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Metastatic Leiomyosarcoma of Gall Bladder: Rare Case Report

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Abstract: ***Introduction: Primary sarcomas of the gallbladder (GB) as a rare disease, the incidence of which is more in women than men. Case Report: A 52 -year-old female patient presented to the outpatient clinic with complaints of intermittent pain in the right upper quadrant of the abdomen, nausea and some weight loss for 2-3 months. Ultrasound revealed Gall bladder mass with liver invasion. CT and PET-CT scan revealed Periportal and omental deposits . Final diagnosis was reached by FNAC and IHC as Metastatic Leiomyosarcoma of Gall Bladder. She was advised palliative chemotherapy as the tumor was unresectable and in advanced stage. She refused for chemotherapy and eventually died 3 months after diagnosis. Discussion: LMS of Gall Bladder is a rare entity. Patient presents with complaints of right upper quadrant pain, nausea, vomiting. It may be accompanied by cholelithiasis or cholecystitis or both. It is usually diagnosed at an advanced stage when there are only palliative options left. Survival rate is very poor. Conclusion: Leiomyosarcoma of gall bladder should be kept in mind when evaluating a patient with gall bladder mass.***

1. Introduction

Griffon and Segall first described the primary sarcomas of the gallbladder (GB) as a rare disease[1], the incidence of which is more in women than men (Ratio 5:1)[2] . Mostly, Primary Sarcoma of gall bladder appears in the 6th or 7th decade of life with cholecystitis (with or without cholelithiasis) as a primitive diagnosis[3].

More than 40% of tumour like lesions of gall bladder are adenomatous hyperplasia, which is the most common benign mesenchymal proliferation [4]. Though, the malignant degeneration of adenomatous hyperplasia is rare, the malignant mesenchymal neoplasms are extremely rare. Leiomyosarcoma of GB is rarely diagnosed preoperatively. The accurate diagnosis is only established with histological and immuno-histochemical techniques. [5]

2. Presentation of Case

A 52 year old female patient presented to the with complaints of intermittent pain in the right upper quadrant of the abdomen, nausea and weight loss for the last 2-3 months. On physical examination, abdomen was soft, non distended with tenderness in the right hypochondrium region with Hepatomegaly and an irregular palpable mass in the right hypochondrium till 2 finger breadths below the costal margin and hard in consistency. No audible bruit was heard and bowel sounds were present.

Abdominal ultrasound was suggestive of over-distended GB, loss of GB and liver fat plane, Hepatomegaly with multiple ill-defined mass lesions? Secondaries, with multiple GB calculi largest measuring 17mm. (Figure 1)

Liver function tests were normal except Serum Alkaline Phosphatase which was mildly raised. The tumor markers were within normal range (carcinoembyonic antigen, carbohydrate antigen 19-9, alpha-fetoprotein).

Contrast enhanced computed tomography (CECT) of abdomen demonstrated a mass measuring 10.7 x 7.9 x 12.4 cm involving segments V and VI of liver extending into adjacent GB lumen and peri-cholecystic fat region (multiple enhancing nodular lesions, largest measuring10.4 x 8.8 mm) with non enhancing hypodense cysts in segments II, IV and VII of liver. (Figure 2)

The 18F-fluorodeoxyglucose positron emission tomography-CT (18-FDG PET-CT) was done to evaluate for distal metastasis, it revealed liver mets in segment IV, V andVI with ill-defined fat planes with gall bladder and GB wall thickening. It also showed peri-portal and omental metastasis of the tumor. (Figure3)

2.1 Pathological Findings

It showed spindle shaped cells in fascicular architecture with mild nuclear pleomorphism and mitotic figures, suggestive of Spindle Cell Sarcoma of GB.

2.2 Immuno-histochemistry (IHC) marker study findings

It showed smooth muscle actin - positive, Desmin - positive, H-Caldesmon - positive, Dog-1
- negative, CD-117 - negative, Ki67 index - 20%). These findings confirmed the diagnosis of Leiomyosarcoma of GB. (Figure 4)

The patient was advised palliative chemotherapy after consulting an Oncologist.

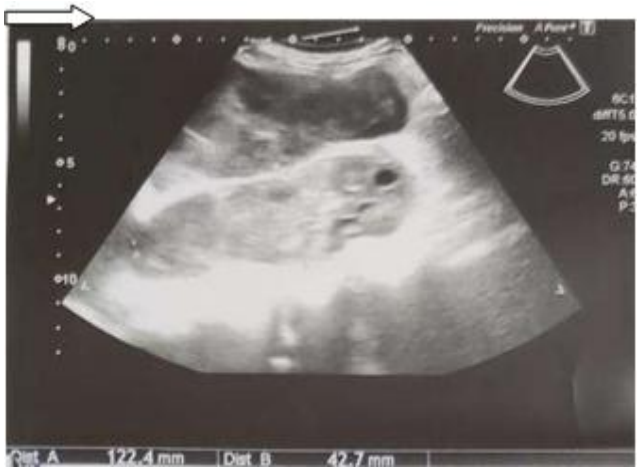

Figure 1: Multiple ill-defined mass lesions ? secondaries, over-distended GB with discontinuation of GB and liver fat plane with multiple GB calculi

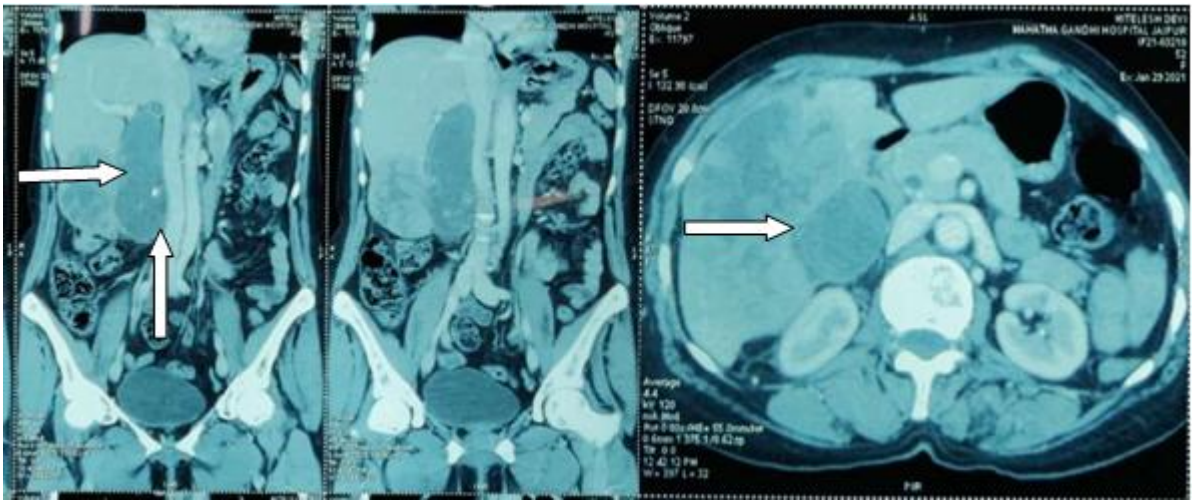

Figure 2: CECT Abdomen showing a GB mass measuring 10.7 x 7.9 x 12.4 cm involving segments V and VI of liver extending into adjacent GB lumen and peri-cholecystic fat region (multiple enhancing nodular lesions, largest measuring10.4 x 8.8 mm) with non enhacing hypodense cysts in segments II, IV and VII of liver

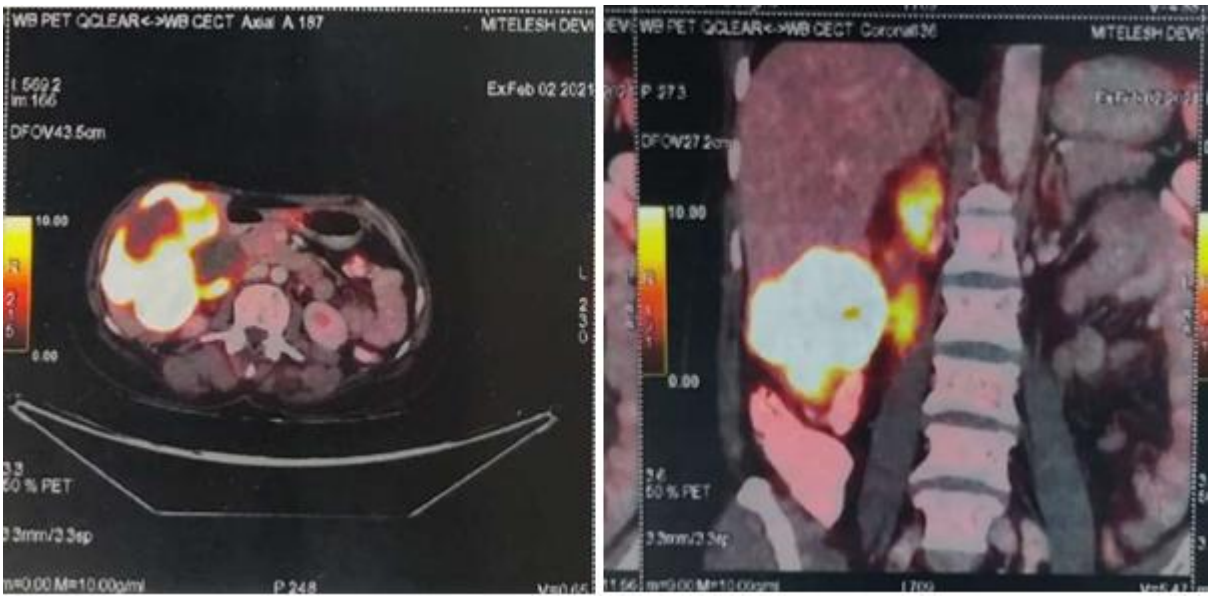

Figure 3: The 18F-fluorodeoxyglucose positron emission tomography/CT that was performed to check for distal metastasis revealed liver lesions in the segments IV, V, VI of liver parenchyma with ill defined fat planes with GB wall thickening with periportal deposits.

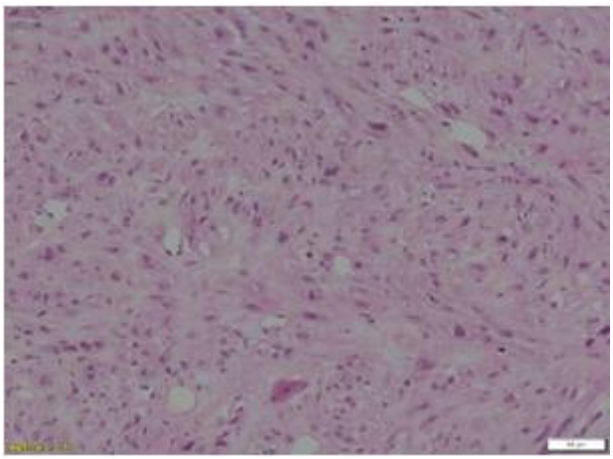

Figure 4 (A): H&E Slide

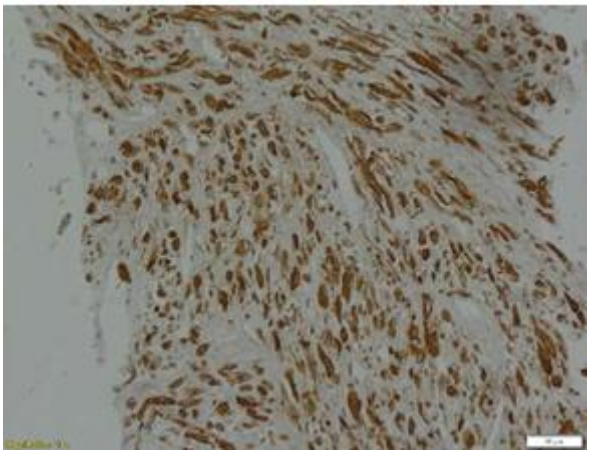

Figure 4 (B): Smooth muscle actin positive

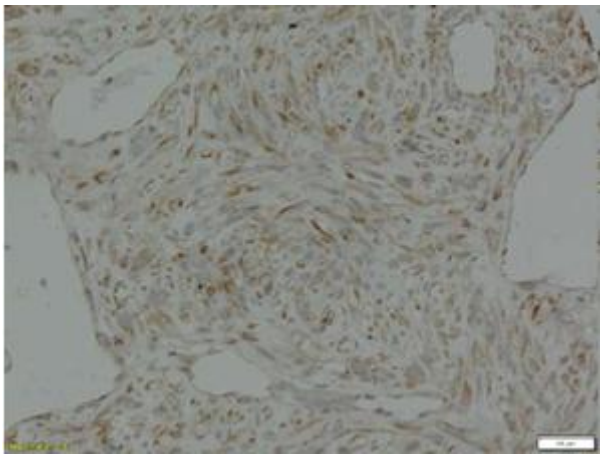

Figure 4 (C): Desmin Positive

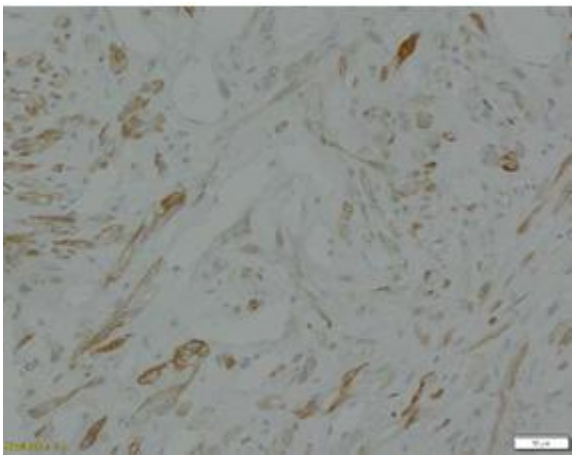

Figure 4 (D): H-Caldesmin - Positive

3. Discussion

Leiomyosarcoma GB, primary sarcoma is a rare malignancy with an incidence of 1.4 per 1000 malignancies of the GB[7]. World Health Organisation classification for soft tissue tumours is used to establish the diagnosis of a Leiomyosarcoma[8]. Microscopically, it consists of intersecting, marginated groups of spindle cells. It is rarely diagnosed pre- operatively. Patients who suffered from a LMS of the GB often present with abdominal pain, fever, jaundice and history of weight loss [6]. In certain cases, as the one presented here, cholelithiasis led to the diagnosis of a GB tumor. Predisposing factors regarding the pathogenesis of the LMS maybe gallstones and acute/ chronic inflammation of the GB [6]. Previous reviews revealed that the majority [11,12] of patients were referred to the hospital suffering from gallstones with acute cholecystitis. [2, 3, 7, 9, 13, 14, 15, 16, 17, 18, 19].

For diagnosis, an ultrasonography, a CT scan and a PET-CT scan are recommended. The LMS may occur as a polypoid mass protruding into the lumen with an irregularly thickened wall. Nevertheless, the lack of specific radiological features makes the differentiation from an adenocarcinoma difficult [20]. Differential diagnosis to consider are adenocarcinoma, rhabdomyosarcoma, liposarcoma, Kaposi sarcoma, and angiosarcoma of the Gall Bladder [11, 14].

The approach for treatment depends on tumor extension. In a non-metastatic stage, Paasch et al (2020) [21], recommended that cholecystectomy combined with a wedge resection of the surrounding liver tissue, along with lymphadenectomy of the hepato- duodenal ligament seems to be a sufficient surgical approach. There is not sufficient evidence in the literature regarding the effectiveness of adjuvant chemo- or radio- therapy. However, many authors recommended palliative surgical bypass due to its poor prognosis, with liver involvement in almost 75% of cases and a five-year survival rate of less than 5%. Adjuvant chemotherapy (doxorubicin, mitomycin C) has prolonged the survival rate in some cases .The LMS of the GB has a very poor prognosis, particularly in a metastatic stage [2, 9, 22]. This was seen our patient who died within 3 months after diagnosis.

4. Conclusion

When diagnosing a tumor of Gall Bladder, Leiomyosarcoma should be kept in the differential diagnosis. In a non-metastatic disease, radical cholecystectomy with lymphadenectomy of the hepato-duodenal ligament is a sufficient surgical approach but in a metastatic disease, chances of survival are very less. Chemotherapy can be tried but it has no major benefit.

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