

# Solitary Fibrous Tumour of Liver: A Case Report and Literature Review

Gaurav Kaushik<sup>1</sup>, Madan Manmohan<sup>1</sup>

<sup>1</sup>Department of Radiodiagnosis, DY Patil School of Medicine, Navi Mumbai.

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## ABSTRACT

Solitary fibrous tumour is an uncommon mesenchymal neoplasm diagnosis of which is difficult because of non-specific clinical and radiological features. A 76 year old male presented with complaints of nausea and vomiting. Physical examination revealed firm mass in the right hypochondrium and epigastrium. CT scan revealed a heterogeneous, lobed and some vascularized mass of size 17.6 x 12.8 cm, situated in the right lobe. After surgery the mass was found to measure 19 cm x 13 cm x 10 cm. Histopathological examination found the tumour cells to be strongly positive for vimentin and CD34, confirming our diagnosis. Microscopic examination revealed haphazard arrangement of spindle cells and collagen bundles between the tumour cells with mild nuclear pleomorphism, and the mitotic count was 0-1/10 high power field. The post-operative course was uncomplicated and the patient was discharged home. After 16 months of follow up examinations, there was no recurrence of disease, local or distant metastasis and the patient remained asymptomatic. The clinical presentation and the radiological imaging can be very non-specific. Suggestive features on CT scan are a single, large, well-circumscribed, heterogeneously enhancing hepatic mass.

**Keywords:** Hepatic, immunohistochemistry, solitary fibrous tumor

## INTRODUCTION

Solitary fibrous tumour is an uncommon mesenchymal neoplasm affecting mainly the pleura and mediastinum. The other sites include the respiratory tract, orbit, peritoneum and central nervous system.<sup>[1]</sup> Usually, Solitary fibrous tumours are benign in nature, but 10-20% of them are malignant which have a tendency to metastasize. Pre-operative diagnosis of Solitary fibrous tumour of liver is difficult because of non-specific clinical and radiological features, which may delay the diagnosis.

## CASE REPORT

A 76 year old male with a medical history of hypertension and diabetes mellitus was referred to our department with a suspicion of liver mass. This suspected liver mass was discovered when the patient presented to our hospital with complaints of nausea and vomiting and underwent examination. There was no history of jaundice, loss of appetite, blood transfusion and weight loss. Physical

examination revealed firm mass in the right hypochondrium and epigastrium. There was no ascites, edema, signs of hepatic failure or cirrhosis stigmata. CT scan revealed a heterogeneous, lobed and some vascularized mass of size 17.6 x 12.8 cm, situated in the right lobe. Laboratory investigations, including liver biochemical profile and tumour markers like Carcinoembryonic antigen (CEA), alpha-fetoprotein, and CA19-9 levels, were within normal ranges. Hematological profile, kidney function, electrolytes and coagulation profile was within normal limits. Magnetic resonance imaging (MRI) and more advanced radiological investigations could not be performed due to financial difficulties of the patient. The patient underwent surgery for removal of the liver mass. On gross examination, the mass measured 19 cm x 13 cm x 10 cm and weighed 1450 gm. External surface was smooth with few congested vessels [Figure 1]. The adjacent liver parenchyma was unremarkable with no evidence of cirrhosis or fibrosis. Surgical resection margin was free of tumour grossly. The resected specimen was sent to department of Pathology for histopathological examination, the tumour cells were strongly positive for vimentin and CD34, confirming solitary fibrous tumour. Microscopic examination revealed haphazard arrangement of spindle cells and collagen bundles between the tumour cells with mild nuclear pleomorphism, and the mitotic count was 0-1/10

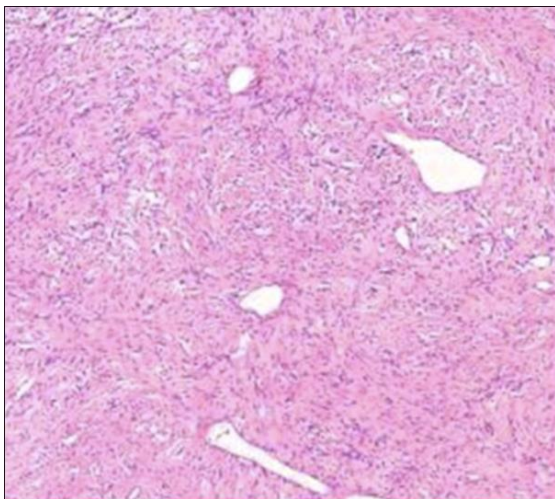
### Name & Address of Corresponding Author

Dr. Gaurav Kaushik  
Department of Radiodiagnosis  
DY Patil School of Medicine,  
Navi Mumbai.

high power field [Figure 2]. The post-operative course was uncomplicated and the patient was discharged home. After 16 months of follow up examinations, there was no recurrence of disease, local or distant metastasis and the patient remained asymptomatic.



**Figure 1: Contrast-enhanced CT scan showing a well-circumscribed mass in right hepatic lobe with widely varying CT attenuation**



**Figure 2. H & E stain showing arrangement of spindle cells surrounding ectatic branching blood vessels with interspersed bands of hyalinised collagen**

## DISCUSSION

Solitary fibrous tumour are rare neoplasms of mesenchymal origin and solitary fibrous tumour of the liver are very rare. Published literature has found twice higher incidence among females as compared to males and is most commonly found in ages of affected individuals ranging from 27 to 83 years.<sup>[2]</sup> Most patients are symptomatic, usually no-specific, as was our case. However, most of these symptoms can be attributed to mass effects of the tumour. Imaging studies are helpful in characterizing these lesions, as solitary fibrous tumours are usually single, well-circumscribed, heterogeneously enhancing, and highly vascular masses. A variety of differential diagnosis should be kept in mind, like

sarcomas, leiomyoma, sclerosed hemangioma, and inflammatory pseudotumour of the liver.<sup>[3]</sup>

Similar to the clinical presentation, radiological imaging of the tumour can also be very non-specific at times. Solitary fibrous tumour typically appears hypoechoic on ultrasonography but areas of myxoid degeneration may make it to appear heterogeneous.<sup>[4]</sup> On CT scan, they appear as a well-defined encapsulated mass with heterogeneous enhancement. Administering contrast may demonstrate a hypervascular tumour and progressive heterogeneous enhancement. CT scan helps to identify necrotic areas within the tumour and an outer pseudocapsule. In most cases, the tumour frequently displaces neighbouring organs, compresses adjacent arterial and venous vessels, and may cause obstruction and dilatation of the common bile duct.<sup>[5]</sup> Solitary fibrous tumour of the liver have low to intermediate signal intensity on T1-weighted images, and they have heterogeneous signal intensity on T2-weighted images. Fibrotic areas is shown by low T2 signal intensity.<sup>[6]</sup> At diffusion-weighted imaging, hypercellularity is indicated in the tumours by increased signal intensity and low apparent diffusion coefficient (ADC). Conversely, increased ADCs indicate a more fibrous tumour.<sup>[7]</sup> However, these findings are suggestive but not diagnostic of hepatic SFT.<sup>[8]</sup>

A definitive diagnosis of Solitary fibrous tumour is established by a core needle biopsy with histopathologic and immuno-histochemistry studies.<sup>[9]</sup> Phenotypes based on immuno-histochemistry include diffuse and strong CD34, BCL2, and STAT6 tumour cell expressions.<sup>[10]</sup> Positive CD34 marker expression can be positive in other tumours as well, but it remains highly characteristic of Solitary fibrous tumour, specially in the absence of expressions of other mesenchymal cell lineage phenotype markers. Moreover, STAT6 is a highly sensitive and specific biomarker of Solitary fibrous tumour, making it helpful to distinguish this tumour from other histologic mimics.

Solitary fibrous tumour of the liver must be considered potentially malignant in all cases, though it is benign in majority of cases. Surgical resection with free margins remains the standard treatment of Solitary fibrous tumour.<sup>8</sup> In cases where tumours are incompletely resected or are unresectable, radiation therapy, chemotherapy, and chemoembolization have been proposed with a multi-disciplinary approach.<sup>8</sup> Rarity of the tumour has not allowed researchers to conduct large sample studies to evaluate the prognosis and overall outcome of the tumour. Also, the best method for follow-up has not been well established, but whole-body CT is suggested in high-risk patients.<sup>[11]</sup>

## CONCLUSION

Solitary fibrous tumour arising from the liver is a rare entity, diagnosis of which can be a challenge. The clinical presentation and the radiological imaging can be very non-specific. Suggestive features on CT scan are a single, large, well-circumscribed, heterogeneously enhancing hepatic mass. Morphologically it may mimic other hepatic neoplasms, making histopathological diagnosis necessary.

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