Adrenal leiomyosarcoma: a rare clinical entity

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Abstract

Adrenal leiomyosarcomas are very rare tumors of mesenchymal origin. We report a 61-year-old male case, with an incidentally diagnosed adrenal mass. MRI revealed a heterogeneous 10cm mass, potentially malignant and the patient underwent an open left radical adrenalectomy. Microscopically, spindle cell type neoplasia was seen. Immunohistochemically, the tumor cells were positive for smooth muscle actin and desmin and the final diagnosis was a well-differentiated primary adrenal leiomyosarcoma. During his 2-year follow-up appointment he had multiple metastases and progressive disease, for which he is still receiving chemotherapy.

Introduction

Tumors arising from the adrenal gland have a significant heterogeneity and arise from either the medulla or the cortex of the adrenal gland. Malignant tumors are rather uncommon and usually manifest as malignant pheochromocytomas and adrenocortical carcinomas. In most cases, malignant tumors are rather aggressive and survival is poor if not detected and treated at an early stage. This is a case presentation of a patient operated for a tumor with preoperative malignant characteristics in which postoperatively an adrenal sarcoma was revealed.

Case presentation

A 61-year-old male presented with a left retroperitoneal mass located above the left kidney, which was incidentally diagnosed on a kidney stone disease follow up. The patient did not complain of any symptoms. Physical examination was unremarkable and a CT scan was performed showing a left adrenal mass measuring...
10x8x8cm with low density characteristics and minor uptake of contrast. MRI was also performed and depicted a heterogeneous mass with low signal intensity on T1-weighted images and high on T2, with no loss of signal intensity in out-of-phase T1 GRE sequence, accompanied by extracapsular nodules consistent with metastatic lymph nodes. The tumor was highly suspicious for adrenal malignancy and the patient was staged with a chest CT which excluded metastatic disease.

The tumor was further biochemically evaluated for hormonal hypersecretion but was proven to be non-functional. The patient underwent left open radical adrenalectomy due to the high possibility of adrenocortical carcinoma (Figure 1).

The pathologic specimen was rather large, 13x11x6.5 cm and weighed over 550gr. The tumor was lobulated and on macroscopic examination had regions with cystic degeneration and necrosis. Normal adrenal tissue was recognized solely in the periphery (Figure 2). Microscopically, spindle cell type neoplasia was seen, accompanied by intermediate nuclear atypia. Mitotic index was 7 mitoses per 10 high power fields. Interestingly, neoplastic cells were in contact with the adrenal vein. The tumor cells were immunohistochemically positive for smooth muscle actin and desmin and negative for S-100, synaptophysin, chromogranin, calretinin and pancytokeratin, while the Ki-67 proliferative index was over 75%. The final diagnosis was a well-differentiated primary adrenal leiomyosarcoma. Finally, satellite nodules of the same histological characteristics were also seen adjacent to the tumor.

Discussion

Adrenal leiomyosarcomas are exceptionally rare with only around 40 cases reported in the English literature. They suggest very aggressive tumors, of mesenchymal origin and are thought to derive from the smooth muscle wall of the central adrenal vein and its tributaries. They are almost always operated on the basis of malignant appearance in preoperative imaging with the clinical and radiographic suspicion of non-secreting adrenocortical carcinoma. In most cases the final diagnosis is established postoperatively, as they do not have specific tumor markers or a specific pattern of imaging characteristics. Adrenal leiomyosarcomas have distinct histological and immunohistochemical
characteristics that differentiate them from other more common adrenal pathologies.

Radical surgery seems to be the cornerstone of treatment. However, even in the absence of preoperative metastases, these tumors show a high incidence of disease progression and post-operative metastatic spread \(^{5,6}\). Patients have a very poor prognosis with the longest reported survival being 36 months \(^7\), while most patients present with recurrence within the first year after resection. Despite the aggressive nature and due to the rarity of this disease there are no data available for the implementation of adjuvant post-operative treatment modalities \(^{8,9}\).

Disclosure

The authors have no conflict of interest to disclose. Patient Consent has been obtained by the patient for publication of these data.