

# Primary Omental Leiomyosarcoma: A Rare Case Report

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## Abstract

Primary leiomyosarcoma of greater omentum is rare and mimics gastrointestinal stromal tumor (GIST) due to their common origin. A 65 years old male presented with a large mass per abdomen of one year duration. Imaging findings were suggestive of extra-gastrointestinal stromal tumor (E-GIST) arising from greater omentum. The mass was excised. On microscopy, the diagnosis of leiomyosarcoma and E-GIST were considered. The tumor cells showed immunoreactivity for smooth muscle actin, muscle-specific actin, heavy-caldesmon and desmin and were immunonegative for CD117, DOG1 and CD34, hence confirming primary leiomyosarcoma of greater omentum. We present this case for its rarity.

**Keywords:** Extra-gastrointestinal stromal tumor, Leiomyosarcoma, Omentum.

## Introduction

Primary tumors of omentum are rare and are usually malignant [1]. Few cases of primary omental leiomyosarcoma have been reported in the literature. Most of these cases originated from the greater omentum. These tumors are generally seen in the

middle age [2,3]. These tumors can mimic other tumors like gastrointestinal stromal tumors (GIST) due to their common origin from mesenchymal stem cells [4]. Here we report a case of primary leiomyosarcoma arising from greater omentum in an elderly male.

## Case Presentation

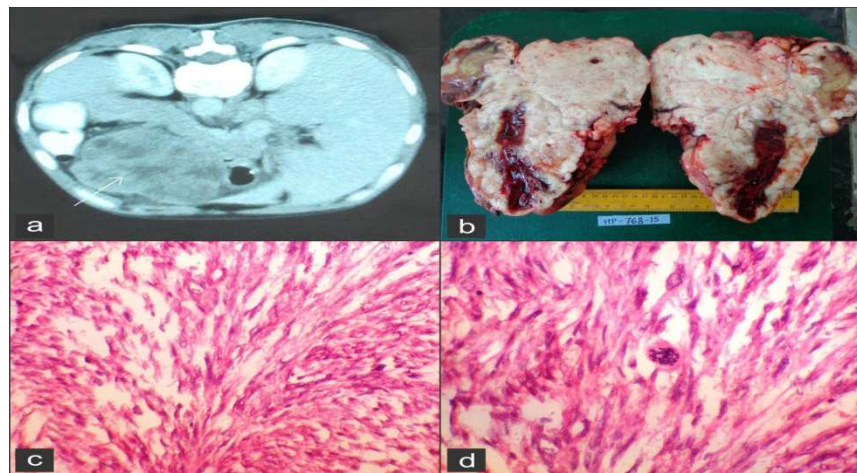
A 65 years old male presented with a large mass per abdomen of one year duration which was gradually increasing and was associated with pain. Patient also had history of significant weight loss and loss of appetite of six months duration. Patient complained of non-projectile vomiting of three days duration. On examination, the thinly built patient had a palpable abdominal mass measuring 21X19cms, involving the epigastric, umbilical, left hypochondriac and left lumbar region.

All hematological investigations were within normal limits and the patient was non-reactive for HIV I and HIV II antibodies. Chest X-ray was within normal limits. Ultrasonography of abdomen visualised a large well-defined hypoechoic mass but failed to determine the exact site of origin.

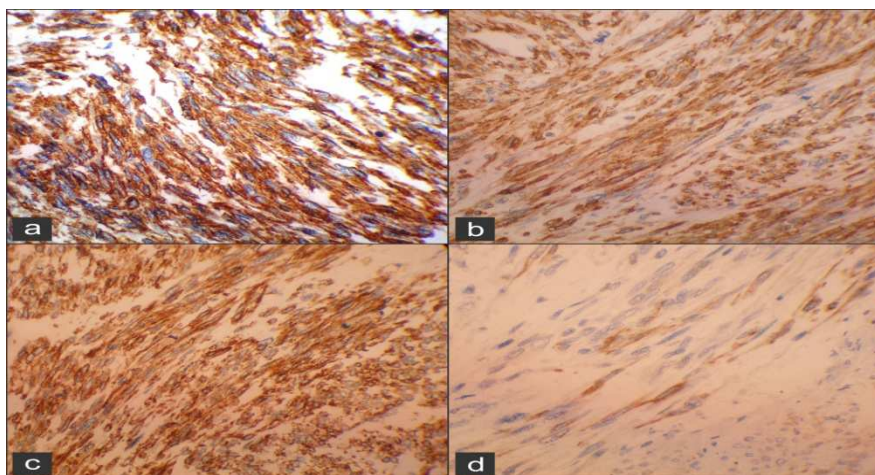
Computed tomography (CT) scan of abdomen showed a large lobulated mass measuring 28x20x18 cm attached to the greater omentum (Figure 1a). The mass showed heterogenous enhancement with multiple small necrotic areas. No obvious infiltration was seen in the pancreas, spleen and adjoining liver. There was no evidence of ascitis and pre-aortic or para-aortic lymphadenopathy. The imaging findings were of neoplastic lesion suggestive of extra-gastrointestinal stromal tumor (E-GIST).

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## Case Report



**Figure-1:** a) CT showing the omental mass (arrow) b) Cut section of leiomyosarcoma c) Microphotograph showing arrangement of tumor cells in fascicles (H and E, x100) d) Microphotograph showing individual tumor cells, few with atypical mitotic figures (H and E, x400).



**Figure-2:** Microphotograph showing tumor cells positive for a) smooth muscle actin (IHC, x400), b) muscle specific actin (IHC, x400), c) heavy-caldesmon (IHC, x400) d) focal positivity for desmin (IHC, x400).

Fine needle aspiration cytology of the mass was suggestive of smooth muscle origin of the tumor. The mass along with the omentum was excised. Rest of the intraabdominal organs were unremarkable.

Grossly the tumor was bosselated and measured 28x20x18cm with attached omentum. Cut surface was grey-white, nodular, firm in consistency with areas of haemorrhage and necrosis (Figure 1b). Few cystic areas were seen. On microscopy, the lesion was partly capsulated with tumor cells arranged in interlacing fascicles of varying sizes (Figure 1c). The individual cells were elongated to spindle shaped, with centrally located blunt ended or cigar-shaped nuclei and moderate amount of eosinophilic cytoplasm (Figure 1d). Atypical mitotic figures 14-16/10 HPF were seen in cellular areas. Areas of coagulative necrosis, hemorrhage and cystic change were also seen.

The rest of the omentum was unremarkable. A differential diagnosis of primary leiomyosarcoma of omentum and E-GIST were made. On immunohistochemistry (IHC), the tumor cells showed strong immunoreactivity for smooth muscle actin (SMA), muscle-specific actin, heavy-caldesmon and focal positivity for desmin (Figure 2). They were immunonegative for CD117 (c-kit), DOG1 and CD34. The final diagnosis of primary leiomyosarcoma of omentum grade 2 was made according to Federation Nationale de Centres de Lutte Contre Le Cancer (FNCLCC) system of grading. The patient had an uneventful postoperative course, and was free of recurrence and metastasis when last seen 18 months after his operation.

## Discussion

Leiomyosarcomas account for 10–20% of all soft tissue sarcomas. They most frequently arise from the uterus, gastrointestinal tract and retroperitoneal region [5]. It is said that smooth muscle tumors arise from mesodermal elements present in the blood vessels, fibrous tissues and the nerves of the omentum. In study done by Dixon et al on omental tumours, nearly 33% were sarcomatous in nature [1].

Only few cases of primary leiomyosarcoma of greater omentum have been reported. Majority of the cases were diagnosed in middle age and there was male preponderance seen in the previously reported cases. The most common presenting symptom was abdominal mass followed by pain abdomen similar to the present case. Ascitis was also reported in few cases [2,3,6]. Our patient did not have ascitis.

CT scan was useful in determining the origin of the tumor. Larger GISTs (>5 cm), like leiomyosarcoma, are frequently lobulated and usually have intratumoral hemorrhage, necrosis, or cystic change. For these reasons, preoperative radiological differentiation between leiomyosarcoma and GIST is difficult. E-GISTs are rare, account for fewer than 5% of cases of GIST; they originate primarily from the mesentery, omentum or peritoneum and are seen in patients over age of 50 years [5,7].

Microscopically leiomyosarcoma can be distinguished from GIST by the cell morphology, absence of skeinoid fibers, high degree of atypical mitotic figures and abundant areas of necrosis. CD117, DOG1 and CD34 are well known specific markers for GIST and are negative in leiomyosarcoma.

Leiomyosarcomas are almost invariably and strongly positive for SMA, but SMA positivity alone is not sufficient. A good majority (70–80%) are also positive for desmin. Most cases are positive for heavy-caldesmon and smooth muscle myosin.

The latter two markers can be useful in differentiating leiomyosarcoma from myofibroblastic sarcomas [7, 8]. Other differential diagnosis which have to be considered are metastatic poorly differentiated carcinomas arising from gastrointestinal tract and pancreas. Due to its malignant behaviour, radical excision of the tumor should be done. Reported cases

demonstrated that 36% of all patients presented with metastatic deposits, most commonly to liver. The long term outcome of patients with omental leiomyosarcomas is unknown. Early reports showed poor outcomes with a high post-operative death rate. Given the poor prognosis of leiomyosarcoma, misdiagnosis may have a detrimental effect on patient outcome [1,2].

## Conclusion

We report this case of primary leiomyosarcoma of greater omentum for its rarity. Although omental leiomyosarcoma is rare, it should be considered in elderly patients presenting with huge abdominal mass arising from greater omentum.

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