

## Case Report

# Calcifying fibrous tumour: a rare case report of an exceptional lesion localized in retroperitoneum, mesentery and pelvis

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### ABSTRACT

Calcifying fibrous tumours are rare benign lesions affecting mostly children and young adults. A 17-year-old female presented with abdominal pain and abdominal distention. Physical examination revealed intra-abdominal mass occupying retro peritoneum and right iliac fossa. Excisional biopsy from peritoneum and mesentery were performed. Histopathologically, it was composed of hypocellular hyalinised collagenized stroma, spindle cells, psammomatous and dystrophic calcification and mononuclear inflammatory cell infiltration. Authors are here in reporting a case of calcifying fibrous tumour and discussing its clinical and morphological features with regard to the literature.

**Keywords:** Benign mesenchymal lesion, Calcifying fibrous tumour, Calcification, Immunohistochemistry, Retroperitoneum

## INTRODUCTION

Calcifying Fibrous Tumours (CFT) are very rare benign mesenchymal lesion. It was described for the first time in 1988 by Rosenthal et al, as a "Childhood fibrous tumour with psammoma bodies" in peripheral axial soft tissue and in 1993, Fetsch et al, re-named it as calcifying fibrous pseudotumour.<sup>1</sup>

"Pseudotumour" was used to reflect belief that the underlying process was most likely inflammatory.<sup>2</sup> However, Nascimento et al, suggested a true neoplasm with a tendency for nondestructive local recurrence.<sup>3</sup> The term "Calcifying Fibrous Tumour" was established in the year 2002 in accordance with World Health Organization (WHO) in the newly published classification of tumours of soft tissue and bone.<sup>4</sup>

Authors here in present a rare case of CFT in the retroperitoneum, mesentery and pelvis and discuss its clinical and morphological features with regard to the literature.

## CASE REPORT

A 19 year old female patient presented to this institute in March 2019 with complaint of abdominal pain and abdominal distention since 1 month. Abdominal pain was dull aching in nature with no relation to intake of meals. Abdominal distention was gradual. There was no comorbidities and no history of previous abdominal surgery. Physical examination revealed a young thin built lady with approximately 31x25 cm intra-abdominal mass occupying retroperitoneum and right iliac fossa.

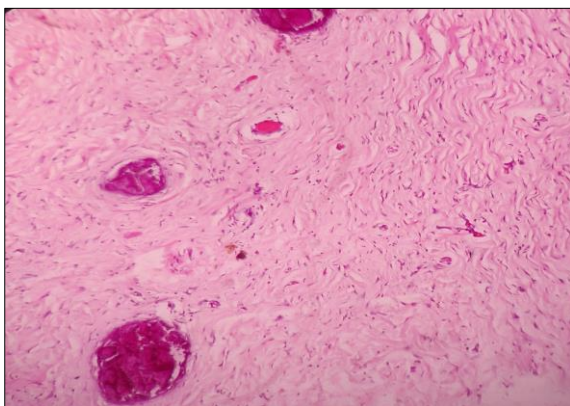
MRI scan was obtained. It showed multiple large well defined lobulated mass in retroperitoneum, mesentery, right iliac fossa and right side of pelvis. Findings were not specific for single etiology and the likely differential diagnosis were mesenteric fibromatosis, small bowel neoplastic pathology with mesenteric lymphnodal mass and mesenteric/retroperitoneal neoplastic lesion.

Biopsy from peritoneum and mesentery were performed for histopathological diagnosis.

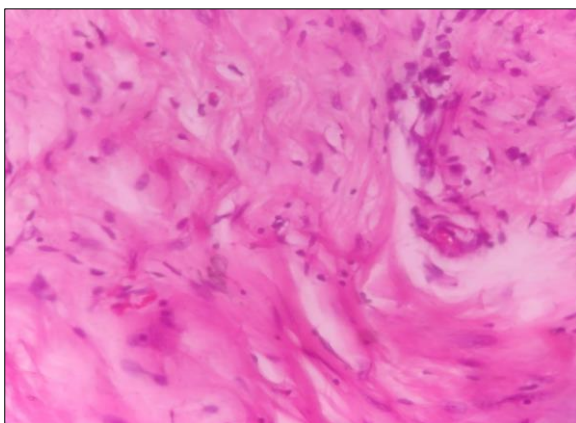
Authors received two soft tissue specimens, one from peritoneum and other from mesentery approximately measuring 3x2x0.8 cm and 3.2x2.5x1 cm in size respectively and firm in consistency. External surface was nodular and brownish-grey in colour. Cut surface was whitish-yellow in colour.

The sections showed predominantly hypocellular hyalinised collagenized stroma with interspersed calcification, chronic inflammatory infiltrate and dilated vascular channels (Figure 1). Collagenous matrix contained bland spindle cells, elongated nuclei, fine nuclear chromatin and inconspicuous nucleoli with scant cytoplasm (Figure 2). Calcification was psammomatous and dystrophic (Figure 3). The inflammatory component was predominantly composed of lymphocytic and plasma cells scattered singly and at places forming aggregates. Scattered mast cells and occasional giant cells were also seen.

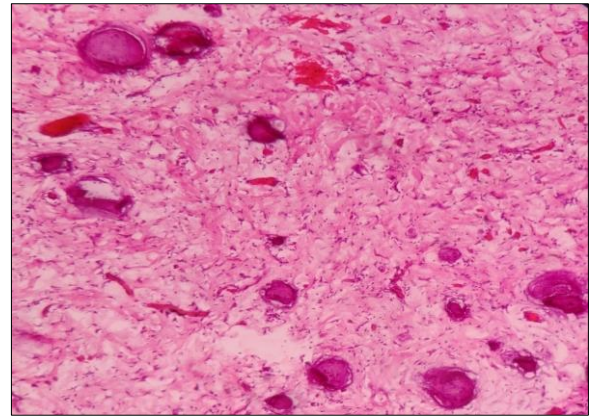
Nuclear atypia, mitosis and necrosis were not seen. Immunohistochemically, spindle cells were immunoreactive for Vimentin (Figure 4) and CD 34 and immunonegative for Desmin, S-100 and CD 117. The lesion was diagnosed as Calcifying Fibrous Tumour.



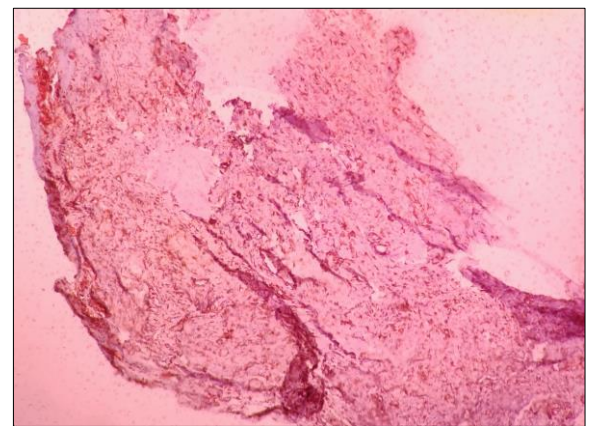
**Figure 1: Hypocellular hyalinised collagenized stroma (H and E stain, 10X).**



**Figure 2: Collagenous matrix containing bland spindle cells (H and E stain, 40X).**



**Figure 3: Abundant Psammomatous and Dystrophic Calcification (H and E stain, 10X).**



**Figure 4: Spindle cells showing immunoreactivity for Vimentin (Immunohistochemistry).**

## DISCUSSION

CFTs are an interesting cases to investigate due to their rare occurrence, unclear etiology and the possibility to occur in nearly every location of the human body and mimic other diseases depending on the location and proximity to other organs.

Calcifying fibrous tumour (CFT) is a rarely seen entity that usually occurs in children and young adults, with a slight increase in risk in women.<sup>5</sup> In the overview of Chorti and Theodossis, there are 3 life-span peaks of occurrence named early childhood, mid-20s and mid-30s.<sup>4</sup> The early childhood peak could represent a genetic or embryologic pathogenical pathway. The second peak could be vaguely associated with a trauma. Concerning the third spike, there are many indications that CFTs are an end-stage presentation of inflammatory myofibroblastic tumour (IMT).

Clinically, it appears as a slowly growing, painless mass in subcutaneous/deep soft tissue which may be associated with systemic symptoms. The lesions most commonly arise in the extremities followed by trunk, inguinal and

scrotal regions, and head and neck area. However, these tumours were gradually reported in other rare areas like visceral regions (pleura, omentum/mesentery and peritoneum) as multifocal lesions, the oral cavity, and the adrenal gland. Very few cases arising from the retroperitoneum have been reported.<sup>5-7</sup> Though unifocal in the vast majority of cases, in a mesenteric presentation the tumour was often observed multifocal.<sup>4</sup>

In this case, it was young female with a very large mass arising from a very rare site - retroperitoneum, mesentery as well as in right side of iliac fossa and pelvis. Authors have no explanation for this tendency of multifocality in this case.

Radiographically, it is non-calcified and well circumscribed. Punctate, thick or band-like calcifications can be seen on Computed Tomography (CT). On MRI, the tumour may be similar to fibromatoses.<sup>7</sup>

Macroscopically, the diameter ranges from 1 to 15 cm. The mass of CFT is well-circumscribed, unencapsulated, lobulated and solid/firm. Uniform greyish white fibrous appearance is appreciated on cut section. It often cuts with gritty sensation due to extensive calcification. In some cases the tumour may have indistinct boundaries; hence infiltration may be indistinguishable from the surrounding tissues.<sup>5,7</sup>

Histopathologically, CFT is characterized by the following 3 components:

- Abundant, paucicellular, hyalinized collagen or birefringent fibrosclerotic tissue;
- Interspersed calcifications; and
- An inflammatory infiltrate.

The collagenous matrix is hyalinized and often exhibits a whorled or storiform pattern but may be haphazard or pattern less. Bland spindle cells are embedded within the abundant collagen. The fibroblastic/myofibroblastic spindle cells are scattered cytologically bland and exhibit ovoid, vesicular nuclei with fine chromatin and inconspicuous nucleoli and abundant eosinophilic to amphophilic cytoplasm. Atypia and mitotic figures are lacking. The calcified component, dispersed throughout the fibrotic areas, may be either psammomatous or dystrophic. The inflammatory component is predominantly composed of lymphocytes and plasma cells infiltrating singly or forming aggregates.<sup>5,8</sup> Although immunohistochemical analysis is not necessary for diagnosis, fibroblasts do express vimentin, CD34 and factor XIIIa.<sup>7</sup> Immunoreactivity is variable in muscle specific actin, smooth muscle actin and desmin. But, ALK stain is consistently negative.<sup>5,7</sup>

The therapy of CFTs is surgical removal. Prognosis of CFT after removal is excellent, few recurrences and no deaths related to CFT are reported.<sup>4</sup>

### ***Differential Diagnosis***

The histopathological features of CFTs are generally easily recognizable from other reactive or benign neoplastic lesions. Inflammatory myofibroblastic tumour (IMT), retroperitoneal fibrosis, reactive nodular fibrous pseudotumour (RNFP), nodular fasciitis, desmoid fibromatosis, fibroma of tendon sheath and calcifying aponeurotic fibroma might be considered in the differential diagnosis<sup>(5,6)</sup>.

### ***Inflammatory myofibroblastic tumour (IMT)***

This is more cellular and shows less hyalinization than CFT. It is composed of spindled myofibroblasts, fibroblasts and inflammatory cells. It typically lacks calcification or occasional stromal calcification is seen in addition to a polymorphic inflammatory cell infiltration. Actin and ALK immunoreexpression is positive in IMT.<sup>5,9</sup> In fact, some authors consider CFT to be a late sclerosing stage of IMT.<sup>7</sup>

### ***Retroperitoneal fibrosis***

Retroperitoneal fibrosis can also be considered as a differential diagnosis for the tumour in retroperitoneal location specially when associated with other fibrosing conditions, such as inflammatory pseudotumours or retractile mesenteries. But in these lesions, there are no psammomatous calcifications, as seen in this case.

### ***Reactive nodular fibrous pseudotumour (RNFP)***

RNFP is less cellular than CFT. Actin, desmin and CD117 immunoreexpression is positive and CD34 is negative in addition to the characteristic histopathological features of CFT.

### ***Desmoid fibromatosis***

Desmoid fibromatosis is less well-circumscribed and more cellular with arrangement in a prominent fascicular growth pattern. Histopathologically, spindle cells typically infiltrate the surrounding soft tissue and microcalcification is extremely uncommon.

### **CONCLUSION**

In summary, Calcifying Fibrous Tumours are rare benign neoplasm with a huge lesion arising from the retroperitoneum as well as in mesentery as described above is extremely uncommon. Though complete surgical excision is the treatment of choice, excellent prognosis after surgical excision and rare local recurrence, in this case patient died due to large sized tumour that may have severely impaired the function of the adjacent organs. Authors are reporting this rare case as lesions like these may be identified more often in the future due to increased awareness and technical advances in diagnosis.

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