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Case Report (Open Access)

MALIGNANT PRIMARY ADRENAL LEIOMYOSARCOMA: AN UNCOMMON ENTITY

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ABSTRACT
A 62 year old gentleman presented with chief complaints of dull aching left loin pain for 2 months. On evaluation with contrast enhanced computed tomography (CECT) of abdomen revealed a left adrenal mass suspicious of malignancy, he underwent laparoscopic left adrenalectomy and histopathological analysis revealed dedifferentiated leiomyosarcoma. PET CT revealed metastatic lesions in the liver. He was referred to the medical oncology department and initiated on adjuvant therapy. He is having a stable disease at 15 months of follow-up.

Keywords*: Malignant Primary Adrenal Leiomyosarcoma, Laparoscopic adrenalectomy*

INTRODUCTION
Leiomyosarcomas (LMS) represent neoplasms originating from smooth muscle tissue. Primary LMS of the adrenal gland are rare with less than 50 cases described in the literature (Sakellariou *et al.,* 2020). This neoplasm exhibits an aggressive biological nature. Diagnosis relies entirely on histological and immunohistochemical assessments and cannot be diagnosed preoperatively (Onishi *et al.,* 2016). In this report, we document a rare case of leiomyosarcoma arising from the left adrenal gland in a 62-year-old man.

CASE
A 62-year-old gentleman, hypertensive for 3 years presented with dull aching pain in the left loin for two months. He did not have vomiting, hematuria, weight loss, or fever. Physical examination revealed no significant abnormalities. CECT of abdomen showed 86.9 x 82 x 81mm heterogeneously enhancing lesion arising from left adrenal gland with relative washout of < 40% suggestive of left adrenal malignant lesion. There was no evidence of lymph nodal or distant metastasis [Figure 1]. Biochemical workup for adrenal tumour was normal. Laparoscopic adrenalectomy was performed and tumor was not found to be extending into neighboring structures [Figure 2].
On macroscopic examination specimen was nodular and measured 11x7.5 x5.5 cm. Cut surface showed a well defined tumor measuring 8 x 5.5 x 6cm with firm grey white bulging nodular appearance with normal adrenal gland over a span of 2.5 x 0.3 cm.
Histopathological examination showed compressed adrenal gland circumscribed by cellular tumour arranged in the pattern of long intersecting fascicles, bundles and nodules. The tumour cells are spindle shaped with moderate eosinophilic cytoplasm, blunt ended nuclei, mild to moderate pleomorphism and increase in mitosis with atypical forms (10-12/HPF). There were focal undifferentiated component. All margins were free of tumour.
Immunohistochemical stains of smooth muscle actin (SMA) and desmin were diffuse and strongly positive in >90% tumour cells and weakly staining in the remaining tumour cells showing undifferentiated component. The IHC markers of C-kit, DOG-1, S-100 protein, and CD34 were negative. The final diagnosis of dedifferentiated leiomyosarcoma was made [Figure 3].
Considering the high grade and aggressive biological characteristics of the tumour patient was scheduled for adjuvant therapy. Prior to commencing chemotherapy, a PET CT scan was performed after 1 month

post surgery, revealing metastatic lesions in the liver. Patient is initiated on chemotherapy with Doxorubicin. On follow-up imaging at 15 months post surgery he is not having any local recurrence and his liver metastasis is remaining stable without increase in size.

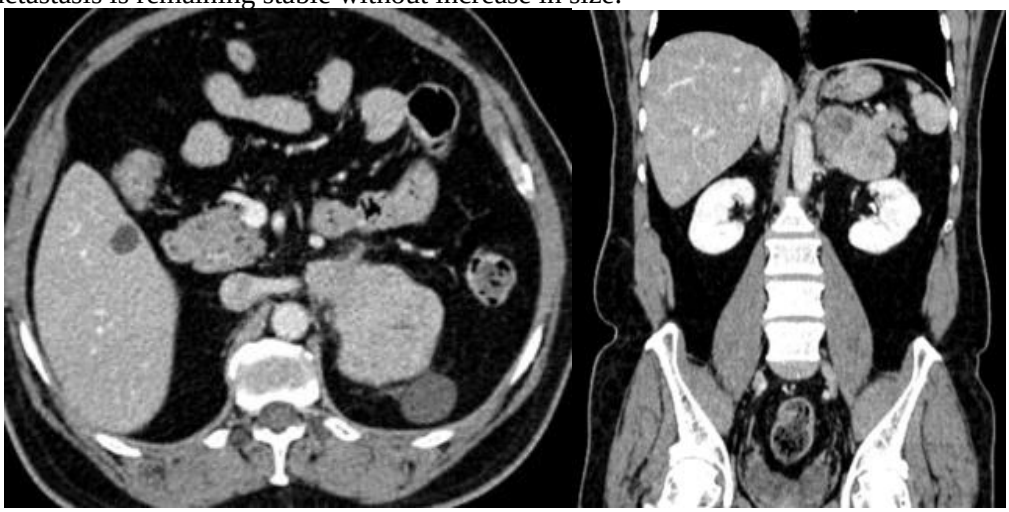

Figure 1.a **Figure 1.b**

Figure 1: Contrast-enhanced CT scan of abdomen axial(a) and coronal(b) sections, showing heterogeneously enhancing lesion arising from left adrenal gland

DISCUSSION

Primary leiomyosarcomas of the adrenal gland (PLAG) are rare. The majority of these tumors are benign (myelolipomas and hemangiomas). Malignant mesenchymal tumors of adrenal gland encompass leiomyosarcoma, malignant peripheral nerve sheath tumors, and angiosarcoma. Among these, PLAG are the most common malignant ones. These tumors are presumed to originate from the adrenal vein or its branches(Nagaraj *et al.,* 2015). Adrenal hormones are usually normal. No specific tumor markers, distinct clinical manifestations nor imaging modalities have been identified(Jabarkhel *et al.,* 2020).

Radical surgery is the mainstay of treatment for localized PLAG. Although there is a lack of available data regarding the outcomes of laparoscopic adrenalectomy in PLAG, it is found to be equally good as open surgery in adrenocortical carcinoma (ACC) (Mpaili *et al.,* 2018).

At present, the diagnosis of PLAG relies on histopathological and immunohistochemical assessments. PLAG typically comprises spindle cells that demonstrate positive staining for one or more markers associated with smooth muscle cells, notably SMA (in 90 to 95% of cases) and desmin (in 70-90% of cases). High grade component lacks evidence of smooth muscle differentiation and is known as dedifferentiation (Venyo *et al.,* 2020; Zhou *et al.,* 2015). In our patient immunohistochemical analysis demonstrated the expression of SMA and desmin in tumor cells of low grade component and absent to weak staining in high grade undifferentiated component.

PLAG exhibits aggressive characteristics. Among the reported cases half of the patients experienced metastasis or mortality within 3 years of diagnosis. Nearly 20% succumbed within 6 months of diagnosis (Chen *et al.,* 2011; Wang *et al.,* 2020).

Adjuvant radiotherapy or adjuvant chemotherapy has been utilized in cases of PLAG especially in cases of invasive tumours that have invaded near-by organs or possess high-grade characteristics. Due to the rare incidence of this disease, no trials or established guidelines are currently available regarding management. The use of chemotherapy with dacarbazine and doxorubicin as first-line followed by second-line treatment with gemcitabine and docetaxel have been reported to prevent progression of disease and survival more than an year in patient with liver metastasis. Immunotherapy targeting PDL-1 has been applied in the treatment of uterine sarcomas; however, there are currently no available reports

regarding the usage of targeted therapy or immune checkpoint inhibitors (ICIs) for PLAG (Oshidari *et al.*, 2022; Mpaili *et al.*, 2018).

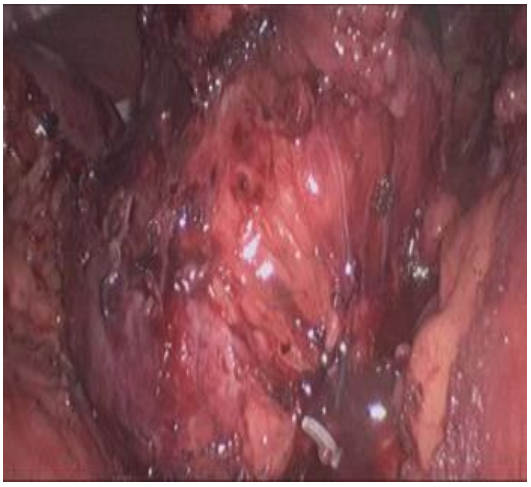

Figure 2: Intraoperative image of left laparoscopic adrenalectomy

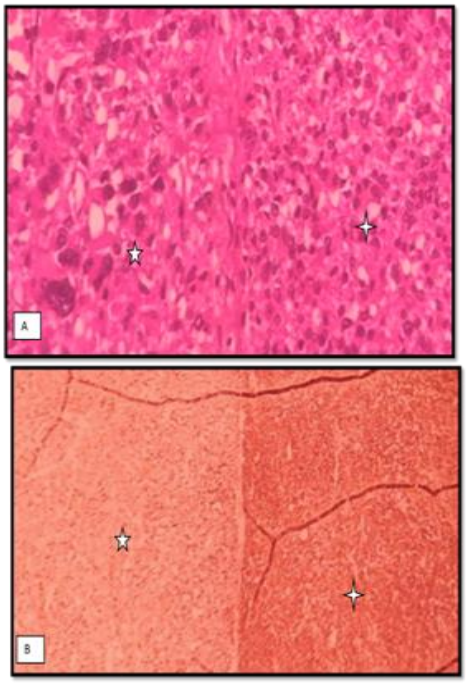

Figure 3: Microscopy A) H&E 40x, Cellular tumor arranged in long intersecting fascicles of spindle shape with moderate eosinophilic cytoplasm, plump blunt ended nuclei, and moderate pleomorphism (✧), with focal dedifferentiated component made of rhabdoid cells (☆).
B) Immunohistochemistry demonstrated the expression of SMA in >90% of tumor cells with low grade component (✧) and absent to weak staining in high grade undifferentiated component (☆).

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