Case Report

Primary Leiomyosarcoma of Liver — Case Report of a Rare Cancer

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Primary Hepatic Leiomyosarcoma are very rare Tumours with only 30 cases reported Worldwide[1]. Patient complains of nonspecific symptoms and often presents with Enlarged Liver. Diagnosis is usually delayed and patients have poor prognosis[2]. 35-year-old woman presented to Outpatient Department (OPD) complaining of dull aching pain and mass in upper part of abdomen for 6 months. On palpation liver was enlarged and lower border extended up to umbilicus. Ultrasonography of abdomen showed Liver enlarged 23 cm with multiple hypoechoic nodules noted in both lobes of liver. Triphasic Contrast-Enhanced Computed Tomography (CECT) abdomen multiple variable sized Peripherally Enhanced Hypodense Nodules in both lobes of Liver suggestive of Multifocal Hepatocellular Carcinoma (HCC)/ Metastasis. Tumour markers S alpha fetoprotein, S. CA 19-9, S. CEA was within normal limits. CECT Chest was normal. Positron Emission Tomography and Computed Tomography (PET-CT) was done, which showed Liver enlarged 24.5cm with FDG avid multiple nodules in both lobes of Liver and multiple enlarged FDG avid periportal, Portocaval and Peripancreatic Lymph Nodes. Rest of body organs were normal. Biopsy from the Liver showed Mesenchymal Tumour composed of spindle cells arranged in fascicular growth pattern, nucleus cigar shaped with atypia and mitotic figures seen. On Immunohistochemistry (IHC) Tumour was positive markers were Smooth Muscle Actin (SMA), VM, Desmin and H-caldestron and negative for CK, S-100, SOX-10, CD-117 and Dog-1. From above work up a diagnosis of primary Leiomyosarcoma of Liver was made. Palliative Chemotherapy was offered to the patient.

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Key words **: Primary Leiomyosarcoma of Liver.**

Sarcomas are tumours which arise from Mesenchymal Tissues. They are very rare and form about 1% of all adult tumours. Leiomyosarcomas constitute 5-10% of all Sarcomas. Primary Hepatic Leiomyosarcomas are very rare tumours. Most of Leiomyosarcomas in Liver present as Metastatic Tumour[3]. Primary Leiomyosarcomas most commonly occurs in Uterus, Retroperitoneum, Lower and Upper Limbs[4]. Hereby, reporting a case of 35-year-old lady presenting as primary Hepatic Leiomyosarcoma with Periportal, Portocaval and Peripancreatic lymph nodes enlargement.

Case Report

A 35-year-old woman presented to Outpatient Department (OPD) complaining of dull aching pain and mass in upper part of abdomen for 6 months. History of decreased appetite and weight loss was present. No history of alcohol abuse and no history of malignancy in family members was noted. On palpation liver was enlarged and lower border extended up to umbilicus 10cm below costal margin. Liver Function Test (LFT) was normal with Alanine Aminotransferase (SGPT) 14 IU/L (14-63 IU/L), Aspartate Aminotransferase (AST) 59 IU/L (45-145 IU/L), Alkaline Phosphatase (ALP) 76 IU/L (28–94 IU/L), Serum Albumin 2.3 g/dL (3.5-5.2 g/dL). Serum Bilirubin, Prothrombin Time, Complete Blood Count, Renal Function Test was within normal. HbsAg, HIV and Anti HCV test was non-reactive. Tumour markers was within normal limits. S. alpha-fetoprotein was 5.5 (0-10 ng/mL), S. CA 19-9 3.78 IU/mL and S. CEA 7.5 ng/mL (5-10 ng/ mL). Ultrasonography of abdomen showed Liver enlarged 23 cm with multiple hypoechoic nodules noted in both lobes of Liver. Intrahepatic Biliary radical and hepatic vasculature was normal. Triphasic Contrast-Enhanced Computed Tomography (CECT) abdomen multiple variable sized peripherally enhanced hypodense nodules in both lobes

Editor's Comment :

- Primary hepatic leiomyosarcoma is a rare condition and diagnosis should be made by ruling out other common conditions like liver metastasis from Gastrointestinal Tract (GIT), uterus,extremities, retroperitoneum and common primary tumours of liver.
- Tumour markers are usually normal and liver biopsy shows mesenchymal cells on histological examination.
- Excision of tumour with negative margins is the treatment of choice.
- Chemotherapy can be offered to patients with positive margins, unresectable and metastatic disease.

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of Liver suggestive of multifocal Hepatocellular Carcinoma (HCC)/ Metastasis. Rest of abdomen was normal. CECT Chest was normal. Positron Emission Tomography and Computed Tomography (PET-CT) was done, which showed Liver enlarged 24.5cm with Fludeoxyglucose (FDG) avid multiple nodules in both lobes of Liver (maximum SUV 18.6) and multiple enlarged FDG avid periportal, portocaval and Peripancreatic Lymph Nodes (largest lymph node 4.1 x 3.6 cm, maximum SUV 10.9). Rest of body organs was normal. Biopsy from the Liver showed Mesenchymal Tumour. Spindle cells were arranged in fascicular pattern. Tumour cells were merged with blood vessels. Nuclei was cigar shaped with variable Atypia, with Cytoplasmic Vacuoles noted at both ends and mitotic figures seen. On Immunohistochemistry (IHC) Tumour was positive markers Smooth Muscle Actin (SMA), Vimentin, Desmin and H-caldestron and negative for CK, S-100, SOX-10, CD-117 and Dog-1. From above work up a diagnosis of Primary Leiomyosarcoma of Liver was made. Palliative Chemotherapy was offered to the patient (Figs 1-5).

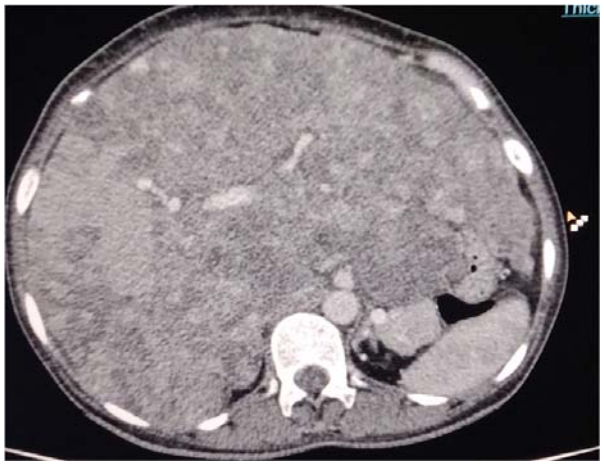

Fig 1 — CECT showing peripherally enhanced hypodense nodules in both lobes of liver

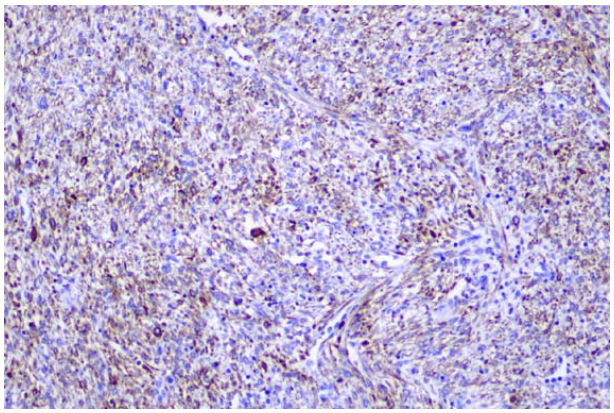

Fig 3 — Tumour cells were positive for desmin on IHC (10x)

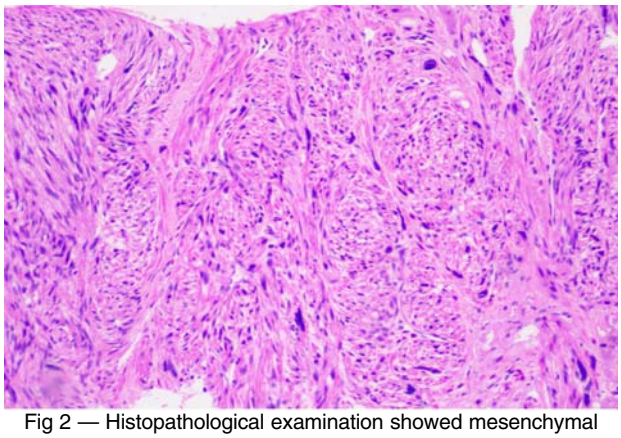

Fig 2 — Histopathological examination showed mesenchymal tumour composed of spindle cells arranged in fascicles (10x)

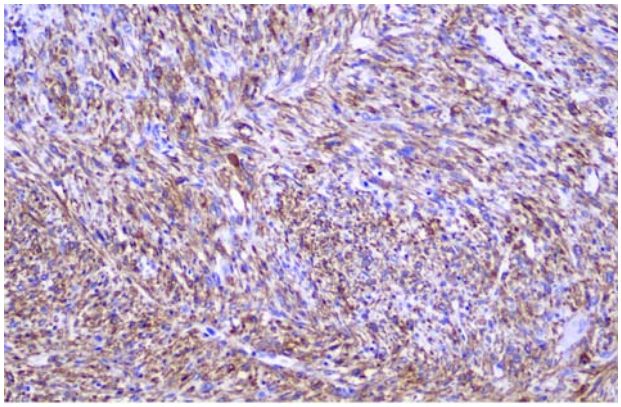

Fig 4 — Tumour cells were positive for H-caldestron on IHC (10x)

Discussion

Sarcomas occur rarely in Liver and they form only 1-2% of Malignant Tumours of the Liver. Hepatic Leiomyosarcomas mostly present as secondary with primary being elsewhere like Gastrointestinal Tract, Retroperitoneum, Uterus, Major Blood Vessels and Genitourinary Tract[3]. Extensive imaging is needed in form of CECT abdomen, Chest, Endoscopy and PET-CT to rule Metastasis from other organs. Therefore, Primary Hepatic Leiomyosarcoma should only be diagnosed after ruling out Metastasis from other organs. Angiosarcoma followed by Leiomyosarcoma forms the most common type of Primary Malignant Mesenchymal Tumour of Liver[4]. Smooth muscle cells in Liver like Intrahepatic Blood Vessels or Biliary Ducts gives origin to Primary Hepatic Leiomyosarcoma[5,6]. Association between Leiomyosarcoma of Liver and Immunosuppression is well documented in literature. AIDS, Epstein-Barr Virus (EBV) infection and immunosuppression in post transplant patients are risk factors for leiomyosarcoma of Liver[7-9]. Primary Liver Leiomyosarcomas usually present with non-specific symptoms and the diagnosis is often delayed resulting in poor prognosis[2]. Abdomen tenderness and enlargement are most common signs[10]. Patient may rarely present with haemorrhage from tumour site[11]. Presentation usually Mimics Hepatocellular

Carcinoma but Serum Alpha-fetoprotein is usually normal as noted this patient. On Biopsy and Histopathology Examination shows tumuor with intersecting bundles of spindle-shaped cells which helps to differentiate from Hepatocellular Carcinoma. On Immunohistochemistry tumour tests positive for Desmin, Vimentin and SMA and tests negative for Keratin, S-100 Protein and Neuron-specific enolase[12]. The above-mentioned finding was noted in this patient. Another uncommon finding noted in this patient is enlargement and enhancement periportal lymph nodes which is usually seen in Cholangiocarcinoma[13] but serum CA19-9 and S. Carcinoembryonic Antigen (CEA) was normal. Standard guidelines for treatment of Hepatic Sarcomas are not well defined but it is well documented that resection with negative margins gives patient maximum survival[14,15]. Evidence for role of Adjuvant Chemotherapy is lacking. Chemotherapy be given in R1 resection, unresectable and Metastatic Disease[16]. Liver transplantation is hepatic Leiomyosarcoma has shown long term survival[17,18].

Conclusion

Primary Hepatic Leiomyosarcoma is rare Cancer and diagnosis is usually made by ruling out Metastasis to Liver from GIT, common primary tumours of Liver and Metastasis to Liver from common sites of Leiomyosarcoma like Uterus, Retroperitoneum and Extremities. It may mimic Hepatocellular Carcinoma or Intrahepatic Cholangiocarcinoma by presentation but Serum Tumour markers usually will be normal. Extensive imaging, Biopsy, Histopathological Examination and immunohistochemistry helps in diagnosis. More reporting is required for better understanding of this rare variety of tumour.

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