A Ball in the Belly: Calcifying Fibrous Pseudotumor of Abdominal Cavity – A Rare Entity

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Abstract
Calcifying fibrous pseudotumor is a rare clinically benign tumor first reported by Rosenthal and Abdul-Karim in 1988. Although rare, the tumor occurs most commonly in children and adolescents. Here we present a case of a 47-year-old male who reported to our institution with vague complaints of intermittent abdominal discomfort which was of mild to severe intensity, diffuse in presentation for a duration of 6 months. The pain was associated with subjective feeling of a “Wandering Lump” in the lower abdomen. This case report presents how this entity was managed along with review of literature. The novelty in this case report was it was a very peculiar case in the sense of its vague presentation, as the patient complained that he feels a lump and discomfort in right lower quadrant of his abdomen in the morning which moves up in the epigastric region; by evening, it comes to lie in the right lower quadrant of abdomen. So, anybody can discard this vague presentation, but in this case, we showed very high index of suspicion and evaluated him. The laparotomy confirmed its vague but true presentation as the lump was actually a freely lying ball which was wandering freely in the abdomen probably by peristalsis and patient’s routine activities. Surgical specialists are therefore advised never to miss such vague presentations, and index of suspicion has to be kept at all times.

Introduction
Calcifying fibrous pseudotumor (CFP) is a rare clinically benign tumor first reported by Rosenthal and Abdul-Karim [1]. The tumor occurs most commonly in children and adolescents [2]. The tumor has a broad anatomic variability in its distribution including the superficial and deep soft tissues, neck, pleura [3] and less commonly gastrointestinal tract [4], adrenal gland [5], and lungs [6]. However, one rare case report of myocardial calcifying fibrous tumor had also been reported. We describe a case of abdominal cavity CFP which presented with features of abdominal discomfort. The tumor has variable spectrum of clinical presentation depending upon the site. It can be present in specific site for a variable time span and unlikely to attract attention unless it manifests itself...
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symptomatically or clinically; the chances of malignant transformation are not noted in literature so far. It has unique histological and immunophenotypical features comprising dense, well-circumscribed mass consisting of hyalinized, paucicellular lamellar collagen with abundance of lymphoplasmacytic infiltrates, bland spindle cells, lymphoid aggregates, and psammomatous or dystrophic calcifications. This histological overlapping with other mesenchymal tumors makes the diagnosis of CFT challenging. Etiopathogenesis of the tumor is unclear, but it has been postulated that CFT may represent a sclerosing end stage of inflammatory myofibroblastic tumor; Larson et al. [7] suggested that CFT could be an unrecognized lesion with IgG4-related disease; Mehrad et al. [8] found copy number losses on chromosome 8 and deleterious mutations in ZN717, FRG1, and CDC27 genes, all of which are novel findings for studies of CFTP tumorigenesis. However, it still continues to evolve. The prognosis is good if complete resection is done.

Case Report

Here we present a case of a 47-year-old male who reported to our institution with vague complaints of intermittent abdominal discomfort which was of mild to severe intensity, diffuse in presentation for a duration of 6 months. The pain was associated with subjective feeling of a “Wandering Lump” in the lower abdomen. It was not associated with any constitutional symptoms. On clinical examination, per abdomen patient had a firm, nontender, oval lump of 10 cm × 8 cm in left iliac fossa. The overlying surface was smooth, margins were well demarcated, and the lump was mobile in all directions. USG abdomen revealed 11 cm × 9 cm round, predominantly pelvic lesion exhibiting extensive reverberation artifacts and provided a differential diagnosis of dermoid, teratoma, or fatty lesion (Fig. 1). Plain CT scan of the abdomen showed a round, well-circumscribed, hyperdense mass with a maximum cross-sectional area of 11 cm in the left iliac fossa with mean HU 436 with central core most likely mesenteric cyst, dermoid (Fig. 2).

Hematological and biochemical evaluation were within normal limits. After complete work up, written and informed consent was taken from patient for surgery and anesthesia. The abdomen was painted with 10% betadine solution and prepped from subcostal margins to groin. The abdominal cavity was exposed using standard midline approach (Fig. 3). Intraoperative findings revealed solid, spherical pearly white mass approximately 10 cm in diameter lying freely in the peritoneal cavity in the left iliac fossa; the lump was retrieved subsequently (Fig. 4). Thick fibrous adhesions in left supracolic compartment were present without mesenteric lymph nodes or ascites.

Postoperative period was uneventful. Patient was started on soft diet on postoperative evening and subsequently was discharged after 4 days. Patient was on regular follow-up for 3 years. Our patient has been pain free since then and was monitored by imaging surveillance which was normal.

Discussion

CFT is an extremely rare, benign lesion, which shows a predilection for subcutaneous and deep soft tissue. They are histologically distinct and characterized by the presence of hypocellular, densely hyalinized collagen with psammomatous or dystrophic calcification and mononuclear inflammatory infiltrate. They were first described in
1988 by Rosenthal and Abdul Karim [1] as a “childhood fibrous tumor with psammoma bodies” in 2- and 11-year-old girls in peripheral axial soft tissue. Fetch et al. [9] reported ten cases of a distinctive benign fibrous lesion and first used the term “calcifying fibrous pseudotumor.” The term “Pseudotumor” was used to reflect the belief that the underlying process was most likely fibro-inflammatory and reactive. However, Nascimento et al. [10] suggested CFP to be a true neoplasm with a tendency for local recurrence. Subsequently, the World Health Organization established the name for this lesion in 2002 as “Calcifying Fibrous Tumors” in the newly published classification of tumors of soft tissue and bone [11]. Calcifying fibrous tumor seems to have a female predilection (ratio 1:1.27). In a review of the 157 cases reported internationally, there is an estimated 1 case of abdominal CFT per year worldwide [12]. Abdominal CFTs are proposed to be the late, “burned out” manifestation of inflammatory myofibroblastic tumors. Abdominal CFT can become symptomatic and present as a surgical emergency mimicking commonest causes like acute appendicitis, torsion of mesenteric or ovarian cyst. Age distribution seems to be trimodal with one spike at 0–4 years, a second one in the mid-20s, and a third one in the mid-30s. This trimodal distribution may reflect different pathogenesis in the 3 different groups. Concerning the third spike (around thirties), there are many indications that CFT results as a late sclerosing stage of myofibroblastic tumor [13–16]. Trauma may also be the basis of pathogenesis of the second spike [16]. Concerning the early first spike, most acceptable contributors are genetic and/or embryologic factors. Van Dorpe et al. [17] and Chen [14] had reported series of familial multifocal abdominal cavity CFPs. These unique occurrences of familial peritoneal multifocal CFT also suggest that there may be a genetic susceptibility to this disorder, although chance and common environmental etiologic factors cannot be excluded.

The novelty in this case report was it was very peculiar case in the sense of its vague presentation, as the patient complained that he feels a lump and discomfort in right lower quadrant of his abdomen in the morning which moves up in the epigastric region; by evening, it comes to lie in the right lower quadrant of abdomen. So, anybody can discard this vague presentation, but in this case, we showed very high index of suspicion and evaluated him. The laparotomy confirmed its vague but true presentation as the lump was actually a freely lying ball which was wandering freely in the abdomen probably by peristalsis and patient’s routine activities. Surgical specialists are therefore advised never to miss such vague presentations, and index of suspicion has to be kept at all times.

**Conclusion**

CFTs are extraordinary rare, benign distinct soft tissue tumors of uncertain histogenesis with typical histological appearance. Our review of 57 previously reported cases suggests that a good outcome is expected when diagnosis of CFP is made and complete resection with margin of
normal tissue is done. The chances of local recurrence are almost nil. In our opinion, pathologists and surgeons should be made aware of multiple CFPs, since it is possible to confuse CFPs macroscopically with disseminated metastatic sarcomas or diffuse malignant mesotheliomas during surgery.

Statement of Ethics

This study protocol was reviewed and approved by the Institutional Ethical Committee with approval decision number January 2021 dated June 1, 2021. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Data Availability Statement

All data generated or analyzed during this study case report are included in this article. Further inquiries can be directed to the corresponding author.