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Case Report

Not all Gastric Mesenchymal Tumors are Gists

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Abstract

Calcifying fibrous tumors (CFT) of the gastrointestinal tract are very rare soft tissue neoplasms, with only few cases reported in literature. A new case is described with review of the literature.

ABBREVIATIONS

GIST: Gastro-Intestinal Stromal Tumor; CFT: Calcifying Fibrous Tumor

INTRODUCTION

Gastric mesenchimal tumors are most commonly represented by gastro-intestinal stromal tumors (GISTs) and leiomyomas. However other uncommon mesenchimaltumours can arise in gastric wall. Among them, calcifying fibrous tumors (CFT) of the gastrointestinal tract are very rare soft tissue neoplasms, with only few cases reported in literature.

CASE PRESENTATION

A41 year old man was admitted to our General Medicine Unit for alcohol-related liver disease. During work up, a MRI was performed, showing a gastric nodular lesion with calcifications. The lesion measured 3.9 cm in maximum diameter and was localized within gastric fundus wall. A clinical diagnosis of GIST was made and resection of the mass with part of gastric wall was performed.

Gross examination revealed a well-circumscribed, non-encapsulated, firm, white mass (Figure 1A-1B), located in the muscle wall (Figure 1C-1D) with extension into the sub-serosa; the lesion was covered by sub-mucosa and mucosa with no evidence of ulceration. At histology, the lesion was predominately composed of densely hyalinized collagenous fibers, with a vaguely wavy-storiform pattern and with sparse spindle cells without atypia or mitoses (Figure 2A). Calcifications were diffuse, with different recognizable patterns: longitudinal calcifications, following the contour of collagen fibers with progressive deposits of calcium within the collagen fibers itself (Figure 2A); psammomatous bodies (Figure 2B-2C); large dystrophic calcifications (Figure 2D). A mononuclear inflammatory infiltrate, composed predominantly of B and T lymphocytes was present; nodular lymphoid aggregates were also seen (Figure 1E). The tumor was immunohistochemically negative for CD117, DOG1, smooth muscle actin, desmin, S100 protein, CD34, ALK and IgG4 while expressing vimentin. The lesion was diagnosed as calcifying fibrous tumor (CFT).

DISCUSSION

CFTs are rare soft tissue tumors, found ubiquitously, with benign behavior but a recognized tendency for local recurrence [1]. The pathogenesis of CFT is still unknown; isolated case reports considered CFT to be the sclerosing end stage of

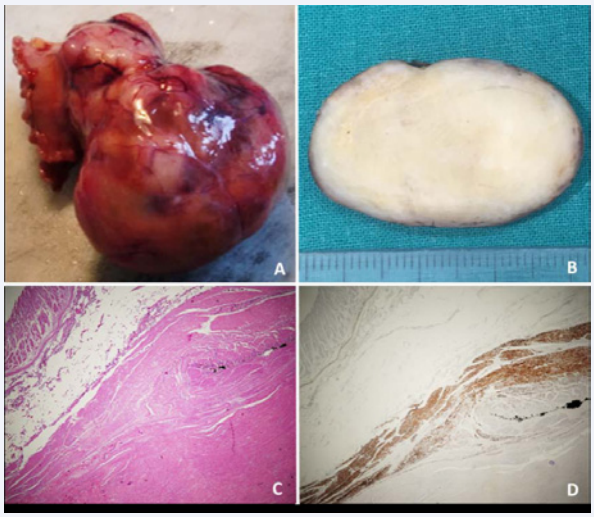

Figure 1 A) Gross aspect of the lesion, with sub-serosal extension. B) Gross transverse section of the tumor showing a well delimited white lesion with small yellowish calcified areas. C-D) Haematoxylin and Eosin (C) and desmin (D) stained section (magnification x4) showing normal overlying mucosa and submucosa and the lesion (desmin negative) within the muscle wall (desmin positive).

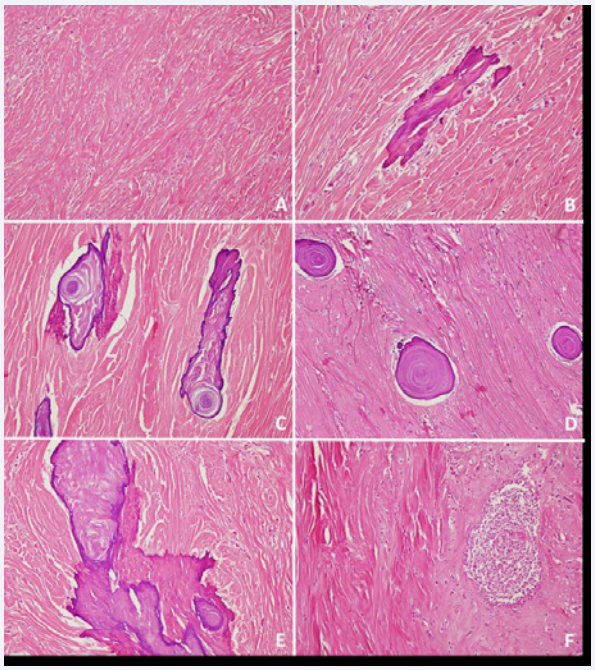

Figure 2 Haematoxylin and Eosin stained section of the tumor. A) Microscopic aspect of CFT showing densely hyalinized collagenous fibers, with a vaguely storiform growth pattern (magnification x10). B-C-D-E) Different patterns of calcification with linear-longitudinal (B), transitional linear to psammomatous (C), psammomatous (D) and large dystrophic calcifications (E) (magnification 10x and 20x). F) Lymphoid aggregate within the collagenous hyalinized fibers of CFT (magnification 10x).

inflammatory myofibroblastic tumor [2], but this has not been confirmed in larger series [1].

Gastrointestinal CFTs are rare; 23 cases originating from the gastric wall are described, mainly as single case reports except for a larger series of 7 cases by Agaimy [3]. About half of the reported cases have been described after the publication of the Agaimy series, thus probably reflecting a major attention for this under-recognized and often misdiagnosed lesion (as sclerosing calcified GIST or sclerosing leiomyoma). Summarizing the information available from the literature and the current case, CFTs show no sex predilection (12M/12F), with a mean age of 51 yrs (range 37-77), and a mean size of 2.1 cm (range 0.8-3.9). CFT are more frequently located in the gastric body or fundus (13/16 cases) with variable involvement of the gastric wall both as polypoidsubmucosal lesions and subserosal masses.

Main differential diagnoses include: sclerosing calcified GIST (which is a common aspect mainly in incidental small lesion), sclerosing leiomyoma and inflammatory myofibroblastic tumor: typical morphology of CFT coupled with immunohistochemistry negativity for specific markers are necessary for diagnosis.

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