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Calcifying Fibrous Tumor of Peritoneum in a Case of Ulcerative Colitis: An Unusual Coexistence

KEYWORDS: Calcifying; Dystrophic; Peritoneum; Ulcerative.

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ABSTRACT *Calcifying fibrous tumor is a rare, benign mesenchymal tumor of children and young adults, usually occurring within the subcutaneous and deep soft tissues of the extremities, trunk, neck, inguinal and scrotal region. Its occurrence in peritoneum is rare. Here, we report a case of calcifying fibrous tumor, which incidentally presented as peritoneal mass at the right hepatic flexure of colon in a 30-year-old female who underwent laparotomy for ulcerative colitis and was operated for the same. The histopathological and immunohistochemical findings of the peritoneal mass were consistent with calcifying fibrous tumor, comprising of spindled fibroblasts, abundant hyalinized collagen, several foci of dystrophic and psammomatous calcifications with focal lympho-plasmacytic inflammation and positive vimentin and factor XIIIa immunoexpression.*

INTRODUCTION:

Calcifying fibrous tumor (CFT), originally reported as a pediatric fibrous tumor with psammoma bodies, is a rare benign soft tissue tumor resulting from reactive fibro-inflammatory process[1]. CFT have been uncommonly documented in peritoneum and its coexistence with ulcerative colitis is rarely reported in literature. Here, we report a case of CFT, which incidentally presented as peritoneal mass at the right hepatic flexure of colon in a 30-year-old female who underwent laparotomy for ulcerative colitis.

CASE PRESENTATION:

A 30-yr- old female presented with the complaints of pain in abdomen, loose stools with passage of blood and mucus and fever off and on from the past 1 year. Stool examination of the patient showed presence of occult blood. A lower gastrointestinal (G.I.) endoscopic biopsy was suggestive of ulcerative colitis involving rectum and sigmoid colon. However, the patient was unresponsive to medical treatment and was taken for procto-colectomy. On laparotomy, an incidental peritoneal mass was identified near the hepatic flexure of colon that was adherent to the colonic mesentry and was found to be arising from the covering peritoneum. The mass was excised and submitted for the final diagnosis.

Grossly, the excised mass was well-circumscribed and globular, measuring 3.5 x 2 x 2 cm with attached fat. The consistency of the mass was firm to hard. Cut-section revealed presence of cream-grey heterogenous areas with whorled appearance (Figure1a). Histopathological examination of Hematoxylin and eosin (H&E) stained sections from the mass showed spindled fibroblastic cells in an irregular fascicular pattern with areas of abundant hyalinized collagen and dystrophic and psammomatous calcification and focal lymphoplasmocytic inflammatory infiltrate, consistent with CFT (Figures 1b,1c&1d) No necrosis or mitosis was appreciated. Immunohistochemically, the spindle cells of the CFT showed cytoplasmic immunopositivity to vimentin (figure 2a) and factor XIIIa (figure 2b). Smooth muscle actin (SMA), desmin, cytokeratin, S-100, CD-117, CD-34 and Anaplastic lymphoma kinase-1 (ALK-1) were found to be non-reactive. Congo red staining of the tumor section was negative. Post-operative recovery of the patient was uneventful. No recurrence have been observed during the last 6 months of follow-up of the patient.

DISCUSSION:

The findings in the present case were diagnostic of calcifying fibrous tumor (CFT). Though CFT has been reported to occur in coexistence with other diseases like castleman's disease or as an incidental finding at cholecystectomy, its coexistence with ulcerative colitis is not documented in the literature[2, 3]. The cause and pathogenesis of CFT is not fully known, though initially it was postulated as a reactive fibro-inflammatory process as a result of response to tissue injury[1]. Although, rare familial occurrence of peritoneal CFT have been reported, the histological evidence of presence of lymphoid and plasma cells infiltrate in CFT and its recently documented association with IgG4-related chronic inflammatory or sclerosing diseases represents an unregulated and exaggerated immune response[4]. CFT may, thus, be proposed as another manifestation of immune mediated disease in patients with ulcerative colitis, which need further evaluation and studies to confirm this association.

The differential diagnoses in the present case included an inflammatory myofibroblastic tumor (IMT), calcifying aponeurotic fibroma, nodular fasciitis, calcifying granulomas, amyloidoma, reactive nodular fibrosing pseudotumor, solitary fibrous tumor (SFT) and desmoids fibromatosis.

CFT was previously proposed to represent the sclerosing end stage of inflammatory myofibroblastic tumor (IMT) but recently distinct differences have been documented between these two different entities[5]. IMT do not show dystrophic or psammomatous calcification and demonstrates immunohistochemical positivity for SMA, ALK-1 (Anaplastic lymphoma kinase-1) and variable positivity for CD34 in contrast to the present case which showed negative SMA, ALK-1 and CD-34 immunoexpression[5].

Calcifying aponeurotic fibroma shows band-like calcification, catilagenous metaplasia and multinucleated gi-

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ant cells, which were absent in the present case. Nodular fasciitis displays tissue culture-like spindle cells in myxoid stroma and lacks calcification. Calcified granulomas usually show residual histiocytes and multinucleated giant cells. Amyloid fibroma show positive staining with Congo red and was thus excluded.

Reactive nodular fibrosing pseudotumor shares some overlapping features with CFT but it lacks calcification and shows SMA immunopositivity[6]. Similarly, solitary fibrous tumor (SFT) may present as peritoneal mass but lacks prominent calcification and show consistent CD-34 immunopositivity[7].

Intrabdominal desmoid fibromatosis shows infiltration of surrounding tissues and positive SMA immunoreactivity and lacks calcification[8].

Complete surgical excision is the curative treatment for CFT with only rare cases reported to show recurrence so far.

CONCLUSION:

CFT can occur in association with inflammatory bowel disease (IBD) and may be missed during work-up, only to be identified incidentally. Therefore, the possibility of CFT should always be considered in patients of IBD, not responding to usual treatment. This coexistence may represent an unregulated and exaggerated immune response in the patients with ulcerative colitis. The diagnosis of CFT should be concluded after appropriate histological and immunohistochemical work-up to distinguish it from closely resembling tumors.

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FIGURES CAPTIONS:

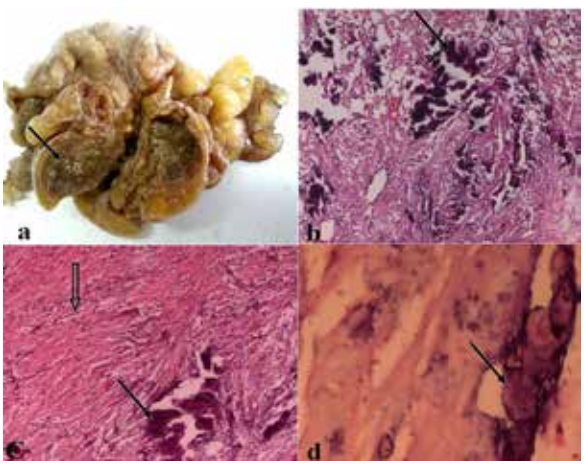

Figure 1: a) Gross specimen showing globular mass with attached fat with cream-grey heterogenous areas with whorled appearance (arrow), b) and c) showing areas of dystrophic calcifications (arrow) with abundant fibro-collagenized tissue (double arrow), d) showing psammomatous calcification (arrow).

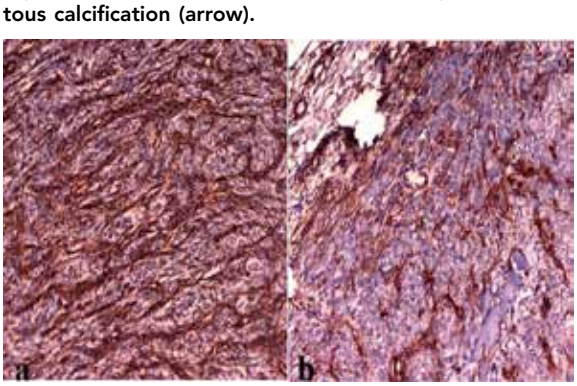

Figure 2: a) and b) Showing positivity of spindled tumor cells to vimentin and factor XIIIa respectively.