MEDVIX PUBLICATIONS

Journal of Medical and Clinical Case Reports
Case Report | Open Access | ISSN 2997-6022 | Volume 01 | Issue 01

Calcifying fibrous tumor of the stomach – a case report

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Received date: 21 January 2024; **Accepted date:** 05 February 2024; **Published date:** 08 February 2024

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Citation: Firdos Saba, Rzan Alsayegh, Tahira Hamid, Shafaq Mujtaba, Deeba Mushtaq. Calcifying fibrous tumor of the stomach – a case report. Journal of Medical and Clinical Case Reports 1(1). https://doi.org/10.61615/JMCCR/2024/FEB027140208

Abstract

A calcifying fibrous tumor (CFT), also called calcifying fibrous pseudotumor, is a rare benign lesion that occurs in the deep soft tissues of the limbs and trunk [1]. Gastric CFT is very rare [2], with less than 50 cases reported in the literature [3]. It is most often asymptomatic and incidentally detected by endoscopy, and it can be cured by local resection with a low chance of recurrence [1].

Here, we report a case of gastric CFT Incidentally detected after the patient underwent sleeve gastrectomy.

Keywords: calcifying fibrous tumor, stomach.

Introduction

A calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor affecting children and young adults [4] and shows a predilection for deep soft tissues of the extremities and the abdominal cavity [2,4]. CFT of the tubular gastrointestinal tract is very rare [5].

CFTs are histologically characterized by fibroblastic spindle cell proliferation [6] within a hypocellular densely hyalinized stroma [6,7], psammomatous or dystrophic calcifications, and lymphoplasmacytic infiltrates [2].

Case Presentation

A 24-year-old male patient with morbid obesity, weighing 143 kg and a BMI of 48, came to the Bariatric surgery department for sleeve gastrectomy. He had no obstructive sleep apnea or GERD. Complete blood counts were within normal limits. Thyroid function tests were also normal. Liver enzymes were near normal. Lipid profile showed reduced HDL (36mg/dl) and increased non-HDL (137 mg/dl). The patient underwent laparoscopic sleeve gastrectomy.

The specimen consisted of a closed tubular segment of the stomach measuring 17x3 cm. External surface smooth gray tan, on cut section submucosal nodule, was seen measuring 1.0x0.6 cm. The cut section of the nodule was gray-white, which lies 2.5 cm from the stapled margin. Microscopy showed sections from the gray-white submucosal nodule, a tumor composed of sparse spindle cells embedded in dense hyalinized collagenous stroma with areas of chronic inflammation and psammomatous calcification. The tumor cells were wispy with tapering ends in parallel to the collagen bundles. The inflammatory cells were composed of a mixed population of lymphocytes, plasma cells, and a few eosinophils. At places, few lymphoid follicles were noted. The tumor was located in the submucosa, with intact, unremarkable surface epithelium. No evidence of gastritis, H. pylori-like organisms, or malignancy was seen.

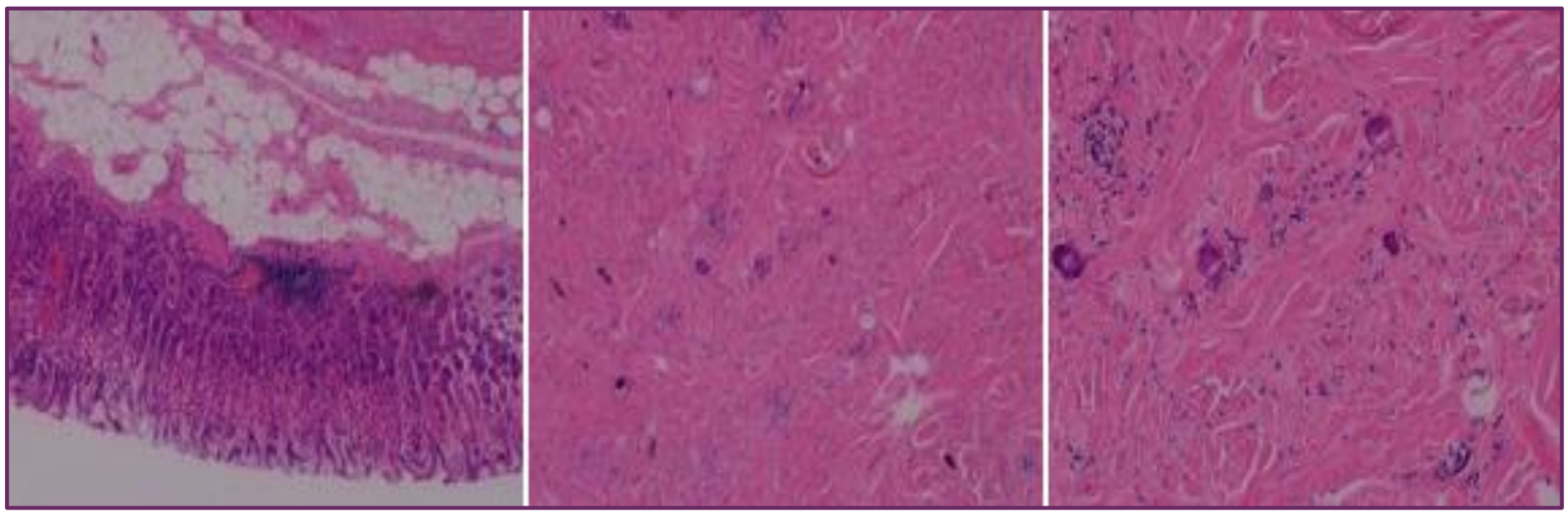

Fig.1 Histologic findings of submucosal tumor. **(A)** Gastric mucosa. **(B)** Tumors are composed of sparse spindle cells embedded in a dense hyalinized collagenous stroma. **(C)** Areas of chronic inflammation and psammomatous calcification.

Properly controlled immunohistochemical stains showed the thin spindle cells embedded in the hyalinized stroma to be positive for Vimentin and CD34 (patchy, weak) while negative for CD117, DOG1, S100, EMA, PR, Desmin, SMA, and MSA. A diagnosis of Calcifying fibrous tumor was offered. Calcifying Fibrous Tumor is a benign lesion, so pathological staging was not done. Lymph nodes were not commented on, as they are benign lesions and do not metastasize. Cancer biomarkers were not needed for this case as the lesion was benign and incidentally detected. Differential diagnosis considered was Gastrointestinal stromal tumor (GIST), which was ruled out by negative CD117 & DOG 1 IHC. Leiomyoma was ruled out by negative Desmin IHC.

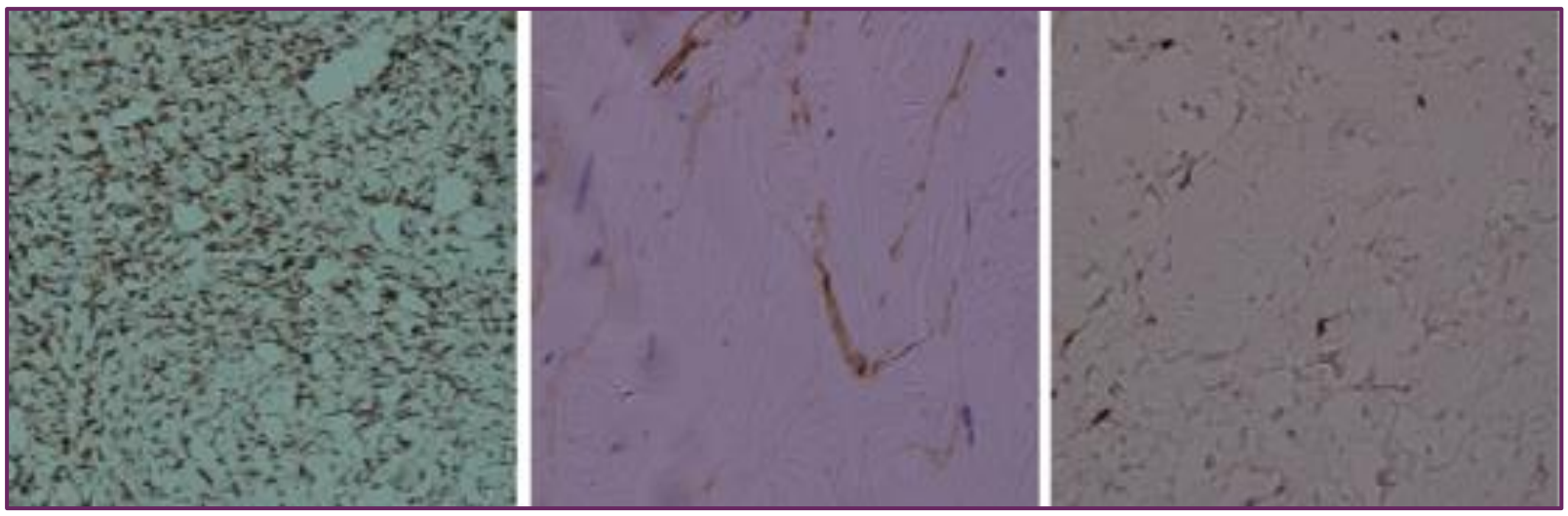

Fig.2 Immunohistochemical analysis of submucosal tumor. **(A)** Vimentin positive. **(B)** CD34 positive. **(C)** CD117 negative.

Discussion

A calcifying fibrous tumor (CFT) was first described by Rosenthal et al. in 1988 as a childhood soft tissue tumor with psammoma bodies [3]. It was previously named a calcifying fibrous pseudotumor [1] and was thought to represent a reactive process resulting from abnormal tissue healing [5].

Reported cases of gastric CFTs have been documented with a mean age of 53 years, no sexual predominance, and involving the body of the stomach and the posterior wall [5]. There are 48 cases of gastric CFT reported in the literature [3], including gastric CFT presenting as an ulcer [5].

CFTs have been documented in various anatomical sites, including the mesentery, peritoneum, mediastinum, pleura, lung, adrenal glands, paratesticular, and spermatic cord. Although CFTs have been reported in various organ systems, the gastrointestinal tract is rarely involved [7,8].

The pathogenesis of CFTs remains unclear, but it has been hypothesized that CFT may represent a late sclerosing stage of inflammatory myofibroblastic tumor (IMT) [2]. Evidence has suggested that these lesions may be associated with IgG4-related diseases [6].

Gastric CFTs are most often detected incidentally and present without symptoms or with nonspecific symptoms such as abdominal pain, dyspepsia, flatulence, nausea, vomiting, altered bowel habits, loss of appetite, weight loss, and fatigue. Gastric CFTs may occasionally be associated with more severe manifestations, including gastric ulcers and bleeding per rectum [6].

Imaging findings of CFT show coarse calcification on conventional ultrasound and peripheral hypoenhancement without central enhancement on ultrasound with contrast. A CT scan shows a mildly enhanced, well-circumscribed homogenous mass with calcification [6]. On MRI, the lesion appears as an isointense signal mass on gadolinium-enhanced T1-weighted imaging and hypointense on T2-weighted imaging [9].

The differential diagnosis of gastrointestinal CFT includes gastrointestinal stromal tumor (GIST), schwannoma, leiomyoma, solitary fibrous tumor, inflammatory myofibroblastic tumor (IMT), plexiform fibromyxoma, fibromatosis, and reactive nodular fibrous pseudotumor (RNFP) [6]. CFTs are difficult to distinguish from GIST [1] as they are frequently hyalinized with dystrophic calcifications. Lymphoplasmacytic infiltrate and psammomatous calcifications are not features of GIST [4].

Entity	Similar histological features to CFT	Differentiating histological features from CFT	Positive immunostaining	Negative immunostaining
Calcifying fibrous tumor (CFT)	Spindle cell proliferation Densely hyalinized stroma Scattered calcifications Lymphoplasmacytic infiltrate		CD34, vimentin, factor XIIIa	SMA, ALK, desmin, S100, cytokeratin, DOG1, c-kit
Gastrointestinal stromal tumor (GIST)	Uniform, bland spindle cells in short fascicles	Collagen deposition with low cellularity Calcifications	CD34, c-kit, DOG1	

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Schwannoma	Spindle cells with tapered ends in micro trabecular pattern Peripheral lymphoid cuff Scattered atypical cells		S100	
Leiomyoma	Spindle cells with elongated nuclei and abundant cytoplasm	Hyalinized stroma Rare mitosis and atypia	SMA, desmin	CD34
Solitary fibrous tumor (SFT)	Staghorn vessles Perivascular hyalinization	Spindle cells Thick bands of hyalinized collagen	CD34, CD99, BCL-2, EMA	
Inflammatory myofibroblastic tumor (IMT)	Myofibroblastic spindle cells Abundant blood vessles Rarely calcification	Lymphoplasmacytic infiltrate	SMA, ALK	
Plexiform fibromyxoma	Multinodular growth pattern Myxoid stroma Small vessles	Hypocellular spindle cell proliferation	SMA	CD34, DOG1, S100, c-kit
Desmoid tumor (neurofibromatosis lesions)	Sweeping fascicles of bland spindle cells with infiltrative growth pattern		Nuclear b-catenin	
Reactive nodular fibrous pseudotumor (RNFP)	Paucicellular fibroblast proliferation	Hyalinized collagenous stroma Lymphocytic inflammatory infiltrate	Vimentin, SMA, desmin	CD34

Table 1: differential diagnosis of gastrointestinal CFT [6].

Recently, gastric CFT has been thought to represent a GI lesion of IgG4-related disease, which is characterized by the presence of diffused or localized masses in single or multiple organs with the elevation of serum IgG4 levels above 135mg/dL, in addition to histological findings of fibrosis and abundant infiltration of IgG4-positive plasma cells and lymphocytes [1]. Cases of CFT with positive staining plasma cells for IgG and IgG4 have been reported [6]. However, the diagnosis of IgG4-RD was not confirmed. It is important to suspect IgG4-RD in patients with gastrointestinal CFT and monitor serum IgG4 levels [1], as CFT may represent a stage of IgG4-associated disorder, and steroid therapy should be considered in clinical management [3].

CFT has also been associated with Castleman Disease, a benign lymphoproliferative disorder that occurs in lymph nodes. In some cases, both entities have been found within the same lymph node or adjacent lymph nodes, raising the possibility of representing different stages of the same reactive process. An additional association of CFT has been made with sclerosing angiomatoid nodular transformation (SANT), a non-neoplastic vascular proliferation of the spleen. Rare cases of SANT co-existing with CFT suggest a possible common etiology. CFT has also been reported to develop in patients after sustaining trauma in the same anatomic site, supporting the theory that CFT may be reactive proliferation resulting from trauma.[6]

Surgical or endoscopic local resection is the main mode of treatment for gastric CFT [5]. The recurrence rate has been estimated to be 10%, and follow-up after excision is recommended. Some authors have suggested that long-term follow-up is sufficient management for patients with small asymptomatic CFTs [6].

Gastric CFT is a benign lesion with a rare tendency for recurrence [5,10]. It shows excellent prognosis [10] and is unlikely to cause serious complications [3]. There are no reported cases of malignant transformation or metastasis of CFT. [6]

Conclusion

Gastric CFT is under-recognized by pathologists due to its low incidence and small size [2]. It is important to distinguish gastric CFT morphology from other stromal lesions in order to determine prognosis and management [6]. The most important entities to exclude in diagnosis are GIST and IMT, as they have worse prognoses and more aggressive progression.[6]

Summary

A calcifying fibrous tumor (CFT) is a rare benign fibrous lesion [4] most often incidentally detected and cured by local resection [6]. CFT of the stomach is very rare [4]. In this article, we presented a case of a gastric calcifying fibrous tumor incidentally detected in a 24-year-old morbidly obese patient who underwent laparoscopic sleeve gastrectomy. This lesion must be differentiated from other spindle cell tumors, particularly gastrointestinal stromal tumors (GIST) of the stomach [5]. Given the rarity of gastric CFT, it is particularly important to enhance clinicians' understanding of this lesion for prognostic and therapeutic purposes [3,5].

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