Primary adrenal schwannoma with catecholamine hypersecretion

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Schwannomas are benign tumors of the nerve sheath Schwann cell or nerve cell sheath, so the tumor is also called neurilemmoma. Although this tumor can involve any place where Schwann cells are present, it arises predominantly in the head and neck region or the flexor surfaces of the extremities. Schwannomas may also occur in the retroperitoneum, and they account for approximately 5% of the primary neoplasms in this area [1]. However, primary schwannomas of the adrenal gland are extremely rare. Primary adrenal schwannomas occur most often as a rare type of incidentaloma with no evidence of hormonal hypersecretion [1–8]. When adrenal neoplasia with catecholamine hypersecretion is found, pheochromocytomas/paragangliomas need be differentiated. If pheochromocytomas/paragangliomas are overlooked or improperly treated, the clinical outcomes almost invariably prove fatal [9]. However, this case is of interest in that schwannoma arising from the adrenal gland with catecholamine hypersecretion, and its clinical presence and imaging features, must be carefully evaluated and further diagnosed.

A 41-year-old woman presented with 2 months history of left upper quadrant abdominal discomfort. Her past medical or surgical history was not significant. Physical examination revealed left upper quadrant and left flank tenderness on deep palpation, and no palpable masses were identified. The results of hormonal laboratory tests found that cortisol levels were elevated (adrenaline 569 ng/l, norepinephrine 483 ng/l). Serum aldosterone, electrolytes, urinary metanephrine, and vanillylmandelic acid were within the normal range. Multi-detector computed tomography (MDCT) examination detected a well-defined mass in the left adrenal gland, measuring approximately 7.7 × 6.4 × 7.1 cm. Computed tomography (CT) images showed heterogeneous density of the mass with dot-like calcification (Figure 1). The left kidney and pancreas in the vicinity were compressed. No sign of bony or visceral involvement was revealed. Local excision was performed under anesthesia. A well-demarcated tumor in the left adrenal gland was identified and entirely removed without complication. Histopathologic examination confirmed that the tumor exhibited Antoni A areas composed of well-defined fascicles of spindle cells and loosely organized Antoni B areas. Immunohistochemical staining demonstrated that the tumor was positive for S-100 protein (Figure 2). Based on these findings, the final diagnosis was benign...
adrenal schwannoma. Her catecholamine level normalized immediately after the operation. Nineteen months after the excision, no recurrence or metastases have been reported.

Schwannoma is a neural sheath tumor arising from the Schwann cell or nerve-cell sheath. Although the tumor can involve any place where Schwann cells are present, involvement of the adrenals is uncommon [1–4]. Histologically, most adrenal neoplasms arise from constituent cortical cells, but adrenal schwannomas are thought to originate from nerves in the adrenal medulla. The hypothesis is supported because the adrenal medulla is innervated by two groups of myelinated nerve fibers: one is derived from the sympathetic trunk or vagus nerve; another is from the phrenic nerve. The tumors are usually benign and slow-growing, but rarely may be malignant. Most malignant neurilemmomas are associated with von Recklinghausen’s disease (VRD) or other types of neurofibromatosis [2, 3].

Clinically, adrenal schwannoma is usually asymptomatic, but some patients can present with either abdominal or flank pain. Laboratory studies are typically unremarkable. When adrenal tumors secrete catecholamines, pheochromocytomas and paragangliomas should be considered for differential diagnosis. Pheochromocytomas and paragangliomas are tumors arising from sympathetic and parasympathetic paraganglia, which can be located in the adrenal medulla or in extra-adrenal abdominal, intrapelvic, or thoracic locations [10, 11]. Additionally, adrenal medulla hyperplasia may produce symptoms similar to those of pheochromocytoma. Although schwannomas are usually nonsecreting tumors, those tumors may occasionally have endocrine function. So far, to the best of our knowledge, only one case of retroperitoneal schwannoma with endocrine function has been reported in the literature. Hori et al. [7] reported a 66-year-old woman with asymptomatic retroperitoneal schwannoma, but her serum noradrenalin and vanillylmandelic acid levels were increased remarkably. Our case is extremely rare owing to the adrenal schwannoma with serum adrenaline and norepinephrine overproduction confirmed by laboratory examination.

Imaging plays a key role in the diagnosis and evaluation of an adrenal mass. Although some patients may present with clinical symptoms secondary to the mass effect of tumor, most of them are discovered incidentally by imaging examina-
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Conflict of interest

The authors declare no conflict of interest.

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In conclusion, the current study demonstrates, for the first time, that a case of adrenal schwannoma with catecholamine overproduction is an extremely rare retroperitoneal neoplasm. Clinical and morphologic diagnosis of primary adrenal schwannoma may be difficult. Although extremely rare, schwannoma should be considered in the differential diagnosis of primary adrenal lesions, especially in patients with catecholamine hypersecretion.

Acknowledgments

Jianglong Hou and Lizhi Zhang contributed equally to this work and should be considered co-first authors.