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CASE REPORT

Primary Gastric Inflammatory Myofibroblastic Tumor in an Adult—Case Report With Brief Review

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Abstract The term inflammatory myofibroblastic tumor more commonly referred to as "pseudostumor ", denotes a pseudosarcomatous inflammatory lesion that contains spindle cells, myofibroblasts, plasma cells, lymphocytes and histiocytes. It exhibits a variable biological behavior that ranges from frequently benign lesions to more aggressive variants. Inflammatory myofibroblastic tumor (IMT) of the stomach is extremely rare and its prognosis is unpredictable. We present a 45-year-old diabetic man with a gastric Inflammatory myofibroblastic tumor. The histopathological and immunohistochemical analysis was the key to reach diagnosis.

Keywords Stomach · Inflammatory myofibroblastic tumor · Inflammatory pseudotumor

Introduction

Inflammatory myofibroblastic tumor (IMT) previously known as inflammatory pseudotumor, plasma cell granuloma, inflammatory myofibroblastoma, and inflammatory myofibrohistiocytic proliferation [1]. This distinctive neoplasm is composed of myofibroblastic cells associated to an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils that relapse often and rarely metastasizes [2]. The pathogenesis of IMT remains unclear, although various allergic, immunologic, and infectious mechanisms have been postulated. Clinically, the majority of IMTs are benign but they require adequate surgical treatment because it has a tendency for local recurrence [3–5]. IMT rarely occurs in adult persons, particularly in the stomach, previously described cases of primary gastric IMTs were in the form of case reports or small series [6] [Table 1]. We present a case of a primary gastric IMT in an adult male with recurrence within 1 month of adequate surgery.

Case report

A 45 year old diabetic man presented with weight loss and epigastric pain of 1 year duration with aggravation in symptoms since 3 months compelling him to seek medical attention. He denied other gastrointestinal symptoms such as nausea, vomiting, abnormal bowel habits, melena or haematemesis and had insignificant past medical or familial history. Physical examination showed cutaneous pallor and mild abdominal tenderness in the epigastrium but no palpable mass. CT scan done outside revealed exophytic mass at pylorus (5.7×4.7 cm), Posteriorly abutting normal looking pancreas with maintained fat plains favoring gastrointestinal stromal tumor (GIST) (Fig. 1). The laboratory findings were normal, except for a normocytic, normochromic anemia (hemoglobin: 9.7 g/dL). Gastroscopy revealed submucosal, broad bulge, located in the posterior wall of lower gastric body and first part of duodenum with normal looking overlying mucosa such as a gastrointestinal stromal tumor (GIST) (Fig. 2). Endoscopic ultrasound (EUS) showed an oval

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Table 1 Summarized clinico-pathological characteristics of previously reported primary gastric IMT

Author	Sex/Age	Presenting symptoms	Tumor localization in the stomach	Tumor size (incm)	Mitosis (10 HPFs)	Histologic pattern	Treatment	Follow-up
Bjelovic et al. [1]	W/43	AP, pyrosis, nausea	Distal stomach, below AI	6 cm	1–2	Hypercellular spindle cell proliferation with vague fascicular areas	DG	2 years
Albaryak et al.[3]	W/56	UGH, nausea, vomiting	C extending towards P	11 cm	1–2	Granulation-type and storiform spindle cell proliferation	PG	NED, 8 months
Shi et al. [4]	M/36	AP, AM	Antrum, LC	4.5 cm	1–2	Myxoid hypocellular with some fascicular areas	PG	NED, 5 years
Shi et al. [4]	M/42	AP, UGH, AM	Upper body, GC	8.0 cm	1–2, focally up to 5	Fascicular with some myxoid areas	PG	Recurrence at 12 months after the first surgery, now NED at 2 years (11 month after the second surgery)
Shi et al. [4]	M/40	AM	Upper body, AW	6.3 cm	1–2	Myxoid hypocellular with some fascicular areas	PG	NED 3.3 years
Shi et al. [4]	M/45	AP, AM	Angle	5.5 cm	1–2	Myxoid hypocellular with some fascicular areas	PG	NED, 2.6 years
Shi et al. [4]	W/40	AP, AM	Lower body, PW	5.8 cm	1–2	Fascicular with some myxoid and sclerotic areas	PG	NED, 4 years
Leon et al.[6]	W/50	Vomiting, early satiety, weight loss	PW	7 cm	1–2	Patternless round and spindle cell proliferation	PG	NED,2 years
Park et al.[8]	W/55	AP, hematoperitoneum	Upper body, AW near GC	8 cm	1–2	Vague fascicular proliferation	Gastric wedge resection	/
Katakwar et al.	M/45	AP	AW near LC	5 cm	1–2	Hypocellular, collagenized, myofibroblatic cells	DG	Recurrence at 1 month after the first surgery.

AP abdominal pain; *AM* abdominal mass; *UGH* upper gastrointestinal hemorrhage, *LC* lesser curvature of the stomach; *GC* greater curvature of the stomach; *AW* anterior wall of the stomach; *PW* posterior wall of the stomach; *C* cardia; *AI* angular incisure of the stomach; *PG* partial gastrectomy; *DG* distal gastrectomy; *NED* no evidence of disease

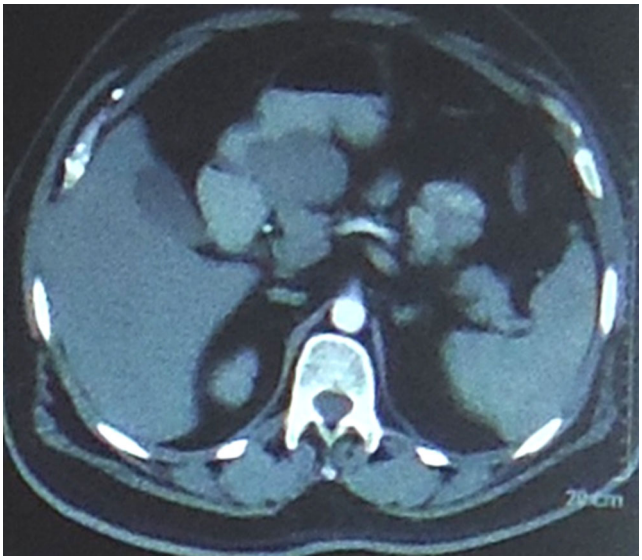

Fig. 1 CT scan showing exophytic mass in pylorus

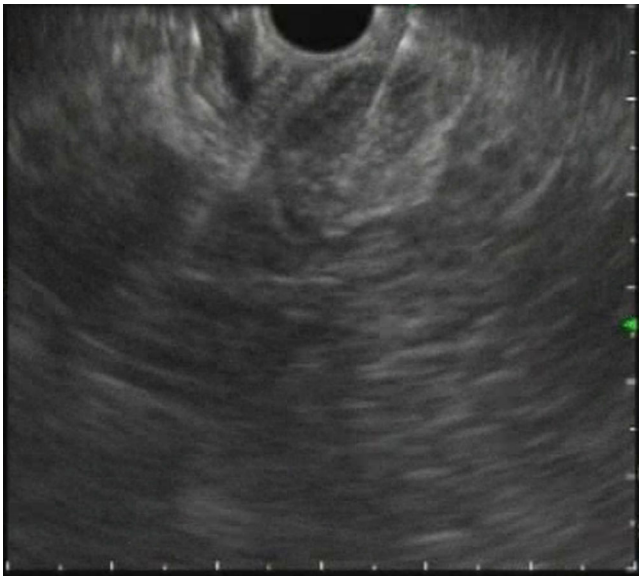

Fig. 3 Endoscopic ultrasound showing mass from lamina propria

Heterogeneous mass 5×5 cm in diameter, arising from the muscularis propria layer (Fig. 3), FNAC was inconclusive. Due to gastric tumor with unclear tumor histology decision for surgical exploration taken, which revealed well encapsulated tumor in the pyloric region close to lesser curvature involving the entire thickness of the gastric wall. There were no signs of loco-regional infiltration or metastatic disease. Distal gastrectomy with Roux-en-Y reconstruction was performed and Nasojejunal tube put across gastrojejunostomy for feeding. Early postoperative course was uneventful and patient was discharged on tenth day following the surgery. Histologically, the tumor was composed of Spindle cell lesions and Area of hyalinization, with no evidence of atypical mitosis suggesting GIST, immunohistochemistry not done due to finances. Twenty days after discharge from hospital patient had Vomiting, Abdominal fullness, Haematemesis and Obstipation. He was admitted and Approximately 700 ml gastric juice aspirated through ryles tube. X ray abdomen and USG were normal.

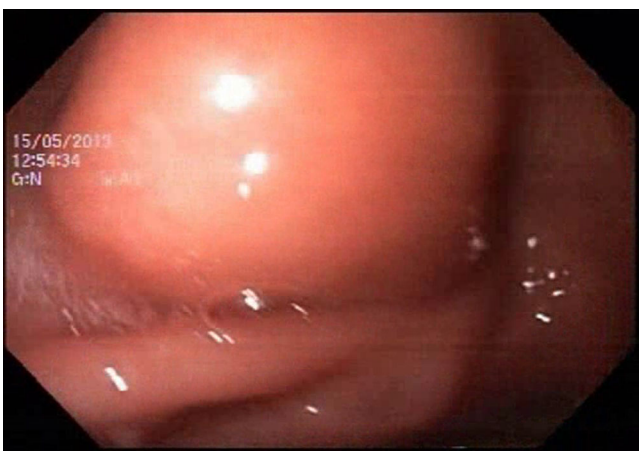

Fig. 2 Gastroscopy showing submucosal bulge

Blood investigation revealed normal electrolytes with mild leucocytosis (11800) and slightly raised serum creatinine (1.9 mg/dl). Gastroscopy revealed stomal ulcer with mucosal edema causing gastric outlet obstruction . Abdominal computed tomography (CT) demonstrated Soft tissue mass at gastrojejunostomy site suggesting recurrence with Nodal involvement. Decision of re-exploration taken in view of gastric outlet obstruction, upon surgery there was dense desmoplastic and fibroblastic reaction at anastomotic site with Omental thickening suggesting exaggerated inflammatory response. As any further dissection at previous gastrojejunostomy would have caused more catastrophes we decided to go for bypass by doing new gastrojejunostomy proximal to previous Jejunojeunostomy with an NJ tube across. Second opinion and immunohistochemistry on initial

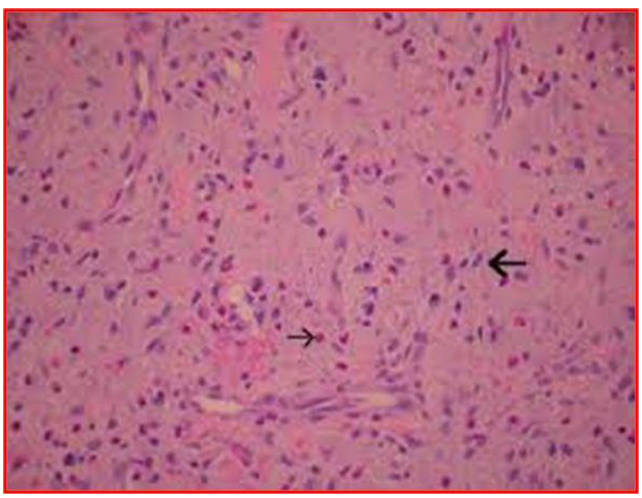

Fig. 4 Histopathology showing Hypocellular, collagenized, myofibroblatic cells

Table 2 Immunohistochemistry review in IMT y

Cytokeratin	SMA	Desmin	Calponin	Caldesmon	MyoD1	S-100	ALK
+ve in 20–40 %	+ve in >90 %	+ve in 70 %	Usually +ve	Usually +ve	-ve	-ve	Usually +ve

SMA smooth muscle actin; *MyoD1* myogenic differentiation 1; *ALK* anaplastic lymphoma kinase

gastrectomy specimen was reported as Hypocellular, collagenized, myofibroblatic cells, Positive for SMA (smooth muscle actin), ALK (anaplastic lymphoma kinase) & beta-catenin. Negative for Desmin, S-100 protein and c-kit Suggesting inflammatory myofibroblastic tumor than desmoid fibromatosis but not GIST (Fig. 4). Patient had fast recovery and Discharged on 8th postoperative day and has been followed up without any further complications.

Discussion

There has been debate as to whether inflammatory myofibroblastic tumor (IMT) is a tumor or inflammation, and also whether is benign or malignant. It is locally recurrent, however it rarely metastasizes [7]. The primary inflammatory myofibroblastic tumor (IMT) is a very rare neoplasm in adults and the exact nature of the disease is not yet completely understood [8]. The first abdominal localization was described in liver by Pack and Baker in 1953 [9]. It was once accepted that IMT is primarily a disease of children and young adults and commonly occurs in the lungs [10]. However, recently the authors verified that the IMT can occur in any organ of the body and in all ages. Cytogenetic abnormalities such as rearrangements of the ALK gene on chromosome 2p23, clonal chromosome abnormalities, DNA aneuploidy, and the role of oncogenic viruses in the pathogenesis of IMT suggest that it is a real neoplasm [7, 11, 12]. Even with a thorough diagnostic workup, which included CT, US, EUS, laboratory analyses, upper flexible endoscopy with FNAC in this case, it was difficult to make an accurate preoperative tumor diagnosis. In most cases IMT features mimic malignancy on upper flexible endoscopy and radiological imaging. For that reason, most IMT cases require surgical exploration and complete resection to obtain an accurate microscopic diagnosis [7]. Due to a good general condition and no signs of metastatic disease, a surgical procedure performed earlier includes a distal gastrectomy with sufficient proximal and distal margins and Roux-en-Y reconstruction. A complete surgical resection remains the only proven mode of cure, and is proposed as the first line of treatment in all IMT cases [3]. Histopathologically IMT is characterized by a myofibroblastic proliferation spindled and/or epithelioid, a lymphoplasmacytic infiltrate distributed among the tumor cells and a myxoid background stroma. Three architectural patterns have been described in IMT: myxoid hypocellular pattern, a cellular fascicular or nested pattern with variable amounts of myxoid stroma, and a sclerotic, hyalinized pattern with minimal myxoid stroma [7]. As far as the differential diagnosis is concerned there are few tumors or lesions in the stomach that must be distinguished from IMT, which include gastrointestinal stromal tumor (GIST), inflammatory fibroid polyp, smooth muscle neoplasm, peripheral nerve sheath tumor, solitary fibrous tumor, fibromatosis and rarely follicular dendritic cell sarcoma [8]. GIST may show cyst formation, hemorrhage, necrosis, occasionally seen in some IMTs. GIST typically does not have the inflammatory background as seen in the IMT [13]. In addition, some GIST cells have cytoplasmic vacuoles, a feature not seen in the IMT. Immunohistochemically GIST is typically positive for CD117 but negative for ALK, whereas IMT shows an opposite profile as seen in our patient [Table 2]. Recently ALK reactivity was found to be associated with local recurrence, but not distant metastasis, which was confined to ALK-negative lesions, suggesting that reactivity may be a favorable prognostic indicator in IMT's [7]. However, other studies did not confirm such an association. Tumor recurrence within a year of a surgery was observed in 15 % to 37 % cases of primary gastric IMT [4, 10]. But none of previous report suggests recurrence within 1 month of surgery. In every primary gastric IMT case a long- term clinical, radiological and laboratory follow-up is indicated.

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