

Gastric Inflammatory Myofibroblastic Tumor in a 10-Year-Old Patient in Macao—Case Report and Literature Review

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Abstract

Inflammatory myofibroblastic tumor (IMT) is a special type of mesenchymal tumors. The tumors can occur all over the body. The most common organs involved were lung, followed by mesentery, omentum, retroperitoneum, and pelvic cavity. But it occurs very rarely in the stomach. This article mainly reports a 10-year-old patient who complained of progressive dysphagia for 4 years. Esophagogram suggests esophageal achalasia. Abdominal CT examination revealed a huge mass with calcification in the cardiac region of stomach with metastatic lymphadenopathy. PET-CT was also performed that consistent with malignant gastric tumor with metastatic lymphadenopathy. Gastroscopy also indicated that there was a huge mass in the cardia that compressed the esophagus, and biopsy was performed to reveal chronic gastritis. Pathological analysis was performed after surgical exploration and tissue samples were taken out and the final pathology was consistent with inflammatory myofibroblastic tumors. The patient was not able to undergo surgical treatment, so crizotinib chemotherapy was used. After treatment, the patient's tumors were significantly reduced, and the effect was obvious. The patient is now in stable condition and continues to follow up. This article hopes to review the literature of imaging diagnosis of inflammatory myofibroblastic tumors through this case report, so as to improve the understanding of this disease.

Keywords

Computed Tomography, Inflammatory Myofibroblastic Tumor, Stomach

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a special type of mesenchymal

tumors. It is mainly composed of spindle fibroblasts and/or myofibroblasts, often accompanied by infiltration of inflammatory cells such as plasma cells, lymphocytes and eosinophils [1]. IMT is more common in children and adolescents, especially in women [2]. IMT can occur in all parts of the body, the most common site is the lung, followed by mesentery, omentum, retroperitoneum, pelvic cavity and so on [3]. It occurs very rarely in the stomach [4]. The main symptoms are gastrointestinal hemorrhage and intragastric mass. It is easy to be confused with gastrointestinal mesenchymal tumors such as gastric stromal tumors or leiomyomas. Now the author reports a case of gastric inflammatory myofibroblastic tumor and reviews relevant literature to analyze its clinical features, imaging manifestations, and prognosis in order to improve the understanding of the disease.

2. Case Report

A 10-year-old male presented with dysphagia for about 4 years and worsened for more than 6 months. After eating, he felt something stuck under his sternum, but his symptoms were relieved after drinking water. Over the past two months, repeated vomiting of food after each meal, worsening for 2 days, even vomiting after drinking water, defecation for about 1/4 - 5 days, urine volume decreased by about 20%, weight loss of about 5 kg in the past two months. The patient came to our hospital for consultation. Physical examination revealed mild dehydration and dry skin. Abdominal examination revealed mild tenderness in the left upper abdomen, no rebound pain, and normal bowel sounds. No hepatosplenomegaly. Laboratory examination showed microcytic hypochromic anemia (Hemoglobin level is 8.8), biochemistry profile showed unremarkably. Barium meal examination revealed dilated esophagus deviates to the right side. Narrowing at hiatus "Bird Beak" sign is noted (**Figure 1**). Esophagus achalasia suggested. Abdominal CT showed the wall of stomach at cardia region is irregular thickening (**Figure 2**). There are enlarged lymph nodes at the peri-gastric region. Gastric malignancy is suspected. Gastroscopy was performed and showed a circumferential neoplasm with irregular overlying mucosa was found at cardia and involved the lesser curvature wall of the upper body, biopsy was performed. The pathology showed chronic gastritis. PET-CT was performed that showed a markedly hypermetabolic gastric cardia lesion is seen with infiltration to the lesser curvature of gastric body and consistent with a malignant gastric tumor and metastatic lymph node at gastrohepatic ligament region suggested. The patient underwent laparotomy. During the operation, a giant mass was found in the cardiac region, about 4 cm in size. The surgeon performed the biopsy and sent for pathological examination. Postoperative pathology suggests inflammatory myofibroblastic tumors. The patient was unable to undergo surgical treatment, so crizotinib was given for chemotherapy. After chemotherapy, the tumors in the stomach were significantly reduced and the effect was obvious. Now the patient is stable and there are no adverse reactions. Patient discharged from hospital regularly follow up.

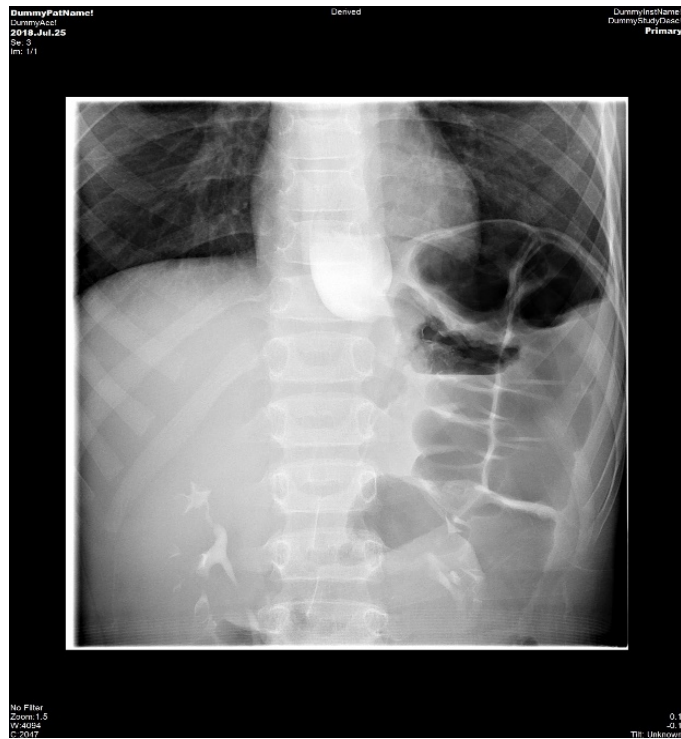


Figure 1. Barium meal examination revealed dilated esophagus deviates to right side. Narrowing at hiatus. “Bird Beak” sign.



Figure 2. Abdominal CT showed irregular thickening with calcification noted on the wall of lesser curvature of stomach.

3. Discussions

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal neoplasm with intermediate malignant potential. It was first discovered in the lung by Phillips in 1937 [5]. IMT mostly occurs in the lung and rarely in the stomach [6]. In 2002, WHO defined it as “tumors composed of differentiated myofibroblastic spindle cells, often accompanied by inflammatory cells such as plasma cells, lymphocytes, and eosinophils”. Biological behavior has been classified as intermediate (occasional metastasis) group [7]. According to the literature, some IMT cases may be related to gene mutation, virus infection or immune system regulation [8] [9] [10] [11]. The tumor is not sensitive to radiotherapy and chemotherapy, and surgical treatment is still the main treatment. IMT is more common in children and adolescents than in women. Most of the cases were in lung, mesentery, head and neck, and soft tissue of limbs. It occurs rarely in the stomach [12]. Clinical manifestations are often nonspecific. The main manifestations were fatigue, fever, abdominal pain, upper gastrointestinal bleeding, abdominal touch mass, weight reduction, gastroesophageal reflux, pallor, ascites [13]. Microcytic hypochromic anemia may ultimately result from chronic hemorrhagic polypoid ulcer in the stomach IMT [14]. It has been reported in literature that a few primary gastric IMTs are characterized by fever of unknown origin which is ineffective in antibiotic therapy. Their clinical course is long, ranging from several weeks to several months [15]. The main clinical manifestations of this patient were progressive dysphagia and microcytic hypochromic anemia. In most cases, endoscopic features of gastric IMTs are similar to those of malignant tumors. The gastroscopic manifestations of this patient are very similar to malignancies.

The radiological characteristics of inflammatory myofibroblastic tumor of the abdomen are nonspecific. Abdominal radiographs may show intestinal segment displacement caused by soft tissue masses and amorphous calcification in tumors. IMT of gastric wall can be determined by gastrointestinal fluoroscopy, but these findings are not specific. The typical image of IMT is thick, banded, irregular or punctate calcification, sometimes difficult to distinguish from achalasia [16]. Computed tomography showed calcification, solid, sometimes uneven, well-defined, sometimes lobular, globular masses. These imaging findings may indicate that invasive or multifocal diseases are difficult to distinguish from metastatic diseases [17]. CT imaging findings of IMT have been reported in literature. The main manifestations were clear subepithelial mass pattern and irregular wall thickening pattern. The mass showed contrast enhancement in varying degrees. The lesion may be accompanied by calcification or even direct invasion of adjacent organs [18]. Marvis *et al.* reported that invasive masses in patients with inflammatory myofibroblastic tumor presented as benign tumors on CT, but lymphadenopathy was not reliably detected, although it may be found during surgery [19]. They also pointed out that all reported IMTs had no CT correlation with lymphadenopathy. In our case, enhanced spiral CT showed signifi-

cant enhancement of multiple lymph nodes around the stomach. The CT findings of this patient were irregular mucosal thickening with calcification. It also accords with the performance mentioned in the literature. On MRI, lesions usually show low signal quality of T1-weighted and T2-weighted imaging and equal signal quality of gadolinium-enhanced T1-weighted imaging [20]-[26]. Endoscopic biopsy is often difficult to reflect the true situation of lesions due to superficial sampling, inadequate depth of biopsy and low volume of biopsy. The results of pathological biopsy are often negative. In our case, the depth of biopsy samples under gastroscope was insufficient, leading to false negative results.

Surgical resection is the preferred treatment for gastric IMT. The appropriate surgical method can be selected according to the location, size, depth of invasion and the patient's physical condition. It is not advocated that extensive lymph node dissection be performed [18] [27] [28] [29]. However, for some patients who fail to undergo surgical treatment, crizotinib can be used as a treatment for IMT. The response rate of crizotinib in the treatment of IMT has been reported to be 86% [30] [31]. The effect of crizotinib treatment was obvious in this case.

4. Conclusion

It is important to consider inflammatory myofibroblastic tumor in differential diagnosis of gastric calcified tumors, especially in childhood gastric tumors. Enhanced spiral CT is helpful in evaluating the location and extent of tumors. Therefore, the correct diagnosis can be made only when endoscopy results are combined with imaging findings.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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