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Case Report

Endoscopic resection of gastric calcifying fibrous tumor: An incidental finding

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ABSTRACT

Calcifying fibrous tumor (CFT) is benign mesenchymal lesion. CFTs of the GI tract are quite rare with less than 60 cases of gastric CFTs reported in the English literature. In a healthy 49 old female, routine Endoscopic ultrasonography (EUS) revealed an internally isoechoic, homogeneous 12mm lesion mainly within the submucosa in the distal body of stomach with no significant past history. An endoscopic submucosal dissection (ESD) was done to rule out malignancy. The histopathological examination showed a hypocellular lesion with few spindle cells interspersed in a hyalinized, collagenous matrix with occasional lymphoplasmacytic cells. Foci of psammomatous, dystrophic calcification were noted. Immunohistochemical examination revealed tumor cells positive for vimentin and negative for CD34, CD117, DOG1, desmin, SMA and S100. Based on histopathological and IHC findings, a diagnosis of gastric calcifying fibrous tumour was made. Only a handful of CFT cases resected by endoscopic submucosal dissection has been reported in the literature. So herein we report a rare case of endoscopically resected gastric calcifying fibrous tumor.

1. Introduction

Calcifying fibrous tumor is a unique entity, which is described by World Health Organization as benign mesenchymal tumor characterized by hypocellular lesion composed of spindle cells, few lymphocytes against a hyalinized collagenous matrix. CFTs originated from the soft tissue like pleura & abdominal viscera has been reported (2,3), but CFT's originated from the stomach are rare, which are often detected incidentally during endoscopic examination. Although endoscopic resection is more suitable for CFTs, only a few cases of CFT's have been reported. So, we present a case report of incidentally detected, endoscopically resected gastric CFT in a 49-year-old female with complete resection of tumor.

2. Case Presentation

A 49-year-old female presented with a complaint of pain abdomen. Physical examination and laboratory tests including CBP, LFT, RFT were within normal limits. She had no significant past medical history. Contrast enhanced computed tomography (CECT) abdomen showed no significant abnormality. Upper GI endoscopy revealed a subepithelial lesion in the distal body of stomach, which is followed by endoscopic submucosal dissection of the lesion. Grossly we found 1.2 × 1 × 0.7 cm, firm, grey-white nodular lesion. Histological examination revealed a circumscribed hypocellular lesion composed of hyalinised collagenous tissue with few scattered spindle cells and lymphoplasmacytic infiltrate. The spindle cell exhibit elongated nuclei with fine chromatin and eosinophilic cytoplasm. Few areas of dystrophic calcification and occasional foci of psammomatous calcification were identified. No significant nuclear atypia or increased mitotic

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activity or any other features of malignancy noted. (Figure 1 A-E) The immunohistochemical examination revealed the tumor cells positive for Vimentin (Figure 2) and negative for Desmin, S100, ALK, CD34, CD117 and DOG-1. Based on our histopathological & immunohistochemistry findings, lesion was diagnosed as gastric calcifying fibrous tumour.

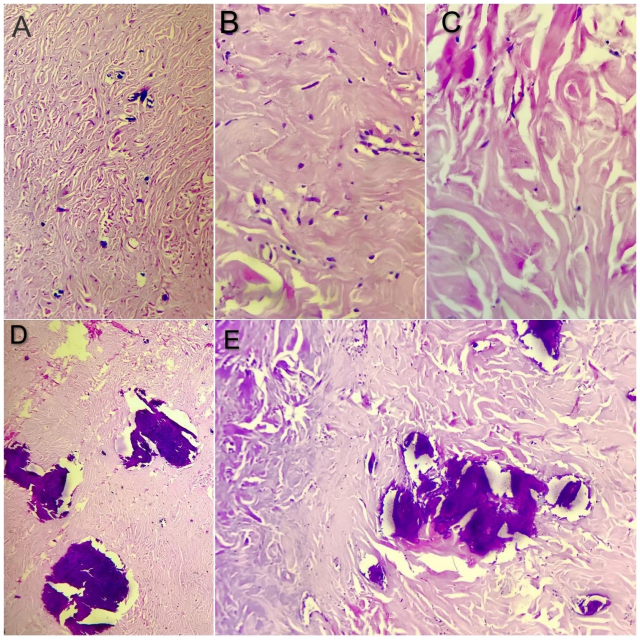

Fig. 1: A-C): (H &E 10X, 40X) shows dense hyalinised collagenous tissue along with few scattered spindle cells; **D-E**): (H&E 10X, 40X) Areas of dystrophic calcification

3. Discussion

Calcifying fibrous tumor is a mesenchymal lesion, which was named by Rosenthal and Abdul-Karim in 1988 as 'childhood fibrous tumor with psammoma bodies',[1] but later few studies confirmed that it can occur in adults as well. Around 75% of the CFTs originates in the peritoneum and abdominal cavity, and less than 60 cases of gastric CFTs has been reported in literature till date.[2]

A recent study reported that CFTs can arise in the different parts of gastrointestinal tract like small bowel, stomach, esophagus, large intestine and appendix and shows female predominance.[3] As previously it was assumed to be rare in the GI tract, current data with improved clinical recognition of this entity has led to the conclusion that the majority of these lesions may originate in the gastrointestinal tract.[4,5]

The pathogenesis of CFT confined to gastric wall remain elusive, few studies showed that these may be pathophysiologically associated with inflammatory lesions such as IgG4-RD, hyaline vascular type Castleman disease, IMT etc.[4]

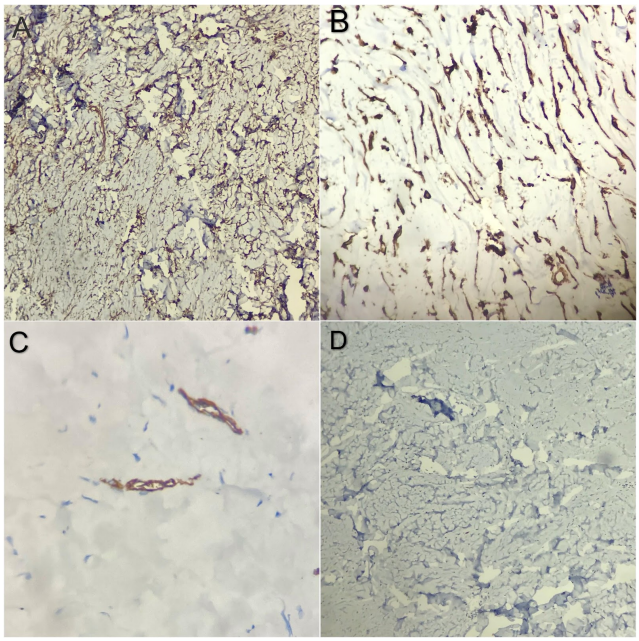

Fig. 2: A,B): Photomicrograph of Vimentin immunostain (100X, 400X) shows cytoplasmic staining in spindle cells; **C**): CD34 (400X) highlights blood vessels and shows spindle cells negativity; **D):** Negative immunostaining for Desmin, SMA, DOG1, CD117, S100, cytokeratin and ALK

Small Submucosal tumors are usually asymptomatic and are incidentally detected by endoscopic examination. A recent study reported incidence of gastric submucosal lesions to be 0.36%.[6] However, the incidence of CFTs in the GIT remains a diagnostic dilemma, first due to the rarity of the lesion and second, due to the presence of many stromal lesions in the gastrointestinal tract with similar overlapping histologic findings.

The differential diagnosis for GI tract CFT's include gastrointestinal stromal tumor (GIST), schwannoma, sclerosing leiomyoma, inflammatory myofibroblastic tumor (IMT), plexiform fibromyxoma and reactive nodular fibrous pseudotumor (RNFP).[4] So CFTs arising from stomach should be differentiated from other lesion like Sclerosing calcified GIST, which lack psammomatous calcifications and shows positive immunostaining for CD117, DOG-1 unlike CFT.[2]

Schwannoma can be found in the GIT including stomach and shows spindle cells with wavy nuclei which shows diffuse S-100 positivity, unlike in CFT. The spindle cells in sclerosing leiomyoma shows positive immunostaining for SMA and desmin. Inflammatory myofibroblastic tumors are composed of fibroblasts/ myofibroblasts, with rare foci of calcification with positive immunostaining for SMA and ALK.[7]

Reactive nodular fibrous pseudotumor is a rare lesion of the gastrointestinal tract, which is large in size and shows

proliferation of fibroblasts in a hyalinized collagenous stroma. The presence of calcification favours CFT and helps in differentiation of these two entities. RNFP shows positive immunostaining for vimentin, SMA and desmin.[4,8]

The morbidity associated with invasive surgical gastrectomy was a major driver for implementing endoscopic resection procedures. Nowadays, endoscopy has been accepted as first-line therapy for early gastric lesions with very low likelihood of lymph node metastasis. CFTs are incidental findings and are most commonly treated by local surgical resection. Till date, there are no cases of malignant transformation or any metastatic disease asssociated with CFT's have been reported.

4. Conclusion

In summary, we conclude that Gastric CFTs are benign lesions that are underdiagnosed quite oftenly. Awareness about this entity would help many histopathologists to diagnose and differentiate this entity from other spindle cell lesions of the GI tract. Making a correct diagnosis is considered as an important key step in management and prognosis. Further, dedicated studies are needed to know the pathogenesis and recurrence rate of CFTs for proper management of patients.

5. Source of Funding

None.

6. Conflict of Interest

None.

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