography; PET-CT: Positron Emission Tomography-Computed Tomography; EGD: Esophagogastroduodenoscopy; PPI: Proton Pump Inhibitors; IHC: Immunohistochemistry; ESR: Erythrocyte Sedimentation Rate; ENA: Extractable Nuclear Antigen; ANA: Antinuclear Antibody; ANCA: Anti-Neutrophil Cytoplasmic Antibodies; PR3: Anti-Protease 3; MPO: Anti-Myeloperoxidase; GBM: Glomerular Basement Membrane Antibody; PCR: Polymerase Chain Reaction; AIP: Autoimmune Pancreatitis; PBP: Plasminogen-Binding Protein; UBR2: Ubiquitin-Protein Ligase E3 Component N-Recognin 2.

INTRODUCTION

IgG4-Related Disease (IgG4-RD) is a chronic multi-organ-involved fibro inflammatory condition characterized by the tendency to form tumefactive lesions, increased serum levels of IgG4 (often but not always) and tissue infiltration by dense lymphocytes and IgG4-positive plasma cells. IgG4-RD is always confused with neoplasm, infectious and inflammatory disorders clinically and radiologically [1-3]. It is estimated that the incidence of IgG4-RD is 3.1 per 100,000 people (statistics from Japan) [4]. Multiple organs and tissues are now known to be involved in IgG4-RD including the pancreas, biliary duct, lacrimal/salivary glands, lymph node, lung, liver, kidney, thyroid gland, gastrointestinal tract, prostate, hypophysis, stomach, skin, aorta, joint, retro peritoneum, meninges and pleura [3,5-8]. IgG4-RD is highly sensitive to glucocorticoids, but it can lead to end stage organ failure and even death if unrecognized. Histopathology remains key to diagnosis because reliable biomarkers are lacking [9]. To our knowledge, there has been no report about IgG4-RD of the stomach manifesting as an ulcerated gastric cancer with mucormycosis infection.

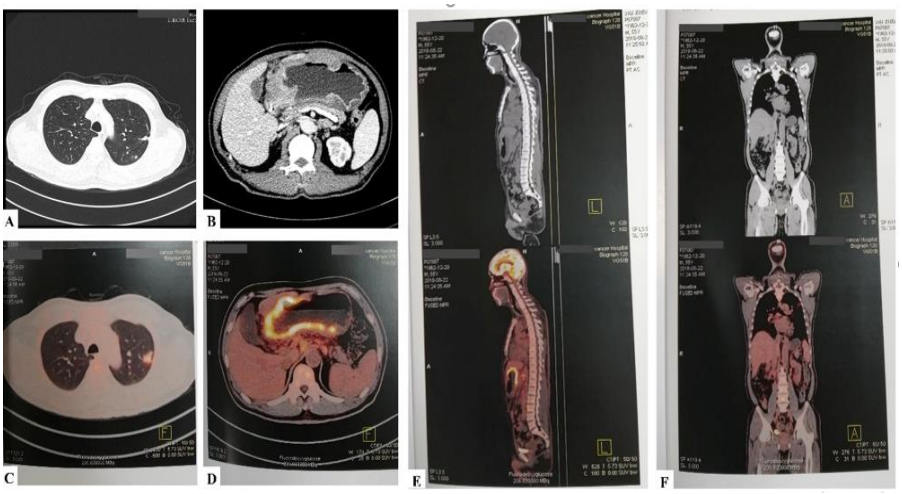
CASE PRESENTATIONS

A 56-year-old male presented with a fever and sought medical advice in the local hospital in August 2018. Laboratory tests revealed anemia (hemoglobin 81 g/L, reference range 130-175 g/L) and the Computer Tomography (CT) scan indicated gastric antral region irregular thickening, dilatation of bile ducts of liver left lobe and nodule in the left upper lung. Positron Emission Tomography-Computed Tomography (PET-CT) showed radioactive accumulations in the above position as well as retroperitoneal lymph nodes (Figures 1A-1F). A huge ulcer with 50 × 45 mm was observed by Esophagogastroduodenoscopy (EGD), whereas no evidence of the pathological diagnosis of stomach carcinoma.

The patient was treated with Proton Pump Inhibitors (PPI) and antibiotics to resist *Helicobacter pylori* for two weeks,

nevertheless, the ulcer still existed (50 × 50 mm) and no evidence of malignant cells pathologically as well. A laparoscopic removal of the mass was conducted with the rapid pathological diagnosis three times, neither the mass nor lymph node was positive for malignant tumor cells. Besides, no tumor cells were found by Immuno Histo Chemistry (IHC) detection in the postoperative tissues. Repeated fever occurred after the operation with intermittent anorexia, meteorism, vomiting, fatigue, diaphoresis and melena until February 2019.

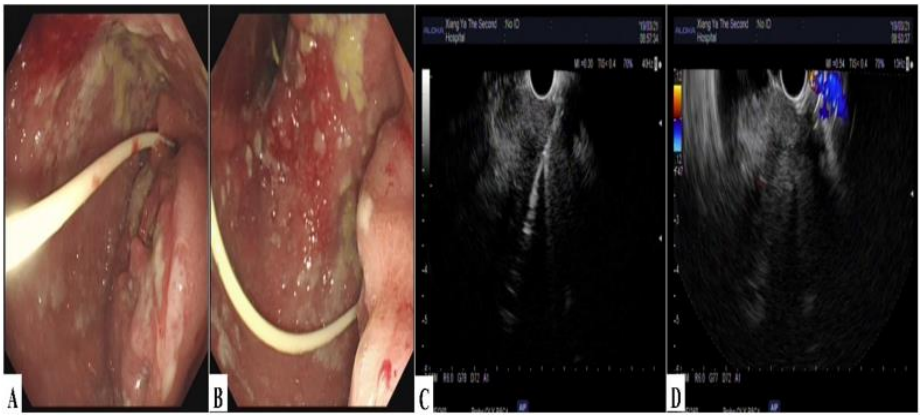
Figure 1. Imaging manifestations of this rare case. **Note:** (A,B) Contrast-enhanced CT indicated gastric wall thickening, perigastric and retroperitoneal lymph node enlargement and nodule in the left upper lung with double lungs exudation; (C-F) PET-CT showed radioactive accumulations in the above position as well as retroperitoneal lymph nodes.



During this period, the patient was in the hospital twice, laboratory tests still revealed anemia (hemoglobin 69 g/L and 74 g/L respectively in January and February 2019) and CT indicated the alteration of body and the antrum of the stomach (February 2019) and mass biopsy observed fungi but no tumor cells once again (February 2019). Treated with PPI, antibiotics, octreotide, nutrition agents and drugs for relieving symptoms, the patient remained to complain uncomfortably. The patient underwent an appendectomy at the age of 24 and he was diagnosed to type 2 diabetes in 2017. Pulmonary shadow was found in 2015 by routine physical examination. Patient had a history of tobacco smoking and alcohol drinking for 27 years and quit smoking for 3 years. There was no history of allergies, genetically related diseases and infectious diseases.

The patient visited the gastroenterology department of our hospital (The Second Xiangya Hospital, Central South University) in March 2019 for a clear diagnosis and treatment. The left subcostal subxiphoid mass could be palpated by physical examination. Main positive laboratory tests were as follows: Hemoglobin 81 g/L, globulin 49.4 g/L (reference range 20-40 g/L), Erythrocyte Sedimentation Rate (ESR) 121 mm/h (reference range 1-15 mm/h), IgG4 22.4 g/L (reference range 0.030-2.010 g/L), IgG 22.6 g/L (reference range 7.51-15.6 g/L), IgM 0.3 g/L (reference range 0.46-3.04 g/L), IgE>6000 ng/mL (reference range 0-691 ng/ml), *Helicobacter pylori* weakly positive (blood test), faecal occult blood tests weakly positive. The following laboratory results were negative: Blood amylase, lipase, G-test, GM test, Extractable Nuclear Antigen (ENA), Antinuclear Antibody (ANA), Anti-Neutrophil Cytoplasmic Antibodies (ANCA), Anti-Protease 3 (PR3), Anti-Myeloperoxidase (MPO), Glomerular Basement Membrane Antibody (GBM), Human Immunodeficiency Viruses (HIV), Hepatitis B Virus (HBV) and Epstein-Barr Virus (EBV). Multiple lymph nodes of bilateral axillas (largest 5 × 4 mm) and inguinal regions (largest 9 × 4 mm) were found by ultrasound (Figures 2A-2D).

Figure 2. Esophagogastroduodenoscopy images of this rare case. **Note:** (A,B) A huge ulcer was found by esophago gastro duodenoscopy; (C,D) The ultrasound gastroscope showed gastric wall thickening of the antrum.

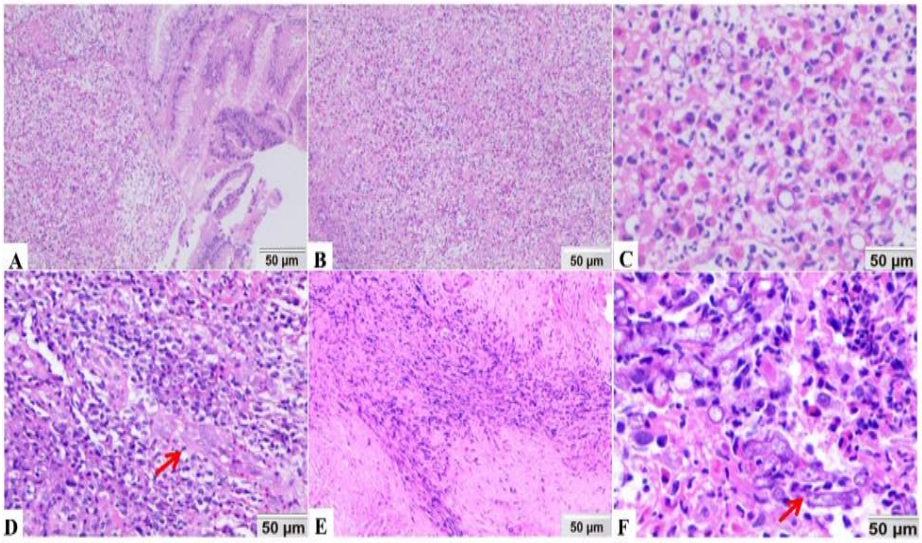


Contrast-Enhanced (CT) indicated the appearance of gastric wall thickening, the status of perigastric and retroperitoneal lymph node enlargement and a nodule in the left upper lung with double lung exudation (Figures 1A and 1B). Sputum smear found Gram-positive cocci and bacillus as well as Gram-negative cocci, but no fungus, whereas esophageal mucosa smear could see hyphae and spores.

Pathological findings

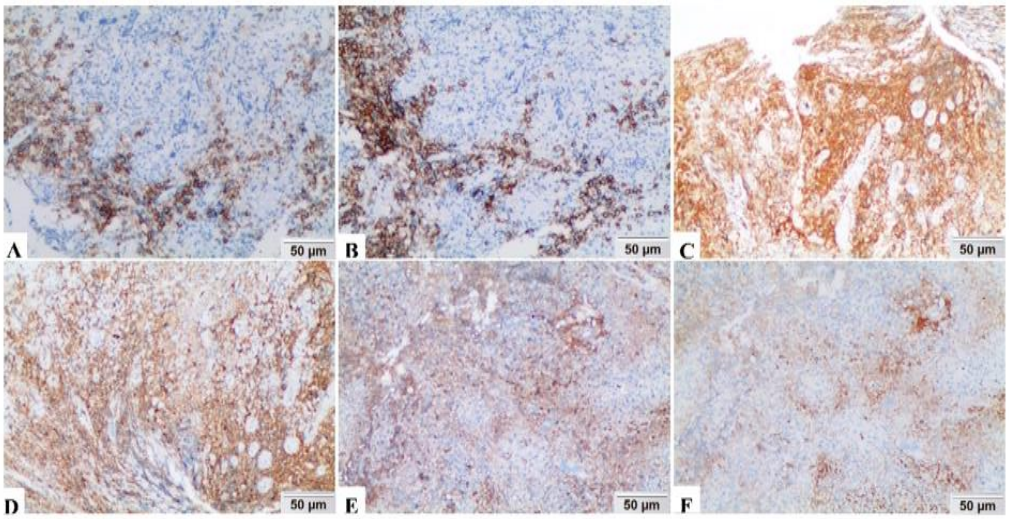
Microscopically, ulcer and inflammatory granulation could be seen with lymphocytes and plasma cells in sheets, obliterative phlebitis, formation of new fibrous tissue and inflammatory necrosis with hyphae and spores also existed (Figures 3A-3F).

Figure 3. The Hematoxylin and Eosin (HE) staining for the typical microscopic findings of this case (biopsy specimen). Microscopically, (A) Ulcer (100X); (B) Inflammatory granulation (100X) could be seen with plasma cells in sheets; (C) Lymphocytes (200X); (D) Obliterative phlebitis (200X); (E) Formation of new fibrous tissue (100X) hyphae and spores also be observed; (F) Inflammatory necrosis (400X).



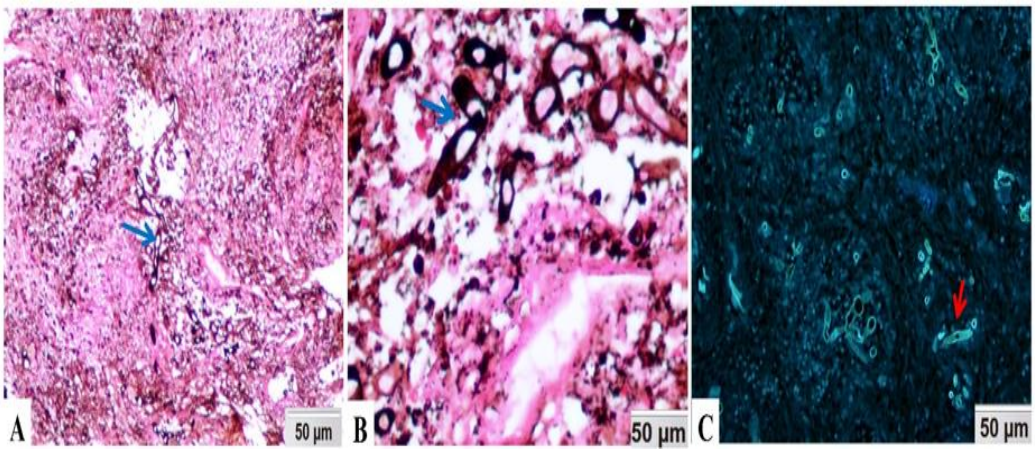
Immunohistochemically, there were cells with positive staining for CD38, CD138, CD10, MUM1, kappa, lambda, IgG and IgG4, focal positive staining for CD5, CD3, Bcl-2 and TIA-1 were observed; there were negative staining for Bcl-6, P53, GrB, PAX-5, CD30, CD20, CD56. The ki-67 index was 40% (Figures 4A-4F).

Figure 4. The typical Immunohistochemistry (IHC) staining for this case (biopsy specimen). The plasma cells had immunohistochemically strong staining. **Note:** (A) CD38 (100X); (B) CD138 (100X); (C) Kappa (100x); (D) Lambda (100x); (E) IgG (100x); (F) IgG4 (100x).



The positive expression of Cytokeratins (CK) in normal gastric epithelial cells, but no tumor cell was found again. Worthy to mention was that IgG4/IgG was more than 40%. Mucormycosis was diagnosed with hyphae and spores confirmed by Gomori's methenamine silver staining (Figures 5A and 5B) and immunofluorescence and confocal laser-scanning microscopy (Figure 5C). Due to the suspicions of lymphoma in clinical and image, we further completed gene rearrangement of B lymphocytes and the result was negative. Therefore, it was highly suspected for this patient of IgG4-RD presented as an atypical gastric ulcer complicated with gastric mucormycosis through the multidisciplinary team discussion.

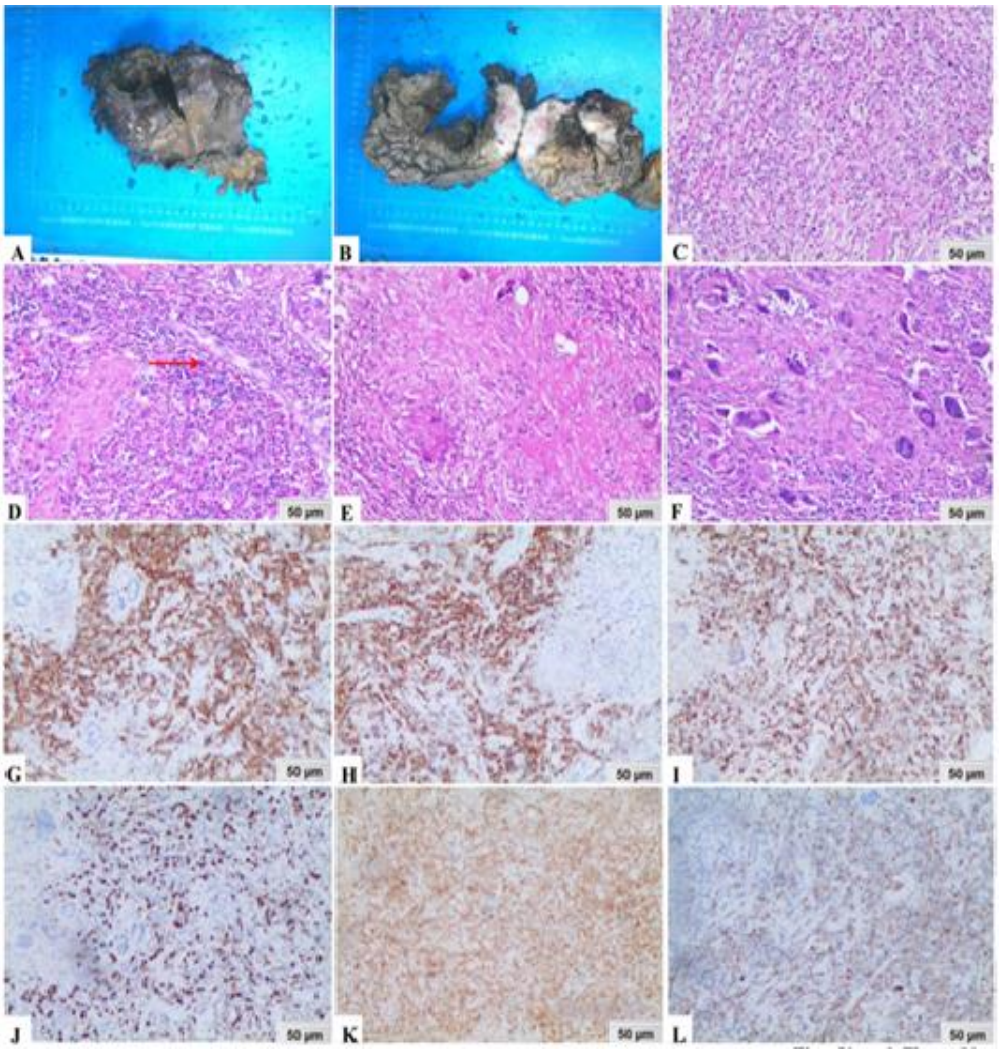
Figure 5. The typical images for hyphae and spores of mucormycosis of this case. Gomori's methenamine silver staining for hyphae and spores of mucormycosis. **Note:** (A) 100X; (B) 400X. Immunofluorescence and confocal laser-scanning microscopy were used to characterize hyphae and spores of mucormycosis; (C) 200X.



The patient vomited blood three times for the following month (dark-red, approximately 150 mL, 10 mL and 70 mL respectively). An emergent exploratory laparotomy was conducted and a mass with 10 × 9 × 8 cm adhered closely to adjacent organs could be touched in the lesser curvature of the gastric antrum. The gastrectomy, abdominal lymph node biopsy and jejunal nutrition tube placement were then conducted. Pathological examination showed the

inflammatory granulation with lymphocytes and plasma cells in sheets, obliterative phlebitis, formation of new fibrous tissue and multinuclear giant cells. The plasma cells had immunohistochemically strong staining for CD38, CD138, kappa, lambda, IgG and IgG4 (Figures 6A-6L). And the IgG4/IgG ratio was more than 40%, the kappa/lambda ratio was approximately 1:1, the ki-67 index was 70%. Tuberculosis was negative by Polymerase Chain Reaction (PCR).

Figure 6. The typical general/microscopic findings of this case (surgical specimen). **Note:** (A,B) The general specimen pictures of this patient; (C) Microscopically, inflammatory granulation could be seen with plasma cells in sheets and lymphocytes (100X); (D) Obliterative phlebitis (100X); (E) Formation of new fibrous tissue; (F) Multinuclear giant cells also existed (100X); (G-L) The plasma cells had immunohistochemically strong staining for CD38, CD138, kappa, lambda, IgG and IgG4 (100X).



Prognosis of the patients

Anastomotic leakage occurred after the surgery, treated with negative pressure drainage and antibiotics (Meropenem, following Levofloxacin) for a month. During this period, no steroids or anti-fungus drugs were used. The latest laboratory tests revealed IgG4 22.4 g/L one month after the surgery. The patient was advised to consult the gastroenterology department for further steroids and anti-fungus drug treatment. The last follow-up was in April 2023. The daughter of the patient disclosed that her father recovered well with occasional hiccups and no other discomfort and currently taking Chinese medicine for treatment.

RESULTS AND DISCUSSION

IgG4-RD is a systemic disorder characterized by the tendency to form tumefactive lesions, elevated levels of serum IgG4 (often but not always) and tissue infiltration by dense lymphocytes and IgG4-positive plasma cells in the affected organs [8]. Auto Immune Pancreatitis (AIP) is one of the most common disease features [10]. Kamisawa et al., first proposed that AIP was not simply pancreatitis but that it was a pancreatic lesion involved in IgG4-RD with extensive organ involvement and they defined IgG4-RD as a new clinicopathological entity in 2003 [11]. Actually, IgG4-RD has been described in virtually every organ system including the pancreas, biliary duct, lacrimal/salivary glands, lymph node, lung, liver, kidney, thyroid gland, gastrointestinal tract, prostate, hypophysis, stomach, skin, aorta, joint, retroperitoneum, meninges and pleura [3,5-8].

It is widely believed that hereditary susceptibility, autoimmunity and infectious agents are potential factors to lead to IgG4-RD [12]. Kawa et al., show that the frequencies of DRB1*0405 and DQB1*0401 are significantly higher in patients with AIP compared with chronic calcifying pancreatitis [13]. Their group also reveals that the Human Leukocyte Antigen (HLA)-linked genetic basis for Automatic Investment Plan (AIP) seems to be controlled by the DRB1*0405-DQB1*0401 mini-haplotype and the Adenosine Triphosphate Binding Cassette Subfamily F Member 1 (ABCF1) gene area [14]. Besides, the Plasminogen-Binding Protein (PBP) of *Helicobacter pylori* and Ubiquitin-Protein Ligase E3 Component N-Recognin 2 (UBR2) expressed by acinar cells of the pancreas are homologous in the peptide. 94% patients with autoimmune pancreatitis (33/35) can be detected antibodies against the Penicillin-Binding Proteins (PBP) peptide, while can be detected by 5% of patients with pancreatic cancer (5/110), indicating that infectious factors and molecular mimicry may initiate IgG4-RD [15].

Moreover, the innate and adaptive immune system might synergize to promote the progression of IgG4-RD and related tissue or organ fibrosis [16]. Microenvironment disorders are of critical importance for the pathogenesis of IgG4-RD, including immune cells including regulatory T cell, regulatory B cell, T follicular helper 2 cell, eosinophilia, and M2 macrophage [17-19]. Inflammatory cytokines also participate in the IgG4-RD such as IFN-α, IFN-γ, IL-4, IL-10, IL-5, IL-13, IL-33 and TGF-β [16,20]. In fact, there are no specific pathogenic factors for IgG4-RD and its cognitive process is dynamic. It has been noticed that immune cells converge on target tissues, interacting with fibroblasts and promoting tissue remodeling, which is coincident with typical pathological manifestations [21]. Although genetic factors, infection and autoimmunity disorder may be involved in the pathogenesis of IgG4-RD, every hypothesis alone is not sufficient to explain it.

Since the incidence of IgG4-RD is quite low with the estimation of 3.1 newly diagnosed cases per 100,000 people (statistics from Japan), sometimes diagnosis may be missed or erroneous [4]. The multi-disciplinary involved method may be valuable for the diagnosis of IgG4-RD. There are no specific clinical symptoms or signs for IgG4-RD, usually common in other diseases or exerting symptoms of complications. Abdominal pain, nausea, vomiting and jaundice are more common in males, whereas swelling of the lacrimal glands and submandibular glands occurs more often in females.

In males, the most commonly involved organs are salivary glands, pancreas and lacrimal glands, while they are salivary glands, lacrimal glands and sinuses in females [22]. The serum IgG4 level is elevated in most patients with increased ESR, CRP, serum IgG level and total IgE level, but there are still some patients presenting normal serum IgG4 levels (largely varied in different research, from 2.5% to 40%) [23,24]. However, increased IgG4 level may occur in other diseases such as allergy and lymphoma.

Organ enlargements of involved tissues are the most common imaging findings. If lungs are involved, honey-combing or ground-glass opacification, pulmonary nodules and Broncho vascular bundle thickening may be found [18]. Both clinical and imaging manifestations are atypical, to some extent, which are likely to be misleading and confusing, especially for single or infrequent involved organs.

Pathological examination is necessary and indispensable for the diagnosis of IgG4-RD [9]. The typical microscopic findings are diffused lymphoplasmacytic infiltration with irregular and storiform fibrosis and obliterative phlebitis, the former two of which occur in most cases. Phlebitis without obliteration of the lumen and elevated numbers of eosinophils also exist sometimes, but no sensitive or specific effects for the diagnosis of IgG4-RD. Necrosis, discrete granuloma and extensive neutrophilic infiltration are important clues to exclude IgG4-RD. Immunohistochemically, staining for IgG and IgG4 is very important. The absolute number of IgG4-positive plasma cells should reach a certain standard in corresponding organs or tissues, moreover, IgG4/IgG positive plasma cell ratio should be more than 40% [19,25,26].

Although the pathological examination is quite important for the diagnosis of IgG4-RD, it can't be confirmed by microscopy alone; multidisciplinary collaboration and comprehensive consideration of clinical and imaging manifestations along with laboratory tests is really important, especially for single or infrequent involved organ. To our knowledge, IgG4-RD rarely involves the stomach. In the few reports available, almost every case is presenting multiple organ involvement accompanied by stomach related and no case with fungal infection. We summarize all literature on the gastric involvement of IgG4-RD (Supplementary Table 1).

In the above table, the majority of cases are multiple organs involved or confirmed by surgical specimen; no patient associates fungal infection. Microscopically, there are four zones in the base of the ulcer of surgical specimen: Inflammatory exudates, fibrinoid necrosis, granulation tissue and mature fibrous tissue. However, pathologists always accept biopsy tissues surrounding the base of the ulcer in the clinical practice for fear that gastric perforation and unnecessary operation, which undoubtedly brings benefit for patients. At the same time, biopsy specimen makes the diagnosis of IgG4-RD involving the stomach difficult, especially presenting with an ulcer lesion. Pathologists must build up a general impression based on the local findings. Patients with gastric IgG4-RD were more likely to be misdiagnosed as Gastrointestinal Stromal Tumor (GIST), gastric cancer or peptic ulcer disease and their clinical course involved resection (51.3%) or even gastrectomy [27].

CONCLUSION

In this case, the fungal infection makes diagnosis more confusing and challenging. Gastric mucormycosis may cause secondary changes for ulcer under the microscope; besides, the identification of specific fungus will influence clinical decision and treatment methods. Therefore, recognizing and understanding IgG4-RD is significant for both clinical and pathological doctors. There are some flaws of the case report. We have attempted to advise the patient conduct regular physical examination and modulate drugs accordingly; However, the patient doesn’t visit doctors regularly and take herbs for treatment. Moreover, the patient was infected the COVID-19 at the follow-up process, but there are no details of the complication of IgG4-RD and the COVID-19. As far as we know, it is the first report about IgG4-related disease manifesting as an ulcerated gastric cancer complicated with gastric mucormycosis. Multidisciplinary team discussion from the gastroenterology, radiology and pathology department is necessary and indispensable for disease diagnosis.

HIGHLIGHTS

The first report about the diagnosis as an IgG4-related disease manifesting as ulcerated gastric cancers complicated with gastric mucormycosis. Systematically reviewed pathological features in biopsy and surgical specimen of IgG4-related disease. Multidisciplinary team discussion is necessary and indispensable for the diagnosis.

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AUTHOR'S CONTRIBUTION

Yuting Zhan designed and revised the manuscript. Xinxue Ma and Shuangshi Fan wrote the manuscript and made the tables. Yang Yang helped to handle the surgical specimen and collect pathological figures. Songqing Fan helped to collect clinical statistics. Li Gu conducted esophagogastroduodenoscopy and provided pictures.

AUTHOR CONSENT

The work was approved by the Ethics Review Committee of the Second Xiangya Hospital of Central South University (Scientific and Research Ethics Committee, No. K022). It was confirmed that informed consent was obtained. It was confirmed that the study complies with all regulations.

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AVAILABILITY OF DATA AND MATERIALS

The dataset supporting the conclusions of this article is included within the article (and its additional file).

CONSENT FOR PUBLICATION

The patients were consent with this study.

COMPETING INTERESTS

The authors have no conflict of interest to disclose.

REFERENCES

1. Lohr JM, et al. IgG4-related diseases of the digestive tract. Nat Rev Gastroenterol Hepatol. 2022;19:185-197.
2. Kamisawa T, et al. IgG4-related disease. Lancet. 2015;385:1460-1471.
3. Avincsal MO, et al. The histopathology of IgG4-related disease. Curr Top Microbiol Immunol. 2017;401:45-60.
4. Masamune A, et al. Nationwide epidemiological survey of autoimmune pancreatitis in Japan in 2016. J Gastroenterol. 2020;55:462-470.
5. Cheong HR, et al. Immunoglobulin G4-related inflammatory pseudotumor presenting as a solitary mass in the stomach. Clin Endosc. 2016;49:197-201.

6. Mack S, et al. Recent advances in the management of autoimmune pancreatitis in the era of artificial intelligence. World J Gastroenterol. 2022;28:6867-6874.
7. Nasrullah A, et al. IgG4 related lung disease-a rare and novel mimic of malignancy and infections-a case series of three patients with a brief review of updated literature. Respir Med Case Rep. 2021;33:101452.
8. Bledsoe JR, et al. IgG4-related disease: Review of the histopathologic features, differential diagnosis and therapeutic approach. APMIS. 2018;126:459-476.
9. Lanzillotta M, et al. Advances in the diagnosis and management of IgG4-related disease. BMJ. 2020;369: m1067.
10. Nista EC, et al. Autoimmune pancreatitis: From pathogenesis to treatment. Int J Mol Sci. 2022;23:12667.
11. Kamisawa T, et al. A new clinicopathological entity of IgG4-related autoimmune disease. J Gastroenterol. 2003;38:982-984.
12. Vaglio A, et al. IgG4-related disease. N Engl J Med. 2012;366:1646-1647.
13. Kawa S, et al. HLA DRB10405-DQB10401 haplotype is associated with autoimmune pancreatitis in the Japanese population. Gastroenterology. 2002;122:1264-1269.
14. Ota M, et al. Two critical genes (*HLA-DRB1* and *ABCF1*) in the HLA region are associated with the susceptibility to autoimmune pancreatitis. Immunogenetics. 2007;59:45-52.
15. Frulloni L, et al. Identification of a novel antibody associated with autoimmune pancreatitis. N Engl J Med. 2009;361:2135-2142.
16. Torre ED, et al. Immunology of IgG4-related disease. Clin Exp Immunol. 2015;181:191-206.
17. Ming B, et al. Role of eosinophilia in IgG4-related disease. Clin Exp Rheumatol. 2022;40:1038-1044.
18. Beck TC, et al. Immunoglobulin G4-related disease-associated dermatitis with pruritus: A positive response to Dupilumab. Life (Basel). 2023;13:833.
19. Kawakami T, et al. Abundant A Proliferation-Inducing Ligand (APRIL)-producing macrophages contribute to plasma cell accumulation in immunoglobulin G4-related disease. Nephrol Dial Transplant. 2019;34:960-969.
20. Minaga K, et al. Identification of serum IFN-α and IL-33 as novel biomarkers for type 1 autoimmune pancreatitis and IgG4-related disease. Sci Rep. 2020;10:14879.
21. Perugino CA, et al. IgG4-related disease: An update on pathophysiology and implications for clinical care. Nat Rev Rheumatol. 2020;16:702-714.
22. Wang L, et al. Sex disparities in clinical characteristics and prognosis of immunoglobulin G4-related disease: A prospective study of 403 patients. Rheumatology (Oxford). 2019;58:820-830.
23. Lin W, et al. Clinical characteristics of immunoglobulin G4-related disease: A prospective study of 118 Chinese patients. Rheumatology (Oxford). 2015;54:1982-1990.
24. Deshpande V, et al. Consensus statement on the pathology of IgG4-related disease. Mod Pathol. 2012; 25:1181-1192.
25. Maccagno A, et al. IgG4-Related pseudotumours: A series of 12 cases and a review of the literature. Pathology. 2022;54:563-572.
26. Chen L, et al. IgG4-related disease: What a hematologist needs to know. Haematologica. 2019;104:444-455.
27. Sawada H, et al. Clinicopathological characteristics of gastric IgG4-related disease: Systematic scoping review. J Gastroenterol Hepatol. 2022;37:1865-1872.