

Calcifying Fibrous Tumor: Report of an Exceptional Lesion Localized to the Jejunum

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Abstract

Background: Calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor; with a predilection for children and young adults that usually arises in the subcutaneous and deep soft tissues, pleura, or peritoneum. CFT of the gastrointestinal tract is exceedingly rare.

Discussion: We describe here a patient with an unusual presentation-intestinal calcifying fibrous tumor. This 26-year-old man came to our emergency department with abdominal distension and intermittent epigastric cramping pain. The physical examination was negative. Colonoscopy showed an intestinal polyp, measuring 3 cm in greatest axis. The macroscopic examination showed a segmental resection of jejunum with pedunculated polyp. The polyp had a smooth, shiny and gray surface, it measured 3 cm. Histologically, it was consisting of hyalinized, hypocellular lamellar collagen, bland spindle cells, chronic inflammatory cell infiltrates, and psammomatous or dystrophic calcifications.

Conclusion: CFT of the gastrointestinal tract is exceedingly rare. We describe a case arising in the small intestine, and discuss the differential diagnosis with other common and uncommon spindle cell lesions.

Keywords: Mesenchymal tumor; Gastrointestinal tract; Immunohistochemistry

Abbreviations: CFT: Calcifying Fibrous Tumor; GIST: Gastrointestinal Stromal Tumor; HE: Hematoxylin Eosin; IHC: Immunohistochemistry

Introduction

Calcifying fibrous tumor (CFT) is a rare benign mesenchymal tumor affecting mainly young people. It is most commonly found in the soft tissues of the extremities and pleura [1]. A total of 157 cases of CFT were identified until May 2016. The most common locations of CFT were stomach (18%), small intestine (8.7%), pleura (9.9%), mesentery (5%), and peritoneum (6.8%) [2]. In the light of an observation and review of the literature, we aim to identify epidemiological, histological, immunohistochemical and prognostic parameters of this rare entity.

Case Report

We report a case of a 26-year-old man with no significant medical history. He consulted for abdominal distension and intermittent epigastric pain with the loss of 4 kg in one month. The abdominal examination was unremarkable. The evaluation didn't include neither abdominal scannography nor echography and laboratory tests were without abnormalities. A colonoscopy was then performed and highlighted a 3 cm jejunal polyp. No biopsy of the polyp was performed at that time and a resection was done during the endoscopy. The macroscopic examination showed a 3 cm, gray, smooth pedicle polyp localized in the submucosa. Microscopic exam noticed a jejunal submucosa containing a well limited benign mesenchymal tumor (Figure 1). The tumor was composed of fibrous hyalinized, hypocellular tissue with a chronic inflammatory infiltrate (Figure 2) and dystrophic calcifications with psammomatous bodies (Figure 3). The tumor cells express strongly the vimentin antibody (Figure 4). Immunohistochemical exam showed that fusiform cells

express CD34 (Figure 5). Moreover, Factor VIII was expressed only in vessels. This tumor doesn't express CD117, dog 1 which eliminates gastrointestinal stromal tumor (GIST). PS100 and bcl 2 was also negative. Furthermore, smooth muscle markers showed negative expression. Thus, histological features and immunohistochemical results confirm the diagnosis of CFT.

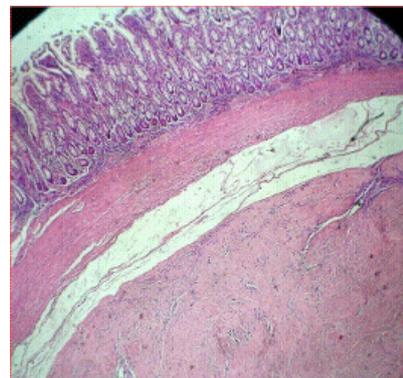


Figure 1: Jejunal submucosa containing a well limited benign mesenchymal tumor (Hematoxylin eosin x 40).

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Discussion

The TFC is recognized as a benign mesenchymal tumor that affects a young population: children and young adults without sex predominance [1]. The predilection sites are the soft tissues of the extremities, pleura and rarely digestive and intra thoracic structures [1]. However, the average age changes according to location: 14.5 years for extra-thoracic location, 38 for pleural and 30 for gastric one [2].

Involvement of the digestive tract is rare. Gastrointestinal CFTs are usually submucosal and range from 0.5 to 11.0 cm in great diameter, with an average size of 2.6 cm. Macroscopically, they are well demarcated, unencapsulated, lobulated masses with a solid and

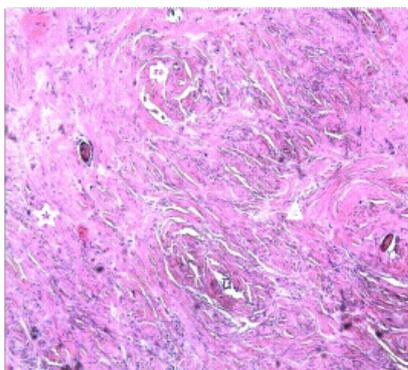


Figure 2: Benign mesenchymal tumor composed of spindle cell proliferation, with dense collagen fibers and loosely aggregates lymphocytes (Hematoxililn Eosin x 200).

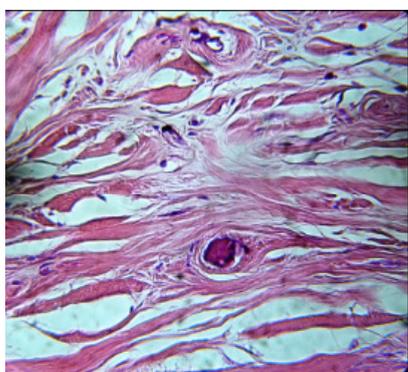


Figure 3: Hypocellular tissue with bands of collagen, chronic inflammatory infiltrate and psammomatous bodies (Hematoxililn eosin x200).

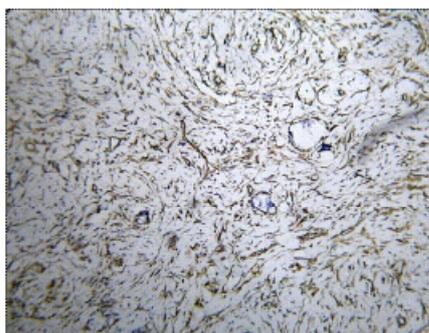


Figure 4: Tumor cells expressing strongly the Vimentin anti-body (Immunohistochemistryx100).

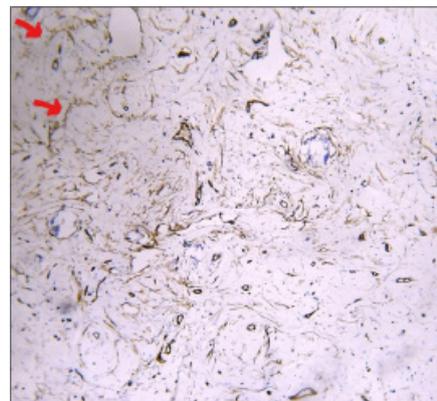


Figure 5: Tumor cells expressing strongly the CD 34 anti-body (arrow) (Immunohistochemistryx100).

white to gray cut surface. Yellow calcifications are sometimes grossly identifiable.

Microscopically, CFT is defined by the following 3 components: (1) abundant, paucicellular, hyalinized collagen; (2) interspersed calcifications; and (3) an inflammatory infiltrate. The collagenous matrix often exhibits a whorled or storiform pattern but may be haphazard or pattern less. Bland spindle cells are embedded within the abundant collagen. The spindle cells exhibit ovoid, vesicular nuclei with fine chromatin and inconspicuous nucleoli and abundant eosinophilic to amphophilic cytoplasm. Atypia and mitotic figures are lacking. The calcified component, dispersed throughout the fibrotic areas, may be either psammomatous or dystrophic. The inflammatory component is predominantly composed of lymphocytes and plasma cells infiltrating singly or forming aggregates [3].

In fact, in digestive localization, CFT represents a real problem of differential diagnosis with gastrointestinal stromal tumor (GIST), schwannoma, the sclerosing leiomyoma and myofibroblastic inflammatory tumor.

GISTs often show hyalinized tumors, dystrophic calcifications but no real psammoma bodies or lymphoplasmacytic infiltrate and it expresses intensely and diffusely the CD117 antibody [4,5].

Schwannoma expresses the PS-100 and the sclerosing leiomyoma expresses smooth muscle actin and H-caldesmon, when inflammatory myofibroblastic tumor expresses smooth muscle actin and desmin [5]. The inflammatory fibrinoid polyp, unlike the CFT, prevails in the antrum. It is in the submucosa with eosinophilic infiltrate and marked hypercellularity [6].

The pathogenesis of CFT is unclear. Researchers suggested that it may be a reaction process between inflammation, a form of fibrous pseudo tumor or the final stage of inflammatory myofibroblastic tumor. However, a study compared 7 cases of CFT and 7 cases of inflammatory myofibroblastic tumor on histology and immunology, no link has been proven. Positive CD34 and collagen hyalinization could link with the solitary fibrous tumor [7].

Surgical treatment is based on the complete excision because the recurrence rate is estimated at 10%. [7]. This lesion is described to have favorable prognosis. No patient presented recurrence particularly for digestive localizations [8].

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