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Case Report

Calcifying Fibrous Tumor Causing Small Bowel Obstruction - Case Report

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Abstract

Background: Calcifying Fibrous (pseudo) Tumor (CFT) is a rare, benign, mesenchymal lesion that has been documented throughout the gastrointestinal tract and may be associated with gastric ulcers, obstruction, intussusception, and volvulus but not short bowel syndrome. Here, we describe the first case.

Case presentation: A 77-year-old male with a history of exploratory laparotomy for an "infection" of the abdomen 10 years ago presented to the emergency department with abdominal pain, bloating, nausea, and vomiting for 4 days. He described the abdominal pain as constant, moderate in severity, diffuse, and without known triggers. He admitted to constipation, shortness of breath, dizziness, and chest heaviness. He denied diarrhea, fever, and weight loss. Computed tomography of the abdomen/pelvis with contrast showed partial bowel obstruction. The patient underwent exploratory laparotomy. Intraoperatively, extensive adhesions from previous laparotomy were found along with a large mass. There were multiple loops of small bowel, which was resected followed by an enteroenterostomy. Pathology report supported CFT given it was positive for Factor XIIIa, CD34, vimentin and negative for nuclear beta catenin and anaplastic lymphoma kinase-1. Postoperative course was complicated by acute kidney injury and *Pseudomonas Aeruginosa* and *Candida Albican* infections.

Conclusion: CFT is a rare, benign lesion that is not known to be associated with short bowel syndrome. Our case illustrates that short bowel syndrome is inevitable in the setting of extensive small bowel involvement with the CFT, who previously had extensive bowel resection.

Keywords: Calcifying fibrous tumor; Short bowel syndrome

Introduction

The phenomenon of Calcifying Fibrous Tumor (CFT) is rare. Since its first description in 1988, only 157 cases of CFT until 2015 have been reported internationally [1-4]. Originally, Rosennthal et al. [4] described CFT as a "fibrous tumor with psammomatous calcifications and lymphoplasmacytic cell infiltrate" in deep soft tissue of children. Overtime, it is now recognized as a rare benign lesion composed of abundant dense well circumscribed hyalinized collagen with lymphoplasmacytic infiltrate, spindle cells, lymphoid aggregates, and psammomatous or dystrophic calcifications [1]. The mean age at the time of diagnosis has been found to be 33.58 years with a female's predilection (ratio 1:1.23) [1]. CFT has been found at various anatomic locations including but not limited to the neck, heart, lung, mediastinum, spine, and most recently gastrointestinal tract from esophagus to large intestine with a distribution of stomach (18%), small intestine (8.7%), pleura (9.9%), mesentery (5%), and peritoneum (6.8%) [1-3]. The exact pathogenesis of CFT is unknown but may be secondary to infection, trauma, and IgG4-related disease [5]. Patients with CFTs can be asymptomatic or symptomatic with complaints of abdominal pain, nausea, vomiting, or bowel dysfunction, which are all non-specific [1,2]. Studies have found that further workup showed CFTs may be associated with gastric ulcers, obstruction, intussusception, and volvulus [2,5,6]. To date, there is no documented report of CFT involvement with multiple loops of small bowel that subsequently resulted in an enteroenterostomy and short bowel syndrome. The authors describe the first case of short bowel syndrome in a patient with CFT proximal to the ligament of Treitz who had undergone a laparotomy 10 years earlier.

Case Presentation

A 77-year-old male presented to the emergency department with abdominal pain, bloating, nausea, and vomiting for 4 days and constipation for the last month. He described the abdominal pain as constant, moderate in severity, diffuse, and without known triggers. He was prescribed colace. He had significant comorbidities of gout, chronic kidney disease, hypertension, hypothyroidism, psoriasis, and prostate issues and a past surgical history of prostate surgery and exploratory laparotomy bowel resection for an "infection" of the abdomen 10 years ago. At the initial presentation, he was following with his oncologist for a possible carcinoid tumor. He admitted to shortness of breath, dizziness, and chest heaviness. He denied diarrhea, fever, and weight loss. His vitals were a temperature of 97.5 F, heart rate of 59 beats/minute, respiratory rate of 18 breaths/minute, blood pressure of 132/59 mmHg, and body mass index of 25.16 kg/m^2. On abdominal examination, the abdomen was soft and mildly distended with minimal abdominal tenderness to palpation and hyperactive bowel sounds were heard. A well-healed midline scar extending from the upper abdomen to the suprapubic area was noted. A Computed Tomographic (CT) scan of the abdomen and pelvis showed fluid distended stomach, duodenum, and proximal jejunum with the rest of the small bowel of normal caliber with diffuse

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wall thickening supporting the diagnosis of proximal small bowel obstruction. Calcifications, retraction of the central mesentery, and previous surgical sutures were also noted. No obstructive uropathy, free air, free fluid, or diverticulitis was seen. Possible etiologies of the patient's small bowel obstruction included postsurgical change, adhesions, mesenteric retraction, neoplasm, fibrosis, and among others. A general surgery consult followed and the patient consented to laparotomy.

Surgery

Intraoperatively, the old midline abdominal scar from his previous laparotomy was excised. Extensive adhesions secondary to previous laparotomy were noted. Multiple small bowel loops were attached to the fascia and each other. Following the mobilization of the adhesions, a large mass was palpated in the mesentery suggestive of malignancy with extensive involvement of multiple loops of small bowel, so the small bowel was resected along with the mass (Figure 1). The exploration revealed a significant section of the small bowel had been resected in the previous laparotomy. The terminal ileum was mobilized and anastomosed to the other end of the small bowel distal to the ligament of Treitz. The enteroenterostomy was successful and the patient tolerated the procedure well.

Pathology

Two specimens of the small bowel were received. The 1st specimen weighed 174 grams and 17.5 cm long. The serosal surface was purple and with focal brown discolorations and the mucosal surface was pink-grey. The 2nd specimen was 16.0 cm × 14.0 cm × 7.5 cm weighing 750 gm with multiple serosal adhesions and tightly coiled bowel loops. During grossing, the bowel loops could not be separated from each other and the soft tissue due to marked adhesions; therefore, the specimen was serially sectioned and revealed a grey-yellowish, not well defined, firm lesion measuring 5.5 cm × 4.5 cm × 2.0 cm with adipose tissue firmly attached to multiple loops of bowel. Trichrome stain showed prominent fibrosis (Figure 2). Immunohistochemistry showed positivity for Factor XIIIa, CD34, vimentin, CD117, Smooth Muscle Actin (SMA) and desmin and negativity for nuclear beta catenin, anaplastic lymphoma kinase-1 (ALK-1), cytokertin AE1/AE3, DOG-1, and chromogranin. Given the above findings, the diagnosis of CFT was favored over other differential diagnoses of desmoid type fibromatosis and Inflammatory Myofibroblastic Tumor (IMT) (Figure 3). The bowel peripheral resections margins were negative for dysplasia or malignancy.

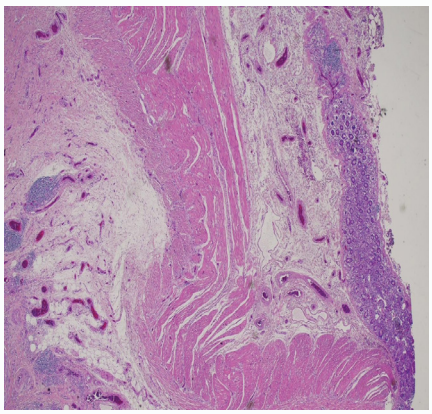

Figure 1: The bowel wall and adjacent calcifying fibrous pseudotumor tumor, 200X.

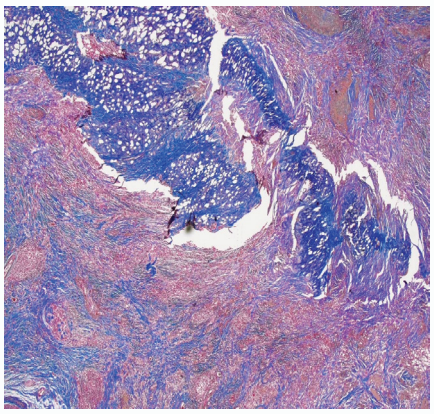

Figure 2: Trichrome stain highlights collagen fibers and spindle cell proliferation, 200X.

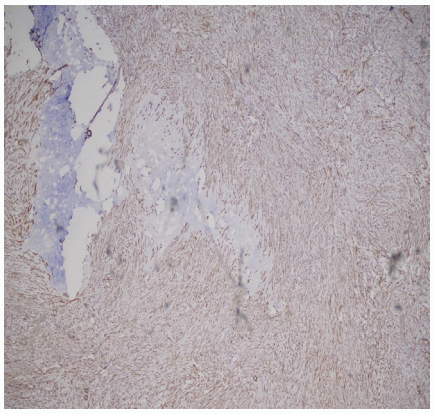

Figure 3: The myofibroblast stained with smooth muscle actin, 200X.

Post-operative course

The patient was started on total parenteral nutrition and antibiotic therapy. He showed signs of acute kidney injury likely secondary to acute tubular necrosis given hypotension noted intraoperatively with systolic blood pressure in the 80s. The patient also became febrile and cultures grew *P. Aeruginosa* which was treated with cefepime and *C. Albicans* infection treated with micafungin. The patient was discharged afebrile and in stable condition.

Discussion

CFT is a rare phenomenon and there is no clinical presentation that is specific to CFT [5]. CFT has been found in various anatomical locations including recently in the terminal ileum mesentery and it has been misdiagnosed as a thyroid nodule and Gastrointestinal Stromal Tumor (GIST) [1-3,7,8]. At the initial presentation, our patient was following with his oncologist for a possible carcinoid tumor. A study reported clear border with coarse calcification on ultrasound and peripheral hypoenhancement without central enhancement on contrast-enhanced ultrasound may be clues to help in arriving at the correct diagnosis of CFT [8]. Our patient's mass was not seen on ultrasound and CT abdomen/pelvis scans due to the extensive multiple loops of small bowel wrapping around it. While the pathogenesis of CFT remains unclear, infection and surgical intervention may be possible etiologies [5]. Thus, our patient's CFT may be secondary to the infection (patient unable to recall the specific pathogen) as well as the laparotomy he had 10 years ago.

The diagnosis of CFT is based on histology and biomarkers because clinical presentations and radiographic findings are nonspecific [9]. Histopathological findings may show calcifications-psammoma bodies, inflammatory cells, and dense hyalinized collagen and immunohistochemical findings may show positivity for vimentin, Factor XIIIa, CD34, or SMA (Figure 4) [1]. The diagnosis of CFT in our patient was made with biomarker studies. Immunohistochemical positivity for Factor XIIIa, CD34, and vimentin strongly supported the diagnosis of CFT while negativity for ALK-1 making IMT less likely as well as negativity for nuclear beta catenin making desmoid tumor less likely [5].

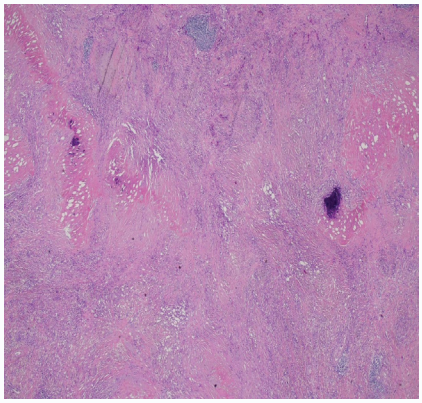

Figure 4: The dense hyalinized collagen of calcifying fibrous pseudotumor with psammomatous calcifications, 200X.

Intraoperatively, the mass was located distal to the ligament of Treitz and the cause of the patient's bowel obstruction and it felt like malignancy. Unfortunately, the multiple loops of the small bowel adhered to the CFT could not be salvaged and the decision to perform an enteroenterostomy was made. It is well-known that a sequel to laparotomy is abdominal adhesions, so the extensive small bowel adhesions to the CFT were more likely secondary to previous laparotomy rather than possibly induced by CFT [10]. Though CFT is a benign disease, the resection of the small bowel in this laparotomy combined with that from the previous laparotomy resulted in Short Bowel Syndrome (SBS). Our patient's enteroenterostomy was between the terminal ileum and the small bowel distal to the ligament of Treitz and he was started on total parenteral nutrition post-surgery.

The clinical outcome is usually favorable for CFT because it is a benign lesion that is cured by local surgical resection with a recurrence rate of approximately 10% [5,8]. No deaths have been directly attributed to CFT [5]. On the other hand, the quality of life for a patient with short bowel syndrome is poorer when compared to that in the general population [11]. Thus, a multidisciplinary care is important in the management of short bowel syndrome to address the physical, psychosocial and financial aspects associated with this medical condition.

Conclusion

Calcifying fibrous (pseudo) tumor is a rare phenomenon. Here, the authors describe the first case in which CFT is associated with short bowel syndrome secondary to extensive abdominal adhesions from previous laparotomy, resection of small bowel, and enteroenterostomy. Patients with short bowel syndrome are more likely than the general population to have poor quality of life and should be managed by a multidisciplinary team.

Author Contributions

ASW and LW drafted the original manuscript. SN, JP, MC, and FBD critically reviewed the manuscript. FBD conceptualized and supervised the project.

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