Primary Adrenal Leiomyosarcoma in an Arab Male: A Rare Case Report with Immunohistochemistry Study

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1. Introduction

Primary adrenal leiomyosarcoma is a rare form of adrenal mesenchymal tumors. Immunohistochemistry (IHC) together with histology takes a major role in determining the tumor type and predicting their biological behavior and differentiating them from adrenal cortical carcinoma. Appropriate radiological investigation is necessary to rule out metastatic disease from primary tumors elsewhere in the body. In this case, we report a primary leiomyosarcoma of the adrenal gland in a 61-year-old Bahraini male clinically presumed to be a renal neoplasm.

Available information on the Internet indicates approximately 20 reported cases of leiomyosarcoma around the world so far. The case of adrenal leiomyosarcoma reported in Bahrain was originally thought to be a renal tumor. However further radiological and immunohistological study led us to correct diagnosis.

2. Clinical History

The 61-year-old Bahraini male patient with a known history of diabetes and hypertension presented with left flank pain of few months duration. He had no other significant complaints or relevant past history. He went initially to a tertiary hospital where he was clinically diagnosed to have a renal tumor. The CT abdomen with contrast showed a large mass in the upper pole of the left kidney with heterogeneous enhancement measuring 16 × 10.7 × 11.7 cm displacing the lower pole of the kidney with mild stranding of the perinephric fat giving a radiological diagnosis of renal neoplasm.

However, the patient went to another hospital for further investigations and treatment. Multiplanar MRI in that hospital using T1 and T2 weighted TSE SPIR postcontrast sequence showed an 11 × 12 × 13 cm oval mass with heterogeneous
Figure 1: In phase axial image: 11 × 12 × 13 cm well defined hypointense lesion anterior and superior to Lt. kidney abutting the renal vessels and Lt. crus of diaphragm with no signal voids indicating calcification and no hyper intense signal indicating fat content or hemorrhage.

Figure 2: Sagittal T2W image: the lesion shows intermediate signal with multiple small hyper intense areas due to necrosis.

elevation with noneenhancing area due to cystic degeneration in the anteromedial aspect of the left kidney displacing it posteroinferiorly. The tumor was seen abutting the splenic vein and abdominal aorta and stretching the left renal vein and artery leading to the diagnosis of adrenal tumor rather than renal origin (Figures 1 and 2). The right kidney, liver, right adrenal gland, and the gall bladder were within normal limits with no enlarged abdominal lymph nodes. Radiological differentials of pheochromocytoma, adrenal cortical carcinoma, and leiomyosarcoma were given.

The complete blood count showed mild normochromic normocytic anemia with Hb of 10.2 × 10^9/L. Urea and electrolytes, PSA, and thyroid function test were within normal limits. The serology including autoimmune markers was negative. The midstream urine (MSU) showed RBC 3–5/HPF, WBC 2–4/HPF, and no protein, cast, crystals, or bacteria were found. Other laboratory investigations were within normal limits and no tumor marker estimations were done.

Prior to operation, according to the hospital protocol, the patient received two units of PRBCs, and then he underwent laparotomy with removal of the adrenal tumor. Intraoperatively, the tumor was arising from the left adrenal gland pushing the kidney inferiorly with no fixation to kidney or other adjacent organs. Excision of the tumor with preservation of the left kidney was performed. The patient withstood the operation well and was discharged on 5th postoperative day without any complication. Postoperatively, we lost contact with the patient as he sought further overseas oncology management.

Grossly, multiple pieces of greyish white friable tissue with attached small amount of fatty tissue were received in the lab altogether weighing 1040 g and measuring 17 × 13 × 8 cm in maximum dimension. The cut surface was solid and grayish white in color with few mucoidal areas. Tiny hemorrhagic areas were present. No normal or residual adrenal gland was identified. Microscopic examination revealed a high grade spindle cell malignant tumor composed of diffuse proliferation of large pleomorphic spindly cells with eosinophilic cytoplasm and spindly, blunt ended vesicular nuclei (Figure 3). Many large pleomorphic cells, few multinucleate giant cells, and brisk bizarre mitosis with widespread apoptosis were present. Large areas of necrosis were noted. Considering the location of the tumor, primary adrenal cortical and medullary tumors and metastatic tumors as well as retroperitoneal tumors were considered in the differential diagnosis. Immunohistochemistry was performed which revealed that the tumor cells were strongly positive for desmin (Figure 4) and vimentin and negative for calretinin, inhibin, chromogranin, synaptophysin, S100, pan keratin, and CD68. No normal adrenal gland was present. The available adipose tissue showed no signs of tumor. Based on microscopic and immunohistochemical findings, together with the imaging studies and the confinement of the tumor...
to the upper pole of the kidney with no involvement of other intra-abdominal structures, a final diagnosis of primary adrenal leiomyosarcoma was given.

3. Discussion

Primary adrenal mesenchymal tumors are very rare and are mostly composed of benign tumors like myelolipomas and haemangiomas [1]. The most common malignant primary adrenal mesenchymal tumor is leiomyosarcoma which usually has asymptomatic presentation. Till now, only around 20 cases have been reported around the world [2–4].

The age of patients ranged from 30 to 73 years with male to female ratio of 3:1 and median size of the tumor is 11–25 cm [2, 4, 5]. With the cut-off size 3 cm for benign tumors, for all larger tumors, preoperative imaging and screening should be performed to assess the resectability of the tumor and the possibility of metastasis from clinically occult tumor [1]. In our case, although initially thought to be a renal tumor, further radiological study confirmed the adrenal origin of the tumor and did not reveal any tumor in other sites.

The origin of the tumor cells is widely presumed to arise from the smooth muscle wall of the inferior vena cava, central adrenal vein, and its tributaries [1, 2, 5–8]. Although the etiologies have not been clearly elucidated, association with HIV and EBV is suggested [2–6, 9, 10]. Scattered case reports suggest severe tissue trauma as the casual or contributing etiopathology in some cases. van Etten et al. suggested that a gunshot wound could be the contributing pathogenetic factor [1].

Morphological changes and clinical behaviors can be related to chromosomal aberrations [3, 6]. For example, abnormalities like 13q14–q21 loss, 5p14–pter gain, RB-1 genes, and RB-cyclinD are some of the frequent genetic abnormalities that are associated with the shorter survival [3].

Although the tumor is uniloculated, it affects both the sides equally. The patients usually present with abdominal flank pain, mass or symptoms of adjacent organ involvement like IVC obstruction leading to lower limb swelling, venous gangrene, and pulmonary embolism. They can also present with symptoms of metastasis to liver, lung, and bone leading to shortness of breath, jaundice, bone pain, and paresis [4–6, 11].

As there are no definitive biomarkers, preoperative diagnosis is difficult [4]. Although radiological features cannot differentiate among the different types of adrenal malignancies, it can be helpful in differentiating adrenal adenomas and nonadenomas by considering the size cutoff, growth rate, and characteristic radiological imagining [1, 4, 12]. Histopathology with immunohistochemistry is mandatory to determine the tumor type along with its grading and aggressiveness [3–5].

Metastatic tumors, malignant melanoma, GIST, MPNST, sarcomatoid renal cell carcinoma, malignant fibrous histiocytoma, and primary retroperitoneal sarcoma infiltrating the adrenal gland should be considered in the differential diagnosis along with the other adrenal and metastatic tumors and appropriate immunohistochemistry.
should be performed to reach a specific diagnosis. Radiological investigations should also be included in the workup to reach an appropriate diagnosis and to assess the prognosis. Even with complete resection, the prognosis is generally poor and appropriate long standing follow-up is required.

**Conflict of Interests**

The authors declare that there is no conflict of interests regarding the publication of this paper.

**References**


