

Primary adrenal leiomyosarcoma

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Abstract

Leiomyosarcomas of adrenal origin occur infrequently, always present as huge abdominal masses, and are associated with poor prognoses when other organs are invaded. Radiologic images of small adrenal leiomyosarcomas have not been published. This report presents magnetic resonance images of a 3-cm left adrenal leiomyosarcoma from a 49-year-old male. Ten months after adrenalectomy, the patient was alive without tumor recurrence.

Key words: Adrenal gland—Leiomyosarcoma—Magnetic resonance imaging

Primary adrenal leiomyosarcomas are extremely rare. Only nine patients [1–9] and two autopsies [10] have been reported in the literature. Most of the clinically diagnosed tumors described in these reports were larger than 10 cm in diameter. To our knowledge, images of primary adrenal leiomyosarcomas smaller than 5 cm in diameter have never been described. This report describes a male patient with a 3-cm primary left adrenal leiomyosarcoma and presents magnetic resonance imaging (MRI) of that tumor.

Case report

A 49-year-old man had left flank pain for 6 months. Ultrasound revealed a left abdominal mass. MRI (Magnetom Sonata, Siemens Medical Solutions) showed a round, left suprarenal tumor with heterogeneously low intensity on T1-weighted images, high intensity on T2-weighted images, and marginal enhancement after gadolinium diethylenetriamine pentaacetic acid administration (Fig. 1). Neither venous thrombosis nor lymphadenopathy was found. The tumor was subsequently removed by laparoscopic surgery, and histologic examination revealed a leiomyosarcoma of the left adrenal gland. The microscopic appearance was that of a

hypercellular tumor with a haphazard arrangement of spindle, oval, or rounded cells intermingled with bizarre tumor cells. After immunohistochemical staining, the tumor was found to be positive for desmin and negative for creatine kinase, myoglobin, S-100, CD34, CD117 (C-kit), and HMB-45. Ten months after surgery, the patient was alive and exhibited no symptoms or evidence of tumor recurrence.

Discussion

Primary smooth muscle tumors arising from the adrenal glands are rare and are believed to originate from the smooth muscle wall of the central adrenal vein and its branches. In almost all clinically reported cases to date, these tumors are larger than 10 cm in diameter. Patients presenting with such tumors range from 30 to 68 years of age. Females and males are approximately equally affected, and tumors occur in the right and left adrenals to approximately the same extent. With invasive diseases that include venous thrombosis, adjacent organ invasion, and distant metastases, the prognosis is extremely poor. Although tumor responses to radiation and chemotherapy are limited, early radical resection is associated with a better outcome. However, identification of the tumor at an early stage is difficult because symptoms are usually not present and because clinically applicable tumor markers are not currently available.

In the case described in this report, the patient presented with left flank pain. The tumor was therefore identified at a relatively early stage. Few MRI scans of primary adrenal leiomyosarcoma have been reported, and images of smaller (<5 cm) tumors are virtually nonexistent. The MRI evidence for extensive necrosis, in the 3-cm tumor described in this report indicates that such small tumors can be extremely aggressive in nature.

In conclusion, the imaging characteristics of adrenal leiomyosarcomas are indistinguishable from those of adrenal cortical carcinomas and metastatic cancers. However, adrenal cortical carcinomas are usually functional and capable of corticosteroid or sex steroid secretion. An early stage, centrally necrotic, and non-functioning adrenal tumor occurring in the absence of a

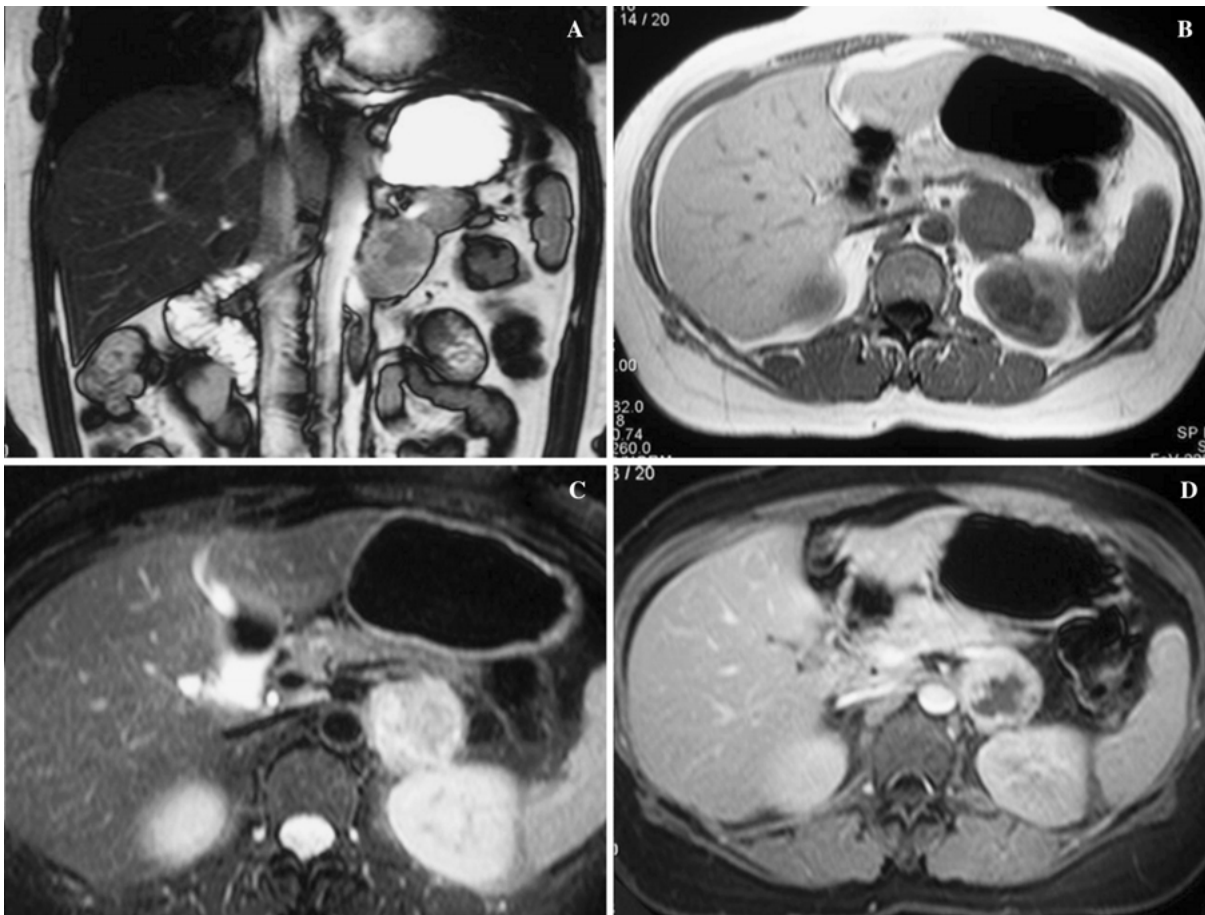


Fig. 1. Magnetic resonance images of a 3-cm left adrenal leiomyosarcoma. **A** T2-weighted, balanced, steady-state, free precession sequence (true fast imaging with steady state free precession [FISP]; repetition time [TR]/echo time [TE]/flip angle [FA] = 4.3 ms/2.1 ms/72 degrees) in coronal section shows a heterogeneous mass in the left suprarenal area. **B** T1-weighted spoiled gradient echo sequence (two-dimensional fast low-angle shot; TR/TE/FA = 150 ms/1.8 ms/70

degrees) in axial section displays a well-defined hypointense mass on the left adrenal gland. **C** T2-weighted turbo spin-echo sequence (TR/TE/FA = 2300 ms/94 ms/150 degrees) demonstrates a hyperintense tumor. **D** After contrast medium administration (gadolinium diethylenetriamine pentaacetic acid), the peripheral portion of the tumor is well enhanced and shows central necrosis.

primary malignancy should alert the physician to the possibility that a leiomyosarcoma is present.

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