Clinical Case Report

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Calcifying fibrous tumor of the terminal ileum mesentery

Case report

Johannes Prucker, MD[a,*], Yarub Salaheddin-Nassr, MD[b], Stefan Leidl, MD[a]

Abstract

Rationale: Calcifying fibrous tumors ("CFT") are recognized as extremely rare mesenchymal tumors with benign biological behavior and low rates of recurrence are seen after removal. The first case of a CFT was reported in 1988 as a possibly inflammatory triggered pseudotumor in deep soft tissue of children. Histologically, the tumor is typically composed of dense hyalinized collagen with paucicellular infiltration of lymphocytes and fibroblasts as well as psammomatous or dystrophic calcifications. It can affect soft tissue in very different anatomical locations, also intrathoracic and intra-abdominal, mimicking various different diagnoses. The etiology is understood to be unclear. Asymptomatic CFTs can be found incidentally on medical images.

Patient concerns: We present the case of a calcifying tumorous lesion found incidentally in the mesentery of the terminal ileum of a 34-year-old male patient in February 2016 undergoing a computed tomography for a urinary tract infection.

Diagnosis: Histopathological and immunhistochemical examination after surgery revealed a CFT.

Interventions: Our patient underwent lower abdominal median laparotomy for tumorectomy.

Outcomes: Two years after surgery the patient is free of a recurrence.

Lessions: We add another case of intra-abdominal CFT to medical literature to provide more information about this very seldom tumor. While the etiology of CFT should be further investigated, diagnosis and therapy seem clarified. CFT should be kept in mind as a rare differential diagnosis of calcifying tumors also in the abdominal cavity. Immunohistological work-up is important for finding the diagnosis and may also help solving pathogenetical questions.

Abbreviation: CFT = calcifying fibrous tumor.

Keywords: calcifying fibrous tumor (CFT), mesentery, soft tissue, mesenchymal tumor

1. Introduction

Calcifying fibrous tumors are very rare benign mesenchymal tumors. There are only approximately 160 cases reported in the international literature. Initially reported in 1988 as a deep soft tissue tumor in children,[1] the tumor was viewed as a pseudotumor[2] and thought to be of inflammatory or traumatic origin. In 2003, the prefix "pseudo" was abandoned and the name calcifying fibrous tumor was given due to the tendency to local recurrence.[3] Calcifying fibrous tumors (CFTs) generally occur in locations where soft tissue is present. CFTs reportedly occurs in the extremities, the abdominal and intrathoracal cavity, the neck and the oral cavity,[4] mimicking, depending on the location and proximity to other organs, various differential diagnoses.[5] Histologically, it is a characteristic for the tumor to demonstrate psammomatous calcification in a hypocellular mass of dense hyalinized collagen with mononuclear inflammatory infiltrates.[1–3,5] Although some histological overlap to other mesenchymal tumors, for example, the inflammatory myofibroplastic tumor is reported, CFTs usually can be clearly distinct but the etiology remains unclear.[4] The therapy consists of the removal of the mostly unifocal lesion, but depending on the location of occurrence, can vary from simple open excision of the tumor to endoscopic surgery. Recurrences are rare and reported with a rate of approximately 10%.[5] Though proven to be a benign disease a CFT can cause complications, for example, intestinal intussusception.[6] Since 2002, the CFT is listed in the WHO classification of soft tissue tumors and there exists a comprehensive overview and comparison of the reported cases in the literature by Chorti and Theodossis,[5] which also provides a good description of the tumor. Nevertheless, due to rare incidences, some aspects remain unclear and it is important to present new cases of CFTs for further investigation of the characteristics and the behaviour of the tumor. We present the probably 8th reported case in the literature of a mesenteric CFT, its process of the diagnosis and its therapy.

2. Case presentation

A 36-year-old male patient of caucasian ethnicity was admitted to our hospital in February 2016 with an incidentally found 6 cm

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[a] Landesklinikum Waidhofen an der Ybbs, Department of General Surgery, [b] Landesklinikum Amstetten, Department of Pathology, Austria.

[*] Correspondence: Johannes Prucker, Landesklinikum Waidhofen an der Ybbs, Department of General Surgery, Austria (e-mail: johannes.prucker@gmx.at).

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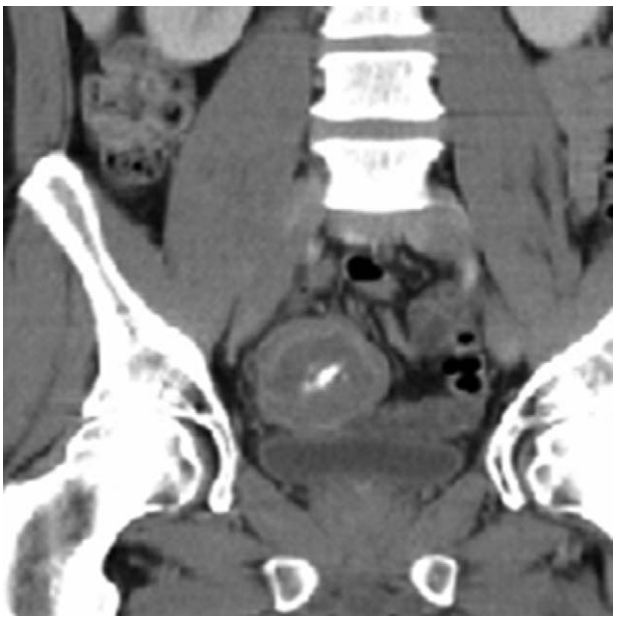

Figure 1. Coronal view of the abdominal computed tomography showing a 6 cm round mass with central calcification.

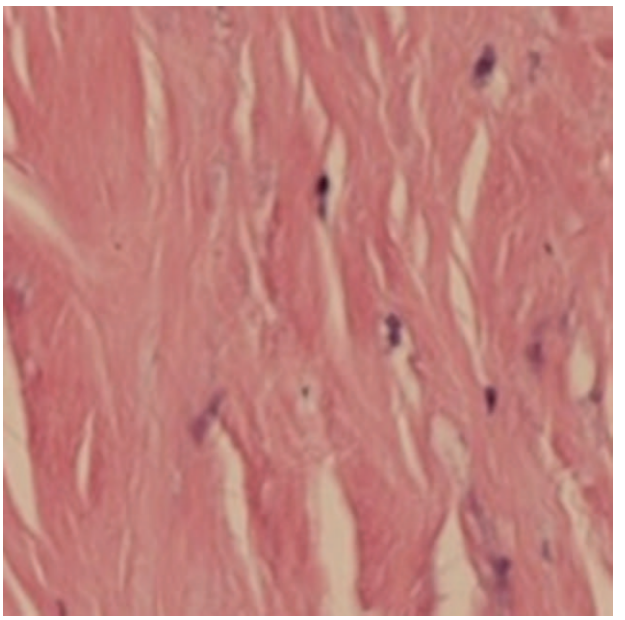

Figure 3. H&E stain (20×) showing hypocellular fibroblastic proliferation with hyalinized collagen representing the basic matrix of the tumor.

roundly mass in the lower right abdomen near the pelvic vessels and the intestine. The mass (Fig. 1) was found when performing a computed tomography extramurally for the investigation of a prolonged upper urinary tract infection of the right side. At the moment of the presentation, the patient was under antibiotic treatment, without symptoms and the physical examination was inconspicuous. Anamnestically the patient reported a recent episode of pain in the right iliac fossa, fitting the diagnosis of a subsiding urinary tract infection. The possible origin of the tumor could not be clarified and because of the visible calcifications, the initial radiological diagnosis was a suspect teratoma. A colonoscopy, laboratory tests, including tumor markers and the patient's personal medical history, brought no further clarification regarding the occurrence of the tumor. An MRI-scan was not obtained. The patient underwent lower median abdominal laparotomy in February 2016 and presented a solid tumor (Fig. 2) in the mesentery of the terminal ileum, partially retroperitoneally spreaded. After the removal of the tumor, the postoperative course was ordinary. The histological examination revealed a stony and hard to cut tumor consisting generally of a collagen matrix glowing under polarisation (Fig. 3). The hypocellular mass showed interspersed plasmocytes, lymphocytes, and the smallest capillaries (Fig. 4). Noticeable were mostly central incorporations of cloudy

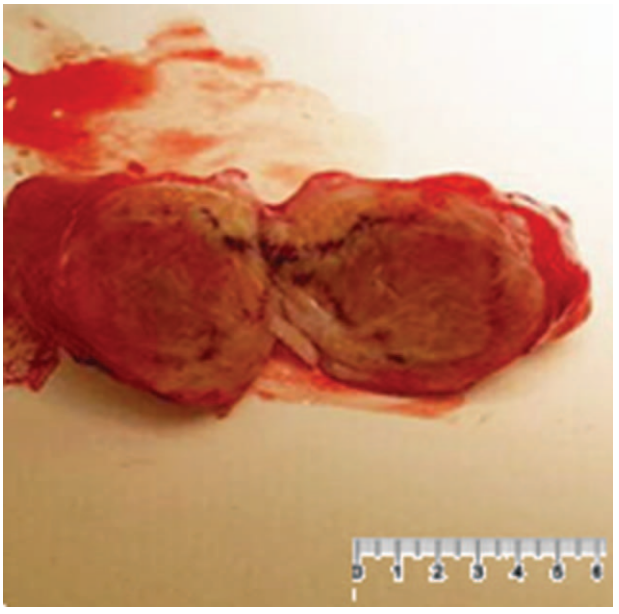

Figure 2. Postoperative cut of the tumor with measuring strip in cm.

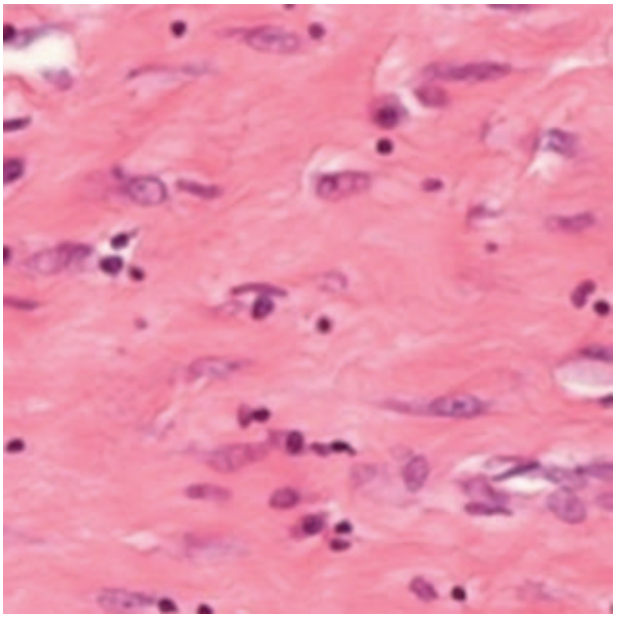

Figure 4. H&E stain (40×) showing interspersed plasmocytes, lymphocyts, fibroblasts, and fibrocytes with a higher cuff-like incidence under the surface of the tumor.

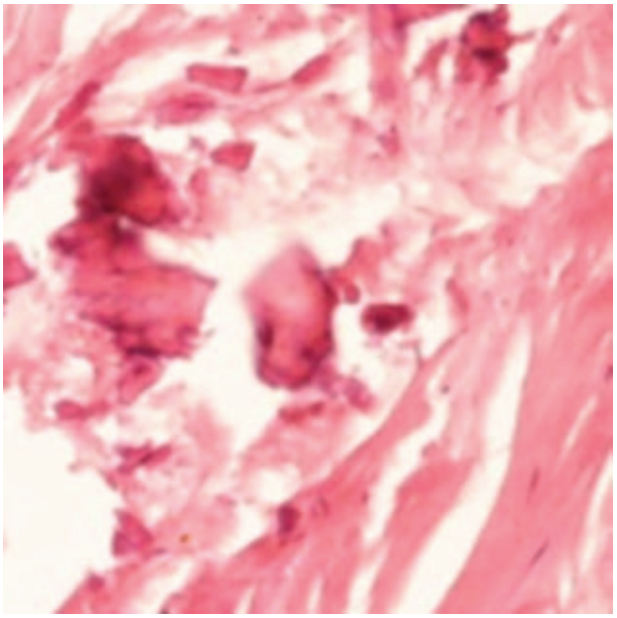

Figure 5. H&E stain (40×) showing central, extended cloudy calcifications.

calcifications (Fig. 5). Under the surface of inconspicuous mesothelium there was a band of a lymphoplasmocytic infiltrate. Immunohistochemically, the tumor showed sparse fibrocytes within the collagen fibres expressing vimentin and some fibers also expressing actin. Caldesmon, CD 34, and Pan-CK were negative. Desmin was negative for 99% of the mass. The follow-up examination after 1 and 2 years showed a symptom-free patient after laparotomy. An MRI scan showed that there was no recurrence of the tumor. In a retrospective case report an ethical approval was waived, but the patient gave informed consent to publish his case.

3. Discussion

CFTs are an interesting subject to investigate, due to their rare occurrence, unclear etiology and the possibility to occur in nearly every location of the human body and mimick other diseases depending on the location and proximity to other organs. In the overview of Chorti and Theodossis[5] there are 3 life-span peaks of occurrence named—early childhood, mid-20s and mid-30s. The early childhood peak could represent a genetic or embryologic pathogenical pathway. This idea is emphasized by findings of diploid cells in tumor fibrocytes and a reported case of familial occurrence.[7,8] The second peak could be vaguely associated with a trauma, and the third peak could confirm the popular and often discussed idea that CFTs are an end-stage presentation of an inflammatory myofibroplastic tumor which has the peak of occurrence several years before. Some authors investigated this theory but have not reached a clear conclusion.[3,9] Furthermore, there were elevated IgG4–levels found in the serum of CFT patients and IgG4 positive plasma cells in CFTs. This could indicate a linkage to IgG4-related diseases, which might be of importance due to the different treatment of CFTs and IgG4 related diseases.[10,11] IgG4-related diseases also tend to form a mesenchymal tumor that can occur at various different locations of the body but is treated with steroids. Unfortunately there is little data on the measurement of IgG and IgG4 in CFTs. Also the ratio of IgG to IgG4 may be of interest,[10] but a recent case of retroperitoneal CFT where those parameters were measured did not confirm this theory.[12] Definite etiology remains to be further investigated. A slight female predisposition in CFTs can be observed, without an explanation for this so far.[5] Though unifocal in the vast majority of cases,[5] in a mesenteric presentation the tumor was often seen multifocal.[6,7,13]The therapy of CFTs is surgical removal. Prognosis of CFT after removal is excellent, few recurrences and no death related to CFT are reported.[5] Laparoscopical removal of CFTs are reported[14] but due to the size of the mass, the hidden localization and unknown pathology the open approach via a lower median laparotomy seemed appropriate in our case. A definite diagnosis was made by pathologists, microscopically seeing a well-defined collagenous paucicellular tumor with central dystrophic calcifications and a few spread lympocytic infiltrates. Furthermore, the immunohistological distinction is very important, showing positivity in most of the cases of CFTs with Vimentin, Factor XIIIa and IgG4. CD34 and SMA are mostly negative but also positivity is reported in some cases.[5] The main intra-abdominal differential diagnosis is a gastrointestinal stroma tumor, but it can clearly be distinguished due to being a cellular mass, lacking collagen and calcifications and being positive to CD 117. Further the above named inflammatory myofibroblastic tumor can have a histological pattern similar to a CFT. It can however be distinguished for its positivity of desmin, and often the oncogenic ALK-1 gene.[3,9] Also the solitary fibrous tumor, which is mostly situated in the thoracic cavity, positive for CD34 and hypercellular, is a potential differential diagnosis regarding mesenchymal tumors in the abdomen.[15] Histologically, similar patterns may also be shown by schwannomas or leiomyomas; however, the schwannoma can be clearly ruled out by S100 positivity, and the leiomyoma via positivity for actin, desmin, and caldesmon. Unluckily, IgG4 was not measured in our case but may also be of diagnostic importance, measured in the lesion as well as the patients serum-levels.[10] Further differential diagnosis are desmoids, fibromatosis, nodular fasciitis, solitary fibrous tumor, and desmoplastic fibroblastoma.[5] In conclusion, the characteristics to differentiate CFTs are advanced with every reported case yielding a benign soft tissue tumor. Our patient, presenting in his mid-30s with a mesenteric CFT, could represent the above-mentioned etiological theory of CFT being the result of an end-stage inflammatory myofibroplastic tumor, which tends to occur in the abdominal cavity and the retroperitoneum and can also show calcifications.[3,5,9] Location in the mesentery is particularly interesting because only about 6% of cases of CFTs were multifocal and most of these cases were either mesenteric or pleural CFTs.[5,6,13] We have no explanation for this tendency of multifocality in mesenteric CFTs and in contrast, our case was unifocal. The etiology of CTF is diversely discussed. A post-traumatic or genetic pathogenesis as well as a relation to inflammatory myofibroplastic tumors or IgG4-related disease seems possible. An important part in clinical practice is to perform a sufficient immunohistochemical work-up. In the past positivity against IgG4 was examined only in a minority of cases of CFTs. IgG4 positivity of the tumor and also the serum IgG4-level should be measured in patients with CFT to elucidate a possible connection to IgG4-related disease.[5,10,11] Immunohistochemical investigation is important not only to confirm the diagnosis, but also to gain more information about the characteristics of CFTs and be able to advance with the research of the yet unclear etiology.

Author contributions

Conceptualization: Johannes Prucker.
Project administration: Johannes Prucker.
Supervision: Stefan Leidl.
Validation: Yarub Salaheddin-Nassr.
Visualization: Johannes Prucker.
Writing – original draft: Johannes Prucker.

References

[1] Rosenthal N, Abdul-Karim F. Childhood fibrous tumor with psammoma bodys. Clinicaopathological features in two cases. Arch Pathol Lab Med 1988;112:798–800.
[2] Fetsch J, Montgomery E, Meis J. Calcifying fibrous pseudotumor. Am J Surgery Path 1993;17:502–8.
[3] Nascimento AF, Ruiz R, Hornick JL, et al. Calcifying fibrous "pseudotumor" clinicopathological study of 15 cases and analysis of relationship to inflammatory myofibroblastic tumor. Int J Surg Pathol 2002;10:189–96.
[4] Bell DM, Dekmezian RH, Husain SA, et al. Oral calcifying fibrous pseudotumor: case analysis and review. Head Neck Pathol 2008;2:343–7.
[5] Chorti A, Theodossis S. Papavramidis, and antonios michalopoulos. Calcifying fibrous tumor. Review of 157 Patients reported in internationla literature. Medicine 2016;95:e3690.
[6] Liang HH, Chai CY, Lee CH, et al. Jejunal and multiple mesenteric calcifying fibrous pseudotumor induced jejunojejunal intussusception. J Formos Med Assoc 2007;106:485–9.
[7] Chen KT. Familial peritoneal multifocal calcifying fibrous tumor. Am J Clin Pathol 2003;119:811–5.
[8] Fukunga M, Kichuchi Y, Endo Y, et al. Case report: calcifying fibrous pseudotumor. Pathol Int 1997;47:60–3.
[9] Hill KA, Gonzalez-Crussi F, Chou PM. Calcifying fibrous pseudotumor versus inflammatory myofibroblastic tumor: a histological and immunohistochemical comparison. Mod Pathol 2001;14:784–90.
[10] Larson BK, Balzer B, Goldwasser J, et al. Calcifying fibrous tumor: an unrecognized IgG4 related disease. APMIS 2014;123:72–6.
[11] Kuo TT, Chen TC, Lee LY. Sclerosing angiomatoid nodular transformation of the spleen (SANT): clinicopathological study of 10 cases with or without abdominal disseminated calcifying fibrous tumors, and the presence of a significant number of IgG4+ plasma cells. Pathol Int 2009;59:844–50.
[12] Erica G, Prohaska , Andrew P, et al. Retroperitoneal calcifying fibrous tumor mimicking an adrenal tumor. J Surg Case Rep 2016;6:1–4.
[13] Tseng IT, Chen ST, Huang ZZ, et al. Multiple calcyfying fibrous tumors in the small intestine and mesentery. J Formos Med Assoc 2012;45:33–6.
[14] Puccio F, Solazzo M, Marciano P, et al. Laparoscopic resection of fibrous pseudotumor of the gastric wall. A unique case report. Surg Endosc 2001;15:1227.
[15] Ge X, Liao J, Choo RJ, et al. Solitary fibrous tumor of the ilium. A case report. Medicine 2017;96:e9355.