Rev Med Chile 2023; 151: 1646-1649

Intestinal perforation as a form of presentation of a small bowel calcifying fibrous tumor

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The authors declare that they have no conflicts of interest.

Author contributions: All authors have made substantial contributions to the conception and design of the study, acquisition of data, analysis, interpretation of data, drafting of the article or revising it critically for important intellectual content, and final approval of the version to be submitted. Authors have read and approved the submitted version of the manuscript.

Recibido el 03 de junio de 2023, aceptado el 27 de octubre de 2023.

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Calcifying fibrous tumor (CFT) is a rare, benign, mesenchymal tumor. It has a slight female predominance, and it can appear in any range of age. It can be in the extremities, neck, and gastrointestinal tract, but it has also been described in other locations. Even though it is a benign lesion, recurrence has been described in some cases in the literature. A free-margin surgical resection is the recommended treatment. We present a 56 -year-old woman who underwent surgery for an intestinal obstruction associated with middle jejunum perforation. Histopathological study described the presence of a calcifying fibrous tumor. Spindle cells were positive for CD34, Factor XIIIa and vimentin. To our knowledge, this is the first case of intestinal perforation secondary to a calcifying fibrous tumor described in the literature.

(Rev Med Chile 2023; 151: 1646-1649)

Key words: *Factor XIII; Surgery; Tumor; Vimentin.*

Perforación intestinal como forma de presentación de un tumor fibroso calcificante

El tumor fibroso calcificante (TFC) es un tumor mesenquimal, benigno poco frecuente. Tiene un ligero predominio femenino y puede aparecer en cualquier rango de edad. Se localiza en extremidades, cuello y tracto gastrointestinal, pero también se ha descrito en otras localizaciones. Aunque se trata de una lesión benigna, se han descrito recidivas en algunos casos en la literatura, por lo que el tratamiento es la resección quirúrgica con márgenes libres. Presentamos el caso de una mujer de 56 años que fue intervenida por un cuadro de obstrucción intestinal por perforación del yeyuno medio, en el que la anatomía patológica describió la presencia de un tumor fibroso calcificante. Las células fusiformes eran positivas para CD34, factor XIIIa y vimentina. Según estos hallazgos, se trata del primer caso de perforación intestinal secundaria a un tumor fibroso calcificante descrito en la literatura.

Palabras clave: *Cirugía; Factor XIII; Tumor; Vimentin.*

Calcifying fibrous tumor (CFT) is a rare, benign mesenchymal neoplasm initially considered a soft tissue tumor. The literature has described a few cases; their etiology and pathogenesis are still uncertain. Rosenthal and Abdul-Karim described it for the first time in 1988[1] and called it a "childhood fibrous tumor with psammoma bodies." Jejunal CFT was reported for

the first time by Liang in 2007 in a case of intestinal obstruction due to intussusception[9]. In 2002, the World Health Organization established the diagnosis of this lesion as a "Calcifying fibrous tumor."

The mean age of presentation is the third decade of life, and the incidence of this condition is higher in women[2]. Although it can occur in any other organ, the most common anatomical location is in the gastrointestinal tract, with the stomach and small intestine the most affected[3]. The histological tumor's characteristics are the presence of a dense, hyalinized collagenous tissue with spindle cells associated with a lymphoplasmacytic infiltrate. The calcifications are very characteristic and may be psammoma bodies or dystrophic calcifications. We report the first case described in the literature of intestinal perforation due to TFC.

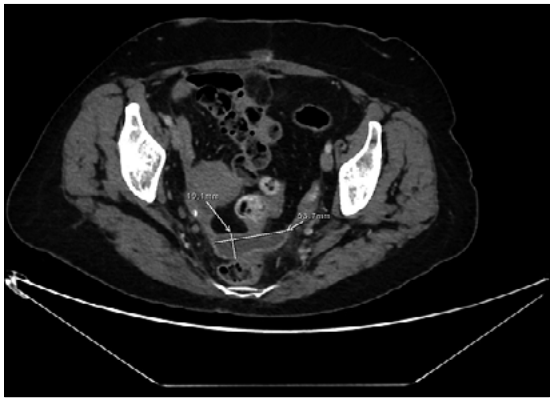

Figure 1. Computerized axial tomography. The white lines point to the described tumor with extraluminal collection and bubbles.

Case report

A 56-year-old woman, a smoker, presented to the emergency department due to the absence of stool and abdominal pain for 24 hours. She was diagnosed with CREST syndrome and was under immunosuppressive treatment with corticosteroids. Her surgical history was remarkable for appendectomy and laparoscopic cholecystectomy. The patient had a blood pressure of 136/85 mm Hg, a heart rate of 98 bpm, and an oxygen saturation of 96%. Physical examination revealed a distended abdomen, diffuse pain on palpation, and rebound tenderness. Blood analysis showed leukocytosis 24.5 10^3/µL [normal values (NV), 4.5 - 11.0] and neutrophilia 91% (NV, 50.0 - 70.0), amylase 89 U/l (NV, 30 - 118), lipase: 106 U/l (NV, 73 - 393) and C-reactive protein: 18.0 mg/l NV, < 10.0). An urgent abdominal computed axial tomography with intravenous contrast was requested, which showed mucosal enhancement in jejunal loops, submucosal edema, decreased caliber, and retrograde dilatation of the rest of the small bowel loops up to a collection of approximately 50 x 50 x 50 mm diameter with presence of air and trabeculation of the adjacent mesenteric fat suggestive of perforation (Figure 1). Given these findings and the patient's clinical presentation, an immediate surgical intervention was decided.

Under general anesthesia, an exploratory laparotomy was performed by a supra-infra umbilical midline incision, revealing generalized purulent

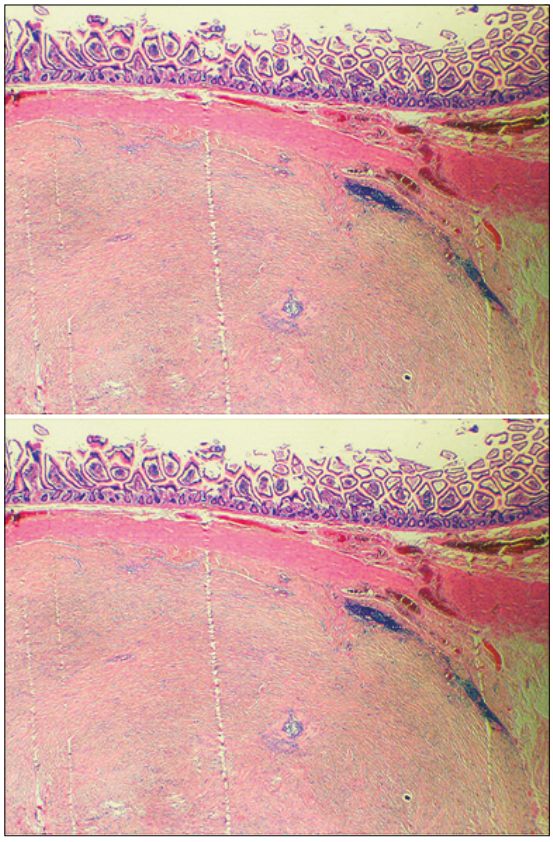

Figure 2. Microscopic image of the piece. Well-demarcated, unencapsulated, hypocellular fibroblastic proliferation growing within a dense collagenous matrix, with foci of dystrophic calcification, surrounded by a lymphocytic cuff. No evidence of atypia, mitosis, or necrosis.

peritonitis as an abdominal collection on the middle jejunum. The abdominal cavity was washed out, and adhesiolysis of the intestinal package was performed; a perforation was located at the antimesenteric side of the mid-jejunum. Resection of the middle jejunum and isoperistaltic manual biplane anastomosis was done.

The patient presented a favorable postoperative course; a paralytic ileus was treated without surgery and was discharged after 11 days of hospitalization.

The histopathological result of the small bowel surgical specimen showed an abundant fibrin deposit on the external surface with a solution of continuity, showing a small whitish fibrous nodule of 0.7 cm in diameter at the mucosae (Figure 2). The histopathological study revealed a not encapsulated, hypocellular, well-demarcated fibroblastic proliferation within a dense collagenous matrix with dystrophic calcification foci surrounded by a lymphocytic cuff, without atypia, mitosis, or necrosis. The immunohistochemistry study was positive for CD34, vimentin, and Factor XIIIa and negative for S100 protein, ALK, Desmin, Actin, CD117, DOG1 and STAT 6 (Figure 3).

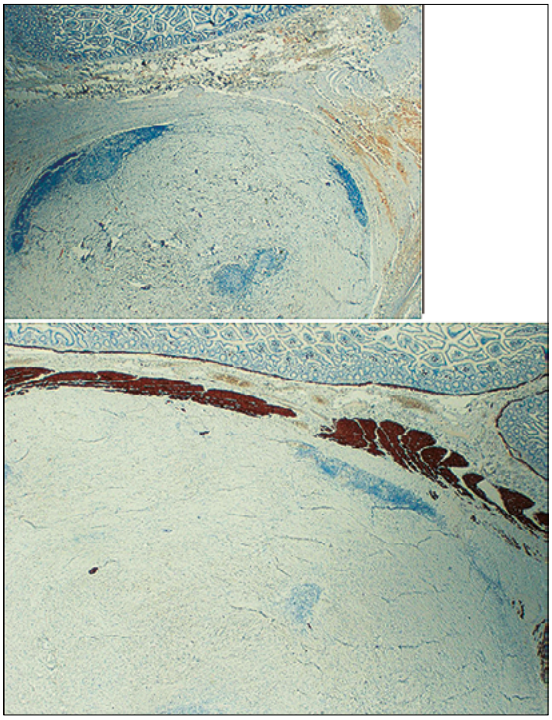

Figure 3. Immunohistochemistry CD34: Positive; S100, ALK, Desmina, Actina, CD117, DOG1, STAT 6: Negatives.

Discussion

A calcifying fibrous tumor is a benign proliferation of fibroblastic cells associated with an inflammatory component and calcifications of unknown origin. Less than 200 cases have been described in the literature[15]. Although its pathogenesis is unknown, some authors hypothesize that CFT may originate from an inflammatory pseudotumor, mediated by an abnormal immune response[14], be related to an excessive inflammatory response following trauma, or may be associated with IgG4-mediated diseases[3,8].

TFC is more frequent in children and young adults, with a slight prevalence in the female gender. The mean age of presentation is in the third decade of life. It is mainly located in the stomach, small bowel, pleura, and neck, but it has also been described in other locations such as the mediastinum, peritoneum, lower limb, adrenal glands, back, lung, and pericardium[2].

Patients are usually asymptomatic. The clinical manifestations are nonspecific and depend mainly on their location, such as intestinal obstruction or mass effect. Incidental diagnosis is common in imaging tests such as ultrasound, CT, or MRI. They are identified as homogeneous lesions on ultrasound, with well-delimited margins, calcifications, and slight enhancement[4]. Magnetic resonance imaging shows a typical hypointense signal in T1 and T2 isointense in T1 after gadolinium administration[2]. Macroscopically, they are well-demarcated, rounded, or lobulated, not encapsulated tumors, firm to cut, and of a whitish, tan, or gray color. Their size varies from 0.5 to 11 cm, but the average size described is 2,6 cm[3]. Although they appear as single lesions, up to 10% may present as multiple lesions at the abdominal level, mimicking a peritoneal carcinomatosis[13].

The preoperative differential diagnosis is difficult. Definitive diagnosis is made by histopathological findings, where intense fibrosis, with inflammation and intercalated calcifications, is observed[3]. Immunohistochemistry is characteristic since they stain intensely and diffusely with vimentin and Factor XIIIa (Figure 4). Differential diagnosis must be made with inflammatory myofibroblast tumors (IMT), IgG4-related disease, leiomyoma, schwan-

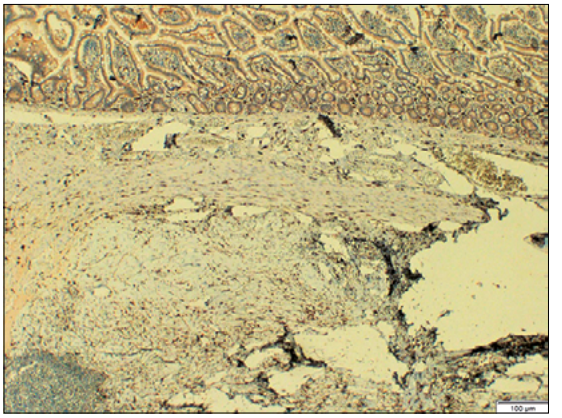

Figure 4. Immunohistochemistry. FACTOR XIIIa positive.

noma, fibromatosis, low-grade fibromyxoid sarcoma and gastrointestinal stromal tumors (GIST)[7]. Staining for CD34, smooth muscle actin, CD 658, and desmin is variable. Some authors believe these tumors may be related to IgG4 disease[5].

The management and treatment depend on its clinical manifestations and localization, with surgical excision being the most appropriate. Despite its benign nature, local recurrences have been described in up to 20% of patients without malignancy or evidence of metastases. Because of this, clinical follow-up is not necessary.

Conclusions

Calcifying fibrous tumors are rare, benign tumors. Their clinical presentation is variable, from asymptomatic to intestinal occlusion and perforation. The differential diagnosis should be made with other neoplastic lesions such as GIST, leiomyomas, and sarcomas. The treatment is mainly surgical. Although recurrence has been described, no malignancy cases are reported in the literature.

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