

88426.docx

**Gastric IgG4-related disease mimicking a gastrointestinal stromal tumor in a child: A case report**

Lin HCA *et al.* Gastric IgG4-related disease

Hsin-Chia Angela Lin, Kam-Fai Lee, Tzu Hao Huang

**Abstract****BACKGROUND**

Gastric IgG4-related disease (IgG4-RD) is rarely encountered in clinical practice, and especially more so among pediatric patients. To our knowledge, this is the first report of IgG4-RD presenting as a calcifying gastric mass in a child. We describe how this entity was difficult to differentiate from a gastrointestinal stromal tumor (GIST) imaging-based approaches. Therefore, this case highlights the importance of considering IgG4-RD in the differential diagnosis of gastric tumor before performing surgical resection, especially to distinguish it from malignancy to avoid unnecessary surgery.

**CASE SUMMARY**

The patient suffered from epigastric pain for several days. Panendoscopy and computed tomography scan revealed a submucosal tumor. Differential diagnoses included GIST, leiomyoma, teratoma, and mucinous adenocarcinoma. However, laparoscopic proximal gastrectomy allowed for the definitive diagnosis of IgG4-related stomach disease.

**CONCLUSION**

We emphasize the importance of considering IgG4-RD in the differential diagnosis of gastric submucosal tumors before performing surgical resection.

**Key Words:** IgG4-related disease; Gastrointestinal stromal tumor; Child; Pediatric; Case report

Lin HCA, Lee KF, Huang TH. Gastric IgG4-related disease mimicking a gastrointestinal stromal tumor in a child: A case report. *World J Clin Cases* 2023; In press

**Core Tip:** Gastric IgG4-related disease (IgG4-RD) is rarely encountered in clinical practice, especially among pediatric patients. To our knowledge, this is the first report of IgG4-RD presenting as a gastric calcifying mass in a child. This entity was difficult to differentiate from a gastrointestinal stromal tumor by imaging-based approaches. Therefore, it is important to consider IgG4-RD in the differential diagnosis of gastric tumor before performing surgical resection, especially to distinguish it from malignancy and avoid non-essential surgery.

## **INTRODUCTION**

IgG4-related disease (IgG4-RD) affects many organ systems, including the pancreato-hepato-biliary system, orbits, salivary glands, retroperitoneum, aorta, kidneys, lungs, and lymph nodes<sup>[1]</sup>. Gastric IgG4-RD is relatively rare, especially in children. Herein, we report an unusual case of definite IgG4-RD mimicking a gastric calcifying mass in a child.

## **CASE PRESENTATION**

### ***Chief complaints***

A 16-year-old girl presented with complaint of epigastric pain that had persisted for several days.

### ***History of present illness***

The patient reported experiencing hiccups, nausea, and vomiting. She did not exhibit fever, abdominal pain, or airway symptoms.

1

### *History of past illness*

The patient had no history of past illness.

### *Personal and family history*

The patient had no relevant personal and family history.

### *Physical examination*

The patient did not exhibit fever, abdominal pain, or airway symptoms.

### *Laboratory testing*

Blood test revealed elevated serum IgG4 concentration (244 mg/dL; normal range: 3-201 mg/dL).

### *Imaging examinations*

Endoscopy was first ordered and revealed a submucosal mass with intact mucosal appearance at the gastric fundus. Subsequent magnetic resonance imaging showed a calcifying submucosal tumor extending from the gastric fundus to the greater curvature of the upper body (Figure 1) and a hypodense lesion in segment IV of the liver. Abdominal ultrasound revealed a 2.72-cm poorly defined hyperechoic tumor in segment IV of the liver. Finally, additional magnetic resonance imaging confirmed the gastric tumor but showed no space-occupying lesion in the liver. According to the image report, the differential diagnosis of the gastric submucosal mass included gastrointestinal stromal tumor (GIST), leiomyoma, lipoma, schwannoma, *etc.*

### *Surgical examination*

Owing to the ongoing uncertainty of the etiology, the patient underwent pure laparoscopic wedge resection of the tumor. A biopsy of the liver lesion was also performed and frozen section histology was negative for malignancy. Histopathological

examination of the gastric specimen demonstrated prominent lymphoplasmacytic infiltration and increased proliferation of spindle myofibroblasts in the fibrotic stroma with focal storiform pattern. Immunohistochemical analysis showed 25-35<sup>3</sup> IgG4-positive plasma cells/high-power field with an IgG4+/IgG- ratio of 50% (Figure 2). Positive staining for smooth muscle actin was observed, but the tumor cells showed negativity for staining of c-kit, ALK, desmin, and S-100. Neither obliterative phlebitis nor tissue eosinophilia were present. Liver biopsy specimens revealed fatty changes. The patient made an uneventful postoperative recovery. Subsequent blood tests revealed elevated serum IgG4 concentrations (244 mg/dL).

### **FINAL DIAGNOSIS**

IgG4-RD of the stomach.

### **TREATMENT**

The patient received prednisolone and azathioprine.

### **OUTCOME AND FOLLOW-UP**

The patient returned for regular follow-up visits for 3 years and remained in stable condition throughout.

### **DISCUSSION**

According to the literature, gastric IgG4-RD mainly presents as wall thickening, gastritis, nodules, polypoid lesions, mass-like lesions, ulceration, vasculitis, and fistula<sup>[1-3]</sup>. The ages of patients reported with IgG4-RD gastric polypoid or mass-like lesions, regardless of other organ involvement, have ranged between 27 years to 77 years<sup>[1,3,4]</sup>. However, to our knowledge, IgG4-RD presenting as gastric mass lesions has not been described in children. Our patient is, thus, the first case of IgG4-RD mimicking a gastric calcifying mass in children, which could be confused with GIST.

GIST is usually treated through surgical resection, whereas the general first-line therapy for IgG4-RD is corticosteroids. Zhang *et al*<sup>[5]</sup> previously reported a case of suspected IgG4-RD presenting as a gastric calcifying fibrous tumor. They postulated that calcifying fibrous tumors may represent different stages of IgG4-RD, even though their case had normal serum IgG4 levels<sup>[5]</sup>. Our case demonstrates that gastric mass lesions of definite IgG4-RD can also occur in children. IgG4-RD is difficult to differentiate from GIST using imaging-based approaches. Therefore, we emphasize the importance of considering IgG4-RD in the differential diagnosis of gastric tumors before performing surgical resection, especially to distinguish it from malignancy to avoid unnecessary surgery.

8%

SIMILARITY INDEX

PRIMARY SOURCES

|   |  |               |
|---|--|---------------|
| 1 | <a href="http://www.wjgnet.com">www.wjgnet.com</a><br>Internet   | 26 words — 3% |
| 2 | <a href="http://f6publishing.blob.core.windows.net">f6publishing.blob.core.windows.net</a><br>Internet | 14 words — 1% |
| 3 | <a href="http://link.springer.com">link.springer.com</a><br>Internet                                   | 14 words — 1% |
| 4 | <a href="http://unte.uk.to">unte.uk.to</a><br>Internet   | 12 words — 1% |
| 5 | <a href="http://worldwidescience.org">worldwidescience.org</a><br>Internet                             | 12 words — 1% |

EXCLUDE QUOTES ON  
EXCLUDE BIBLIOGRAPHY ON

EXCLUDE SOURCES < 12 WORDS  
EXCLUDE MATCHES < 12 WORDS