Clinical Case Report

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Primary gastric inflammatory myofibroblastic tumor

A case report

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Abstract

Rationale: Primary gastric inflammatory myofibroblastic tumor is extremely rare. Only a few cases were reported in the domestic and foreign medical literature with corresponding imaging findings of this disease even more rarely reported.

Patient concerns: We present one case of a 52-year-old female patient with upper abdominal pain, acid reflux, and belching for 2 months.

Diagnoses and Interventions: Electron ultrasound gastroscopy (EUS) revealed elevation of gastric antrum mucosa. A whole abdominal and pelvic multi-slice spiral computed tomography (CT) detected a round nodule in the gastric antrum with considerably delayed enhancement, with initial suspicion of gastrointestinal stromal tumors (GISTS). Then a laparoscopic assisted distal gastrectomy was performed. Finally, the postoperative pathology confirmed the diagnosis of primary gastric IMT.

Outcomes: After 6 months of follow-up, the patient was still alive without any evidence of metastasis or recurrence.

Lessons: Familiarizing with the CT features of this rare tumor may raise radiologists' awareness of the disease and potentially could avoid misdiagnosis.

Abbreviations: CT = computed tomography, EUS = electron ultrasound gastroscopy, HU = Hounsfield units, IMT = inflammatory myofibroblastic tumor.

Keywords: computed tomography, primary gastric inflammatory myofibroblastic tumor

Key Points

- Primary gastric inflammatory myofibroblastic tumor (IMT) is a very rare type of mesenchymal tumor, increased awareness of the CT characteristics of this rare tumor may broaden the radiologist's knowledge base.
- This case report is the first to report the primary gastric inflammatory myofibroblastic tumor with ossification.
- Understanding the imaging features of primary gastric inflammatory myofibroblastic tumor is contribute to diagnose and further to treat it.

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1. Introduction

Primary inflammatory myofibroblastic tumor (IMT) is a very rare type of mesenchymal tumor. As reported, mainly found in children and young adults IMT commonly occurs in the lungs, but in extremely rare occasion, it could occur in adults' stomachs.[1,2] However, Katakwar et al[2] recently confirmed that IMT can occur in any organ of the body and in all age groups. It exhibits variable biological behaviors ranging from often benign lesions to more aggressive variants. IMT is locally recurrent but rarely metastasizes to distant organs.[3] Since patients with this type of tumor seldom cause specific clinical manifestations, right diagnosis in time is challenging. Therefore, imaging characteristics of IMT may be crucial to early diagnosis. Herein, we present a case of an adult woman with primary inflammatory myofibroblastic tumor in the stomach.

2. Case report

A 52-year-old female had developed upper abdominal pain with acid reflux, belching for 2 months. A hard lump with tenderness was palpated under the xiphoid with a clear boundary and little mobility. The size of lump was about 3×4 cm. Initial laboratory work-up revealed no abnormalities. Electron ultrasound gastroscopy (EUS) displayed an elevation of gastric antrum mucosa with suspicion of stromal tumor and non-atrophic gastritis with erosion (Fig. 1). Pathological result confirmed moderate chronic non-atrophic inflammation (gastric antrum) with mild intestinal metaplasia. And the immunohistochemistry of Helicobacter pylori (HP) was negative. However, laboratory tests failed to give us enough clues to confirm a specific diagnosis. The discovery of stomach lesions by EUS warrants performing a routine and contrast-

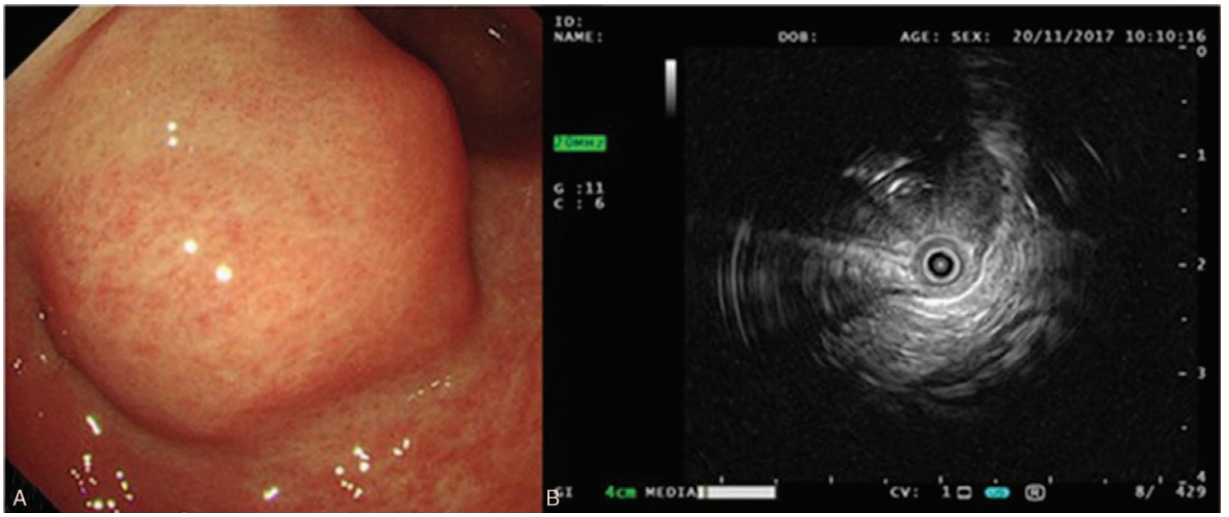

Figure 1. EUS showed elevation of gastric antrum mucosa (A) with an ill-defined heterogeneous hypo-echoic lesion which contains internal echoes (B). There is a heterogeneous rim surrounding the lesions. EUS = electron ultrasound gastroscopy.

enhanced computed tomography (CT) of the chest, abdomen, and pelvis to reveal the distribution of lesions and to identify the possible source (Fig. 2). An exophytic mass with a size of 4.7 × 3.0 cm was detected in the gastric antrum, with an average density of approximately 47 Hounsfield units (HU). Patchy ossification was observed within the lesion. On a contrast-enhanced CT scan, heterogeneous enhancement of the mass was noted, with an average arterial phase density of approximately 81 HU, an average venous phase density of approximately 88 HU and delayed phase density of approximately 106 HU. The lesion had a well-circumscribed margin and a distinct enhancement pattern with clear surrounding fat space. The lesion showed signs prone to be benign, such as no evidence of liver metastases and lymphadenopathy, a well-circumscribed margin and ossification in the mass. Because the nodule is located in the gastric antrum and marked enhanced, we first suspected gastric antrum gastric stromal tumors.

A laparoscopic-assisted distal gastrectomy of the gastric antrum nodule was performed, and an intraoperative frozen section of tumor was analyzed. The tumor was a well-demarcated, exogenous uplift mass of 4.3 cm in diameter. The lining of gastric mucosa was smooth on the surface of the mass. The mass was grayish on the cut surface and hard as bone in gross anatomy. Microscopically, tumor cells were arranged in bundles or braids and the morphology of cells was fusiform (Fig. 3). The cytoplasm was abundant with rod-shaped nucleus mitoses about 0-1/10HPF. In the meantime, a large amount of mature bone-like tissue was seen, which was unevenly distributed. In some areas, the intercellular collagenization was obvious, and no obvious necrosis was seen. Immunohistochemistry showed the results as follows: CD117 (-); CD34 (-); Dog-1 (-); CK (-); S100 (-); EMA (-); Des (-); Ki-67 (3%); ALK (+). Likewise, special staining showed: VG (red), Masson (blue). Finally, a histological diagnosis of gastric IMT without characteristics of malignancy was made according to the above-described morphological and immunohistochemical features. After 6 months of follow-up, the patient is still alive without evidence of metastasis or recurrence.

The patient agreed to authorize us to share the figures and the experiences during his treatment procedure in our department. Informed consent was obtained. The project reviewed by Medical Research Ethics Committee of the First Affiliated Hospital of Nanchang University and met ethical requirements.

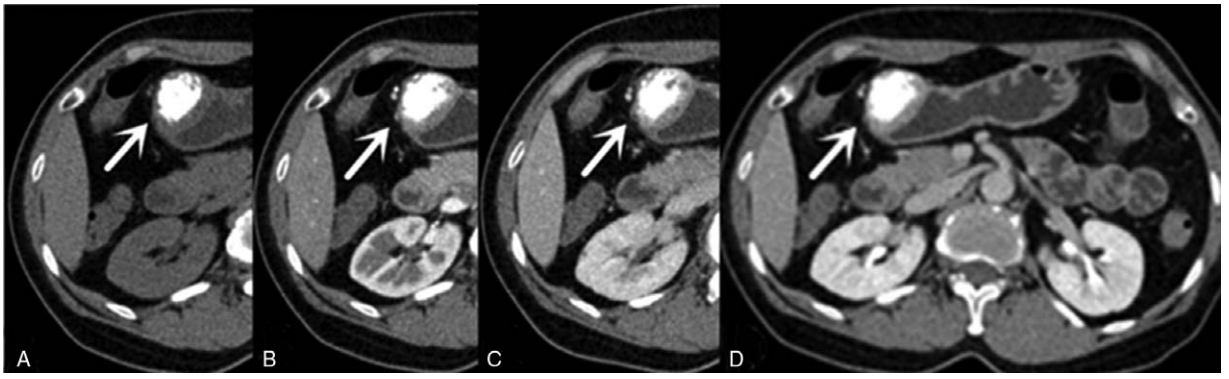

Figure 2. Abdominal CT scan showed a 4.7 × 3.0 cm exogenous raised soft tissue mass in the gastric antrum with a well-circumscribed margin (marked with arrows) and a peripheral enhancement pattern (B-D),and a massive patch of high-density was seen inside (A). CT = computed tomography.

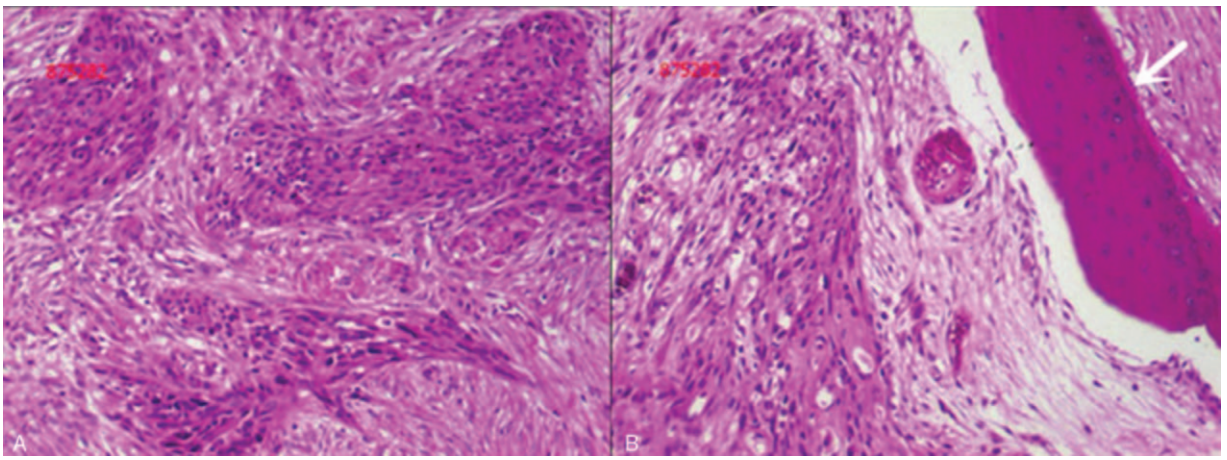

Figure 3. Pathological examination revealed that the tumor cells were arranged in bundles or braids and the morphology of cells was fusiform (A: hematoxylin and eosin, 100×),and a large amount of mature bone-like tissue was seen inside,marked with arrows (B: hematoxylin and eosin, 200×).

3. Discussion

Primary gastric IMT is a very rare disease, which is characterized by fibroblasts and spindle cell fibroblasts, and often accompanied with inflammatory infiltrates of a large number of plasma cells, lymphocytes, and eosinophils.[4] Most reported cases of primary gastric IMT are benign in nature. However, Jun Fan[5] reported the first gastric IMT case with metastasis in an adult. There has been controversy over whether IMT is an inflammation or tumor, benign or malignant, and its etiology and pathogenesis are still unknown. It has been reported that some cases of IMT might be associated with genetic mutation, Epstein–Barr virus (EBV) infection or immune dysregulation.[5–8]

Primary gastri-IMTs do not cause specific clinical symptoms, or abnormal in laboratory examinations. As a result, this type of tumors is prone to delayed diagnosis or misdiagnosis based on clinical and radiological studies. Jadhav et al [9] summarized the data of 34 gastric IMT cases reported in the English literature as follows: IMT was frequently reported in the body of the stomach and presented as an asymptomatic soft tissue tumors on CT, with an average of 6.9 cm.A few cases only gastric wall thickening. However, involvement of lesser curvature with exophytic mass formation was rare.[2,10] Previously reported primary gastri-IMTs showed various enhancement patterns on contrast-enhanced CT images, such as peripheral enhancement,[9,11] irregular enhancement.[12] IMT is divided into 3 basic histologic patterns:

1. myxoid, vascular, and inflammatory areas resembling nodular fasciitis;
2. compact spindle cells with intermingled inflammatory cells (lymphocytes, plasma cells, and eosinophils) resembling fibrous histiocytoma; and
3. dense plate-like collagen resembling a desmoid or scar.[13]

The third type IMT may locally have calcification or ossification. This lump pathology is consistent with the third type IMT and found ossification, which was not discussed in previous related literature. This might be a unique finding for this disease.

Because the histology of the gastric tumor remains unclear, and the patient has good general condition and no metastasis to the surrounding organs, it is necessary for patients to receive prompt early surgical resection. And Surgical resection is the preferred treatment option for IMT,[14] including the following surgical approaches: a distal gastrectomy with sufficient proximal and distal margins and Roux-en-Y reconstruction.

Pathology and immunohistochemistry analysis is the key to the diagnosis of primary gastric myofibroblastic tumors, especially ALK, which can be used as an identifying factors for IMT and gastrointestinal stromal tumors (GISTS), smooth muscle neoplasm, inflammatory fibrous polyps, solitary fibrous tumor, peripheral nerve sheath tumors, fibromatosis, and rarely follicular dendritic cell sarcoma. ALK immunohistochemistry is a useful diagnostic adjunct in the appropriate circumstances, especially positive for ALK rearrangement but focally positive for CD34 and negative for CD21, CD117, CD23, and, S100, which supports the clear pathological diagnosis of IMT and excludes all other possibilities. Although ALK-positive contributes to the diagnosis of IMT, it is only seen in 56% of the cases.[9] It was recently found that ALK reactivity was related to local recurrence but not distant metastasis,[3] which was limited to ALK-negative lesions, suggesting that ALK reactivity may be a favorable prognostic indicator in IMT.[3,15] Our ALK-positive cases showed lymph node metastasis but no distant metastasis, which is consistent with the above literature reports. This case was followed up for 6 months without recurrence and metastasis. However, long-term follow-up is still needed.

In conclusion, for primary gastric tumors with considerably delayed enhancement on contrast-enhanced CT, and particularly accompanied by ossification, the possibility of IMT should be considered.

Author contributions

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Writing – original draft: Bingxue Cheng, Chen Yang.
Writing – review & editing: Li Zhou.

References

[1] Shi H, Wei L, Sun L, et al. Primary gastric inflammatory myofibroblastic tumor: a clinicopathologic and immunohistochemical study of 5 cases. Pathol Res Pract 2010;206:287–91.
[2] Katakwar A, Gedam BS, Mukewar S, et al. Primary gastric inflammatory myofibroblastic tumor in an adult-case report with brief review. Indian J Surg Oncol 2014;5:66–70.

[3] Coffin CM, Hornick JL, Fletcher CD. Inflammatory myofibroblastic tumor: comparison of clinicopathologic, histologic, and immunohistochemical features including ALK expression in atypical and aggressive cases. Am J Surg Pathol 2007;31:509–20.
[4] Coffin CM, Fletcher JA. CDM F, Unni KK, Mertens F. Inflammatory myofibroblastic tumour. World Health Organization classification of tumours. Pathology and genetics of tumours of soft tissue and bone IARC Press, Lyon:2002;91–3.
[5] Fan J, Huang B, Yang X, et al. ALK-positive gastric inflammatory myofibroblastic tumor in an adult with familial adenomatous polyposis and diffuse fundic polyposis. Diagn Pathol 2017;12:68–73.
[6] Khatri A, Agrawal A, Sikachi RR, et al. Inflammatory myofibroblastic tumor of the lung. Adv Respir Med 2018;86:27–35.
[7] Kiryu S, Takeuchi K, Shibahara J, et al. Epstein-Barr virus-positive inflammatory pseudotumour and inflammatory pseudotumour-like follicular dendritic cell tumour. Br J Radiol 2009;82:e67–71.
[8] Vroobel K, Judson I, Dainton M, et al. ALK-positive inflammatory myofibroblastic tumor harboring ALK gene rearrangement, occurring after allogeneic stem cell transplant in an adult male. Pathol Res Pract 2016;212:743–6.
[9] Jadhav M, Harvi R, Patil R, et al. Inflammatory yofibroblastic tumor of the stomach presenting as an exophytic mass—a diagnostic dilemma. Turk Patoloji Derg 2017.
[10] Cho MY, Min YK, Kim NR, et al. Fever of unknown origin as a presentation of gastric inflammatory myofibroblastic tumor in a two year old boy. J Korean Med Sci 2002;17:699–703.
[11] Park SH, Kim JH, Min BW, et al. Exophytic inflammatory myofibroblastic tumor of the stomach in an adult woman: a rare cause of hemoperitoneum. World J Gastroenterol 2008;14:136–9.
[12] Chong A, Ha JM, Hong R, et al. Inflammatory myofibroblastic tumor mimicking gastric gastrointestinal stromal tumor on 18F-FDG PET/CT. Clin Nucl Med 2014;39:725–7.
[13] Coffin CM, Watterson J, Priest JR, et al. Extrapulmonary inflammatory myofibroblastic tumor (inflammatory pseudotumor) A clinicopathologic and immunohistochemical study of 84 cases. Am J Surg Pathol 1995;19:859–72.
[14] Albayrak F, Dursun H, Albayrak Y, et al. Inflammatory myofibroblastic tumor of the stomach in an adult woman: a rare intermittent cause of gastric outlet obstruction. Tumori 2010;96:492–5.
[15] Surabhi VR, Chua S, Patel RP, et al. Inflammatory Myofibroblastic tumors: current update. Radiol Clin North Am 2016;54:553–63.