Calcifying Fibrous Tumor of the Mesentery: A Case Report and a Review of the Literature

Clinical Pathology
Volume 13: 1–3

DOI: 10.1177/2632010X20930689

Derqaoui Sabrine[1], Elouazzani Hafsa[1], Ratbi Amine[2], Bernoussi Zakia[1] and Zouaidia Fouad[1]

[1]Department of Pathology, Ibn Sina Teaching Hospital, Rabat, Morocco. [2]Surgical Department "C," Ibn Sina Teaching Hospital, Rabat, Morocco.

ABSTRACT

BACKGROUND: Calcifying fibrous tumor (CFT) is a rare entity, with a distinctive histological presentation, initially reported as childhood fibrous tumor with psammoma bodies. It is a benign hypocellular fibrous neoplasm calcifications and lymphoplasmacytic infiltrate. The CFTs may involve many sites, including gastrointestinal tract, pleura, abdominal cavity, and neck. The diagnosis might be challenging due to histological overlaps with other mesenchymal tumors. The prognosis is good. We describe herein the case of a 53-year-old woman with an incidentally diagnosed CFT of the mesentery.

CASE PRESENTATION: A 53-year-old woman presented to the surgery department with a 2-year history of an anterior abdominal hernia. A computed tomographic scan of the abdomen failed to demonstrate any evidence of a mesenteric nodule. The patient underwent surgical treatment. Careful exploration during the excision of herniated sac revealed a solitary nodule of the mesentery. Local excision was performed. On gross, it was a well-demarcated nodule. Microscopically, the tumor consisted of an abundant paucicellular hyalinized collagen with calcifications; associated to a sparse mononuclear inflammatory infiltrate.

CONCLUSIONS: Calcifying fibrous tumor is a benign lesion. The diagnosis is based on histology, because clinical and radiological features are nonspecific. Awareness of this entity is crucial to distinguish it from other mesenchymal tumors especially in the gastrointestinal tract.

KEYWORDS: Calcifying fibrous tumors, histopathology, mesentery

RECEIVED: April 20, 2020. **ACCEPTED:** May 9, 2020.

TYPE: Case Report

FUNDING: The author(s) received no financial support for the research, authorship, and/or publication of this article.

DECLARATION OF CONFLICTING INTERESTS: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

CORRESPONDING AUTHOR: Derqaoui Sabrine, Department of Pathology, Ibn Sina Teaching Hospital, Abderrahim Bouabid Avenue, Rabat, Morocco. Email: s.derqaoui@gmail.com

Introduction

Calcifying fibrous tumor (CFT) is a rare entity, with a distinctive histological presentation.[1] It is a benign hypocellular fibrous neoplasm, commonly presenting as a solitary lesion during the second and third decades of life.[2] Originally described as subcutaneous soft tissue tumor, CFTs have been reported in multiple locations (gastrointestinal tract, pleura, abdominal cavity, and neck).[3] Histologically, the presence of calcifications and lymphoplasmacytic infiltrate represents the hallmark of CFT.[2] The diagnosis might be challenging, as histological overlap with other mesenchymal tumors has been reported. Local resection with clear margins is the gold standard treatment for CFT.[3] To date, the results are encouraging as long-term survival is 100% (without any reported deaths).[4] We describe herein the case of a 53-year-old woman with an incidentally diagnosed CFT of the mesentery to provide more information about this rare entity.

Case Presentation

A 53-year-old woman presented to the surgery department with a 2-year history of an abdominal pain, without nausea, vomiting, diarrhea, or constipation. She had no significant comorbidities, except a previous surgical treatment for liver hydatid cysts 9 years ago. Upon physical examination, she was a healthy female with stable vital signs. Her abdominal examination showed a reducible mass on the anterior abdominal wall without strangulation signs. A computed tomographic scan of the abdomen showed evidence of anterior abdominal hernia containing a small bowel loops. Whereas it failed to diagnose any mesenteric nodule because this later was a small size. The patient underwent surgical treatment, consisting of reinforcement of the abdominal wall with a prosthesis for long-term successful repair. Careful exploration during the excision of herniated sac revealed a solitary nodule of the mesentery. Local excision was performed. On gross, it was a well-demarcated, spherical nodule measuring 1.1 cm × 0.6 cm, with a white to gray cut surface. The texture was firm at sectioning. Microscopically, hematoxylin-eosin stained sections revealed an abundant paucicellular hyalinized collagen, with interspersed normal and dystrophic calcifications, associated to a sparse inflammatory infiltrate consisting of lymphocytes and plasmocytes (see Figure 1).

Discussion

Calcifying fibrous tumor is a rare, benign, mesenchymal lesion, which was first described in 1988 by Rosenthal and Abdul-Karim as childhood fibrous tumor with psammoma bodies.[5] It has been reported in larger series in 1993, as calcifying fibrous pseudotumor.[5] Approximately, 161 cases have been reported to

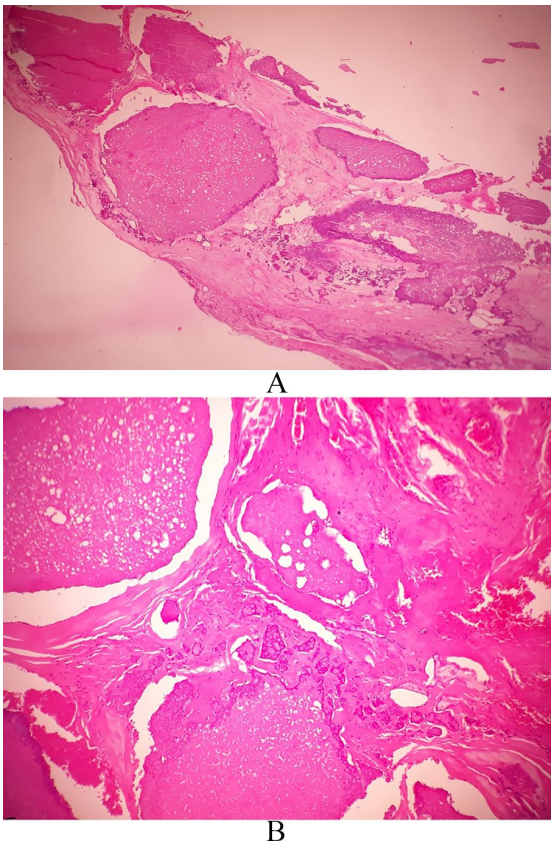

Figure 1. Calcifying fibrous tumor showing abundant paucicellular hyalinized collagen with interspersed normal and dystrophic calcifications, associated to a sparse inflammatory infiltrate consisting of lymphocytes and plasmocytes: (A) H&E 100× and (B) H&E 200×. H&E indicates hematoxylin-eosin.

date.[4,6] Calcifying fibrous tumor has been described in a variety of sites, other than soft tissue and in a wide age range.[7] It mainly affects women with male-to-female incidence ratio of 1:1.27 and an average age at the time of diagnosis of 33.58 years.[7] According to a meta-analysis by Chorti et al, age distribution shows a trimodal pattern with the first pick from birth 0 to 4 years, the second in the mid-20s, and the third in the mid-30s. In addition, the most commonly involved sites are the stomach (18%), the small intestine (8.7%), the pleura (9.9%), the neck (6.2%), the peritoneum (6.8%), the mediastinum (5%), and the mesentery (5%).[8] Most CFTs present as solitary lesions, although multifocal tumors have been reported.[3] Clinically, CFTs are asymptomatic in approximately 70% of the cases and discovered incidentally,[9] if symptomatic; they might present with nonspecific signs (including lack of appetite, fever, weight loss, fatigue, progressive weakness, chest or back pain, and dyspnea) or with specific signs according to the involvement's site.[8] In gastrointestinal tract, CFT might rarely cause serious complications, including bowel obstruction, intussusception, or ischemic necrosis.[9] Imaging studies of CFTs reflect their histological composition, they characteristically present as well-circumscribed mass containing scattered calcifications in ultrasonography as well as in computed tomography.[3] On endoscopic ultrasonography, CFTs may be hypoechoic masses with hyperechoic areas referring to calcifications.[5] Magnetic resonance imaging (MRI) findings include isosignal intensity on gadolinium-enhanced T1-weighted imaging and hyposignal intensity on T2-weighted imaging.[5] On gross, gastrointestinal CFT's size ranges from 0.5 to 11 cm (average size of 2.6 cm); they present as spherical, well-circumscribed, lobulated, solid masses with a white to gray cut surface. Yellow calcifications might be evident macroscopically.[3] The definite diagnosis remains on histology. In fact, CFTs are characterized by the following features: (1) abundant hyalinized paucicellular collagen, (2) psammomatous and/or dystrophic calcifications, and (3) mononuclear inflammatory infiltrate.[3] The collagenous matrix often shows a storiform pattern but may be patternless. The spindle cells embedded within the matrix are ovoid, with vesicular nuclei, fine chromatin, and inconspicuous nucleoli. The cytoplasm is abundant eosinophilic. Neither atypia nor mitotic figures have been reported.[5] Our case showed the classical histological features of CFTs. By definition, the diagnosis of CFT is morphologic; thus, immunohistochemistry might be of use in the differential diagnosis. Spindle cells are strongly and diffusely positive for Vimentin and Factor XIIIa,[5] and rarely positive for smooth muscle actin (SMA).[4] Calcifying fibrous tumor's immunoreactivity for CD34 has been variably reported in the literature.[3] An increased IgG:IgG4 ratio in the plasma cell population has been described, suggesting a potential association between IgG4-related disease and CFT.[3] In our case, the diagnosis was made on morphology findings. Calcifying fibrous tumor must be distinguished histologically from other spindle cell tumors, including sclerosing calcified gastrointestinal stromal tumor (GIST), schwannoma, hyalinized leiomyoma, IgG4-related sclerosing disease, inflammatory myofibroblastic tumor (IMT), solitary fibrous tumor, and reactive nodular fibrous pseudotumor.[9,10] Characteristic morphologic findings of CFTs include circumscription, paucicellularity, collagenous matrix, bland spindle cells, calcifications, and a chronic lymphoplasmacytic inflammatory infiltrate.[3] However, the diagnosis often requires exclusion of more common entities which can be ruled out by a careful histological and immunohistochemical analysis and, in some cases, on genetic analysis.[4,7] The neoplastic nature of CFT is controversial; the first genetic studies have been conducted by Mehrad et al[7] on 3 cases of pleural CFT. Thus, novel deleterious mutations in ZN717, FRG1, and CDC27 genes, associated with copy number losses on 8 chromosomes with a large loss common on chromosome 6, have been identified.[7] The etiopathogeny of CFTs remains unclear; several theories have been advanced comprising a posttraumatic or genetic pathogenesis as well as a relation to IgG4-related disease and IMTs.[4] In fact, a strong relationship between CFT and IMT has been suggested based on morphological studies, and CFT may be the last stage of IMT. By methylation profiling, Tomassen et al[10] showed that CFT and IMT have the same epigenetic profile, although hallmark fusion genes of IMT (*ALK*, *ROS1*, and *RET*) were not found in CFTs. Calcifying fibrous tumor is cured by local excision.[5] Although local

recurrence has been reported, the prognosis is excellent, and the long-term survival is 100%.[8] To date, neither deaths nor metastases have been reported.[3]

Conclusions

Calcifying fibrous tumor is a benign lesion thought of as a soft tissue tumor predominantly. However, several sites have been reported so far. The diagnosis is based on histology, because clinical and radiological features are nonspecific. Awareness of this entity is crucial to distinguish it from other mesenchymal tumors especially in the gastrointestinal tract. There is a need for further studies to understand the pathogenesis of these tumors.

Author Contributions

All authors read and approved the final manuscript.

Informed Consent

Informed consent was obtained from the patient.

ORCID iD

Derqaoui Sabrine https://orcid.org/0000-0002-8935-1346

REFERENCES

1. Nascimento AF, Ruiz R, Hornick JL, Fletcher CDM. Calcifying fibrous "pseudotumor." *Int J Surg Pathol*. 2002;10:189-196. doi:10.1177/106689690201000304.
2. Medina AM, Alexis JB. A 27-year-old woman with incidental omental nodules. *Arch Pathol Lab Med*. 2006;130:563-564.
3. Baumann KB, Orestes MI, Heaton SM, Whiting RE, Wendzel NC, Foss RD. Calcifying fibrous tumor of the neck. *Head Neck Pathol*. 2019. doi:10.1007/s12105-019-01100-7.
4. Prucker J, Salaheddin-Nassr Y, Leidl S. Calcifying fibrous tumor of the terminal ileum mesentery: case report. *Medicine (Baltimore)*. 2018;97:e13351. doi:10.1097/MD.0000000000013351.
5. Larson BK, Dhall D. Calcifying fibrous tumor of the gastrointestinal tract. *Arch Pathol Lab Med*. 2015;139:943-947.
6. Kwan BS, Cho DH. Calcifying fibrous tumor originating from the gastrohepatic ligament that mimicked a gastric submucosal tumor: a case report. *World J Clin Cases*. 2019;7:2802-2807. doi:10.12998/wjcc.v7.i18.2802.
7. Mehrad M, LaFramboise WA, Lyons MA, Trejo Bittar HE, Yousem SA. Whole-exome sequencing identifies unique mutations and copy number losses in calcifying fibrous tumor of the pleura: report of 3 cases and review of the literature. *Hum Pathol*. 2018;78:36-43. doi:10.1016/j.humpath.2018.04.005.
8. Chorti A, Papavramidis TS, Michalopoulos A. Calcifying fibrous tumor: review of 157 patients reported in international literature. *Medicine (Baltimore)*. 2016;95:e3690. doi:10.1097/MD.0000000000003690.
9. Pezhouh MK, Rezaei MK, Shabihkhani M, et al. Clinicopathologic study of calcifying fibrous tumor of the gastrointestinal tract: a case series. *Hum Pathol*. 2017;62:199-205. doi:10.1016/j.humpath.2017.01.002.
10. Tomassen T, Koelsche C, de Leng WWJ, et al. Calcifying fibrous tumor and inflammatory myofibroblastic tumor are epigenetically related: a comparative genome-wide methylation study. *Ann Diagn Pathol*. 2019;41:102-105. doi:10.1016/j.anndiagpath.2019.05.013.