

Solitary Calcifying Fibrous Adenoma of Small Bowel Mesentery - A Rare Case Report

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Abstract: Solitary fibrous tumours are rare mesenchymal tumours that typically arise from the pleura and rarely originate from the mesentery. We herein report a case involving a 24-year-old patient who presented with pain in right iliac fossa, diagnosed as a case of acute appendicitis. Patient underwent an emergency open appendectomy. This mass was found incidentally during an exploration for Meckel's diverticulum as a routine protocol during appendectomy which was resected. The biopsy of the mass revealed that it was a solitary calcifying fibrous tumour originating from the mesentery of the small intestine. The patient was discharged 5 days after surgery and had an uneventful post-operative stay.

Keywords: Acute appendicitis, biopsy, case report, mesentery, pleura, right iliac fossa, solitary fibrous tumour

1. Introduction

Solitary fibrous tumours are a group of rare spindle cell mesenchymal tumours mainly arising from the thoracic pleura. These are rare ubiquitous tumours that originate from the mesenchyme and are characterized by high fibrosity and hypervascularity¹. Although abdominal solitary fibrous tumours have been reported, involvement of the mesentery is rare². Solitary fibrous tumours are usually benign and remain asymptomatic unless they place pressure on adjacent tissues, making them hard to detect. Both computed tomography (CT) and magnetic resonance imaging (MRI) can help to diagnose solitary fibrous tumours; however, a definitive diagnosis relies on immunohistopathology, in which STAT6 and CD34 are used as highly specific markers³. Macroscopically, most solitary fibrous tumours are well-circumscribed and frequently encapsulated masses. Solitary fibrous tumours are typically tan-white, firm and multinodular in appearance and may exhibit haemorrhagic and myxoid changes⁴. In some instances, solitary fibrous tumours may show malignant features and invasion of surrounding tissues⁵. Therefore, complete surgical resection with intact borders is recommended⁶. Because recurrence may occur even 20 years after treatment⁷, long-term surveillance may be beneficial. Although solitary fibrous tumours are well described in the literature, those arising from the mesentery are very rare. To the best of our knowledge, only 22 cases of primary intraperitoneal SFTs of the mesentery have been reported with an associated histopathological diagnosis^{2-4, 7-25}. Here, we report on an extremely rare case of a solitary calcifying fibrous adenoma of the mesentery.

2. Case History

A 24-year-old male presented in surgery outdoor department with complaints of pain in right iliac fossa for 3 days associated with fever with chills for last 3 days, vomiting 3-4 episodes for last 2 days. Patient had no significant past history. On clinical examination: patient was having pulse rate of 103/minutes and axillary temperature was 101.8°F, rest all vitals were within normal limits. Abdominal examination revealed that abdomen was soft, tenderness was present in right iliac fossa, guarding in right iliac fossa, without rigidity and bowel sounds were present. Rest all systemic examination was within normal limits. Investigations were done in which TLC was 15600/mm and other biochemical parameters were within normal limit. Ultrasonography of whole abdomen was done which showed a blind ended, elongated, non-compressible, non-peristaltic structure (approximately 11 mm diameter) was present in right iliac fossa which was suggestive of acute appendicitis.

Patient was planned for open appendectomy and taken for surgery. On exploration, skin incision given at McBurney's point, abdomen opened in layers, appendix identified, transfixed and excised. As a routine protocol, during exploration for Meckel's diverticulum, a swelling of 3×2 cm found 15 cm proximal to the ileo-caecal junction (Figure 1). Swelling was excised (Figure 2). Both specimens i. e. appendix and soft tissue swelling were sent for histopathological examination. On examination of soft tissue swelling grossly it was a single tan-white tissue measuring 3×2×1 cm, which was well circumscribed, firm and non-capsulated. Microscopic examination of the lesion showed that there was densely hyalinized and collagenous stroma with areas of calcification and scanty lymphoplasmacytic

infiltrate. At places of collagen fibres show whirling. Lymphoid follicles were also noted. There was no evidence of tuberculosis in the specimen. Specimen was suggestive of calcifying fibrous tumour of mesentery. Immunohistochemistry was not done due to financial constrain. Postoperative period as well as follow up till 2 months were uneventful.



Figure 1: Lesion found during exploration



Figure 2: Excision of mass

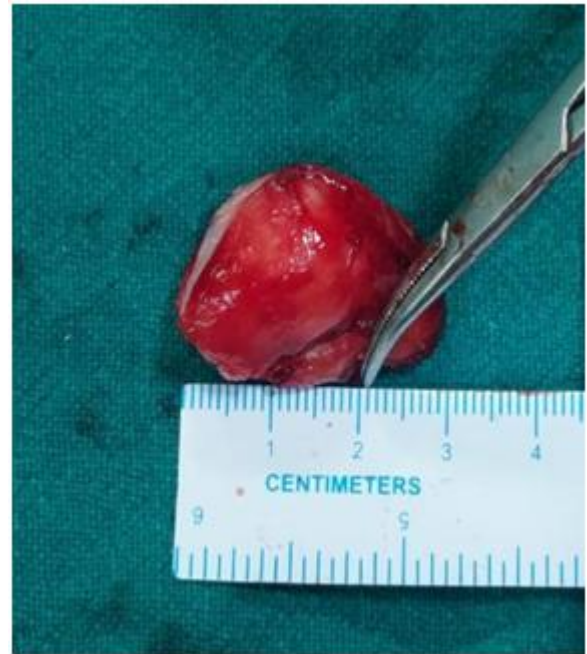


Figure 3: Excised mass from mesentery



Figure 4: Cut section of mass and appendix

3. Discussion

Calcifying solitary fibrous tumour, originally defined by Rosenthal and Abdul - Karim²⁶ as a childhood fibrous tumour with psammoma bodies, was subsequently renamed by Fetsch et al,²⁷ as a calcifying fibrous pseudotumor. They observed that this lesion was not limited to children; the calcifications were not always psammomatous and postulated that this lesion was a result of a reactive fibro-inflammatory process. Both the sexes are equally affected^{26, 27}.

Solitary fibrous tumours most frequently arise from the pleura, but they have also been observed in nearly every anatomical location of the human body²⁸. However, a solitary fibrous tumour arising from the mesentery is very rare⁷. Most were found in men, and the affected patients

median age of solitary fibrous tumours is 54 years (range, 26–82 years).

Calcifying fibrous tumour has been uncommonly documented in mesentery, including rare examples of multifocal involvement, wherein it may present as acute abdomen and even malignancy^{29, 30}. The diagnosis in the present case was achieved on biopsy.

These mesentery SFTs seem to be more commonly occur in the ileum. Based on published reports, it is clear that extra pleural solitary fibrous tumours most commonly occur in adults but have a wide age distribution. Solitary fibrous tumours are usually slowly growing and painless, with no clinical symptoms unless they compress adjacent tissue.⁷

CT and MRI are effective in the diagnosis of solitary fibrous tumours. Most tumours are round or lobulated with clear boundaries and the mass is often large and completely enveloped³¹. On multi - slice CT imaging, solitary fibrous tumours appear as heterogeneously enhancing, multiloculated soft tissue masses. Areas of dense or scattered calcification and central tubular or rounded hypoattenuating regions representing necrosis are also observed. Enhancement is most conspicuous in the arterial and early portal venous phases with washout of contrast in the delayed phase. However, if fibrous elements are predominant, contrast enhancement becomes marked in the delayed phases as well⁴. Multiple peripheral blood vessels were observed in the arterial phase in some cases. This feature is thought to be an important diagnostic factor for solitary fibrous tumours. Solitary fibrous tumours exhibit a heterogeneous appearance on both T1 - and T2 - weighted MRI, with the appearance of the solid areas ranging from isointense to hypointense relative to skeletal muscle on all sequences. In contrast, cystic areas appear hyperintense on T2 - weighted images. The contrast enhancement is similar to that seen on CT imaging.

Differential diagnoses of solitary fibrous tumours include other benign tumours involving the mesentery, such as desmoids, inflammatory pseudotumor, mesenteric fibromatosis and leiomyomas. Gastrointestinal stromal tumours may be identified by the presence of extensive foci of haemorrhage and necrosis. Malignant lesions of the mesentery that need to be excluded before achieving a diagnosis of solitary fibrous tumours include soft tissue sarcomas, leiomyosarcomas, lymphoma and metastases¹⁹.

The presence of rounded necrotic areas, calcifications, and the above - described enhancement pattern in the arterial, portal venous, and delayed phases aid in distinguishing between solitary fibrous tumours and other neoplasms arising from the mesentery.

The most effective therapeutic modality for Solitary fibrous tumour is surgical resection. The role of other treatment modalities in the management of solitary fibrous tumours unclear. Some reports in the literature have demonstrated the effectiveness of radiotherapy for the control of solitary fibrous tumors^{32, 33}. Chemotherapeutic drugs and novel targeted drugs seem to exert some activity on solitary fibrous tumors³⁴. However, a consensus has not been

achieved concerning the effectiveness of radiotherapy and chemotherapy against solitary fibrous tumours.

4. Conclusion

Solitary fibrous tumour of mesentery is very rare condition affecting mesentery and few cases have been reported in literature. However, solitary fibrous tumour is diagnosed histopathologically and radiologically. It usually doesn't require aggressive approach for treatment but if detected should be investigated thoroughly. Further studies may be done to find out some biochemical or pathological or radiological parameter for early diagnosis and aetiology of the disease.

Declaration

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Ethical approval: The study was approved by the Institutional Ethical Committee.

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