CASE REPORT

Primary Adrenal Leiomyosarcoma: A Rare Case Report

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Abstract

Primary adrenal leiomyosarcoma (PAL) is an extremely rare case finding where the mesenchymal tumor is found to be originating from the mesenchymal elements surrounding the adrenal vessels. Herein we report an interesting case of a 60-year-old gentleman, presenting to us with an abdominal lump. Contrast-enhanced computed tomography of the (CECT) abdomen revealed multilobulated heterogeneously enhancing soft tissue lesion of size 99 × 93 × 33 mm in the left supra renal region. After evaluation, the patient underwent an open left adrenalectomy. Postoperatively the histopathological examination showed mesenchymal neoplasm with spindle-shaped vesicular chromatin, conspicuous nucleoli, and eosinophilic cytoplasm with occasional atypical mitosis.

Keywords: Adrenocortical carcinoma, Case report, Primary adrenal leiomyosarcoma.

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Introduction

Leiomyosarcoma is an aggressive type of mesenchymal tumor originating from smooth muscles. Primary adrenal leiomyosarcoma (PAL) is a rare entity, originating from the smooth muscle wall of the central adrenal vein or its branches.[1] Primary adrenal leiomyosarcoma represents 0.1–0.2% of all retroperitoneal tumors.[2] Only a few cases have been reported in the literature till now. Herein we report another interesting case of PAL presenting to us.

Clinical Case

A 60-year-old gentleman presented to us with a lump in his left upper abdomen for 4 months associated with pain, which was moderate in intensity, non-radiating, and having no aggravating or relieving factors. He also gave a history of significant weight loss of 10 kilograms in 4 months. There was no history of loss of appetite, altered bowel habits, vomiting, No history of episodic headache, palpitations, or diaphoresis. He had no other comorbidities. The patient was normotensive. On per abdominal examination, he had a 5 × 5 cm, ill-defined mass in the left hypochondrium, which was hard in consistency and moving with respiration. Finger insinuation between the left costal margin and lump was not possible and it was not bi-manually palpable.

The functional adrenal tumor was ruled out after biochemical evaluation (ONDST-3.10 µg/dL, 24-hour urinary metanephrine and nor-metanephrine values were normal (83.65 µg/24 hrs and 430.15 µg/24 hrs respectively). His CECT abdomen with the adrenal protocol was done, which showed a well-defined multilobulated heterogeneously enhancing soft tissue attenuating lesion of 99 × 93 × 33 mm with internal non-enhancing necrotic areas in the retroperitoneum in the left supra renal location likely origin from the posterior limb of left adrenal. The mass was medially abutting the aorta with maintained fat planes, proximally displacing the left kidney and left renal vessels with no luminal compromise. The lesion showed an absolute washout of 28.9% and a relative washout of 18.15%. Given the large size and heterogeneous lesion with low washout values, it was suspicious of adrenocortical carcinoma.

The patient was optimized and planned for left open adrenalectomy. Given the large size of the lesion, an open approach rather than a laparoscopic approach was planned. Intraoperatively, a 382 gm dumbbell-shaped left adrenal mass with lobulated margins measuring 14 × 10 × 8 cm was found adherent to the left kidney and left renal hilum inferiorly and with multiple neovascularisations from the aorta medially. Anteriorly mass was found to be displacing the splenic hilum and pancreas with preserved planes. Few para-aortic lymph nodes were present, which were excised along with the tumor. The postoperative period was uneventful. The patient recovered well and was discharged on the 5th postoperative day.

His histopathology report was a surprise. The histopathology showed well-encapsulated, non-circumscribed, malignant mesenchymal neoplasm disposed in intersecting fascicles. Individual tumor cells were plump and spindle-shaped with vesicular chromatin, conspicuous nucleoli, and eosinophilic cytoplasm with occasional atypical mitosis with surrounding peritumoral necrosis. There was no neurovascular invasion or perinuclear invasion with circumferential resection margin-free. All lymph nodes were free from tumor invasion (FNCLCC–Grade II).

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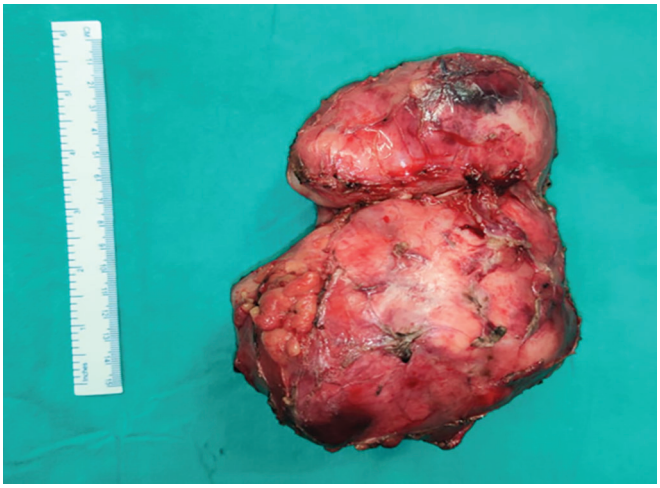

Fig. 1: Postoperative photograph of the resected specimen

Immunohistochemistry (IHC) showed SMA positivity, SOX10 negativity, and Desmin negativity with a Ki-67 index of 40%

The final histopathology was reported to be PAL.

Discussion

Leiomyosarcoma, a malignant tumor arising from smooth muscle cells, is an uncommon neoplasm, especially when found in the adrenal gland. The adrenal gland is not a typical site for leiomyosarcoma development, making primary leiomyosarcoma of the adrenal gland an exceedingly rare entity. The association of leiomyosarcoma with human immunodeficiency virus is well known. About 50% of reported leiomyosarcoma have been associated with human immunodeficiency virus.[3] However, this patient did not have a human immunodeficiency virus.

The clinical presentation of primary leiomyosarcoma of the adrenal gland is nonspecific, often mimicking other adrenal tumors or presenting as an incidental finding on imaging studies. Symptoms, when present, may include abdominal pain, weight loss, palpable mass, and hormonal abnormalities such as Cushing's syndrome or hyperaldosteronism. However, these symptoms are not specific to leiomyosarcoma and can be attributed to various adrenal pathologies.[4]

The diagnosis of primary leiomyosarcoma of the adrenal gland relies on a combination of clinical, radiological, and histopathological findings. Imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) play a crucial role in identifying adrenal masses and assessing their characteristics. However, definitive diagnosis requires histopathological examination of biopsy or surgical specimens, which typically reveals spindle-shaped cells with nuclear atypia and mitotic activity consistent with leiomyosarcoma (Fig. 1).[5]

Treatment of primary leiomyosarcoma of the adrenal gland involves a multimodal approach, including surgery, chemotherapy, and radiation therapy. Surgical resection remains the cornerstone of treatment, aiming for complete tumor excision with negative margins whenever feasible. Adjuvant chemotherapy and radiation therapy may be considered to improve local control and reduce the risk of recurrence, although their efficacy in leiomyosarcoma remains limited.[6] There are no clear guidelines regarding preoperative management with chemotherapy for PAL. However, in cases of metastatic or inoperable leiomyosarcoma, anthracycline or gemcitabine-based chemotherapy regimen is recommended.[7] In cases of inoperable tumors, preoperative identification of PAL is necessary as the management of adrenocortical carcinoma and PAL is different in terms of chemotherapy regimens.

The prognosis of primary leiomyosarcoma of the adrenal gland is generally poor, with a high propensity for local recurrence and distant metastasis. The rarity of this tumor limits the availability of large-scale studies assessing prognosis and treatment outcomes. However, overall survival rates are often dismal, highlighting the aggressive nature of this malignancy and the need for early detection and comprehensive management.

Conclusion

Primary leiomyosarcoma of the adrenal gland is a rare and aggressive malignancy that poses diagnostic and therapeutic challenges. Early recognition and prompt intervention are essential for optimizing patient outcomes, although prognosis remains guarded despite advances in treatment modalities. Preoperative differentiation from adrenocortical carcinoma is necessary. Further research is warranted to elucidate the molecular mechanisms underlying leiomyosarcoma pathogenesis and identify novel therapeutic targets to improve patient survival.[8]

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