Laparoscopic Resection of a Retroperitoneal Ancient Schwannoma: A Case Report and Review of the Literature

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Abstract. *Schwannomas are benign tumors that arise from neural sheath Schwann cells. Solitary benign schwannomas are generally located in the head and neck, often along the cranial nerves, and are a particularly rare neoplasm among tumors of the retroperitoneal space. A 67-year-old man undergoing a general health check was incidentally found to have a mass beside the left kidney. Computed tomography (CT) revealed a cystic adrenal mass located cephalad to the left kidney. Retroperitoneoscopic resection of the retroperitoneal mass was performed. The pathological diagnosis was retroperitoneal benign schwannoma consisting of mixture of a Antoni-A and -B type cells. We report a case of retroperitoneal schwannoma and discuss eleven previous reports treated by laparoscopic surgery in the English literature. Laparoscopic resection may be useful because a retroperitoneal schwannoma, which is commonly localized and hypovascular, can easily be dissected from the adjacent tissues.*

Schwannomas are tumors of the nerve sheath that usually exhibit benign behavior and are found in all organs and in the nerve trunk, although rarely in the retroperitoneum, since these comprise only 3% of all schwannomas (malignant and benign combined) (1). Ancient change in a schwannoma (ancient schwannoma) is a relatively uncommon histological variant characterized by degeneration. The term ancient schwannoma was first suggested by Ackerman and Taylor (2) in a review of 48 neurogenic tumors of the thorax. They reported 10 cases that showed similar features of a typical neurilemmoma, but were distinctive because significant portions of these tumors were composed of only a few cells within a hyalinized matrix. They clarified that these features occurred in the schwannomas of long duration and hence coined the term 'ancient schwannoma'. This type is characterized by diffuse areas of hypocellularity, focal accumulations of hyaline material, and fatty degeneration (3). We report a case of retroperitoneal ancient schwannoma that was discovered incidentally in a patient by abdominal ultrasonographic examination and resected successfully by laparoscopic surgery.

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Case Report

A 67-year-old man had a left renal mass that was found incidentally on abdominal ultrasonography (US) during a medical health checkup and was referred to our institution for additional evaluation. His medical and family histories were unremarkable. At the initial examination, his blood pressure was 132/80 mmHg. Biochemical results, carcinoembryonic antigen, carbohydrate antigen 19-9 and interleukin-2 receptor were all within the normal ranges, including serum catecholamines. CT revealed a well-defined round 80×75×60 mm cystic mass with a rim of soft tissue in the left adrenal region. There were neither calcifications nor evidence of infiltration. Enhancement of the peripheral rim of soft tissue was seen after intravenous administration of contrast material (Figure 1). Magnetic resonance imaging (MRI) showed a smooth marginated low-intensity mass on T1-weighted image and high-intensity on T2-weighted image. Adrenal scintigraphy using ^{131}I-adosterol and ^{123}I-metaiodobenzylguanidine (MIBG) did not show increased activity in the region of the mass. A tentative preoperative diagnosis of a malignant tumor of undetermined origin, schwannoma, nonfunctioning pheochromocytoma or paraganglioma was considered on the basis of the imaging and laboratory results. The patient underwent left adrenalectomy, during which the surface of the tumor was found to be smooth and slightly adherent to the left kidney. The resected specimen was oval and firm; it measured 83×65×45 mm and weighed 147 g. The tumor was well demarcated from the adrenal gland and the cut surface of the excised specimen exhibited red-brownish cyst formation and a yellowish-white rim (Figure 2). Pathological examination showed whorls and interlacing fascicles of schwannoma spindle cells, along with alternating

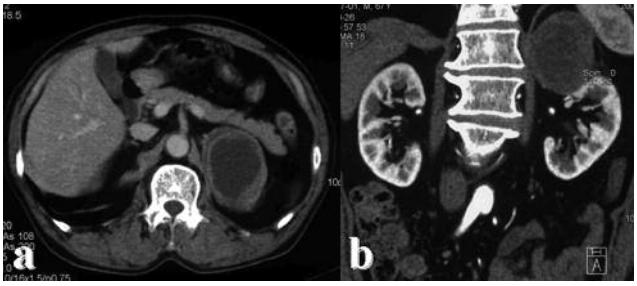

Figure 1. *Computed tomography showed a well-defined round, 80×75×60 mm cystic mass with a rim of soft tissue in the left adrenal region. Slight enhancement of the peripheral rim of soft tissue was seen after intravenous administration of contrast material. (a: Axial image, b: coronal image).*

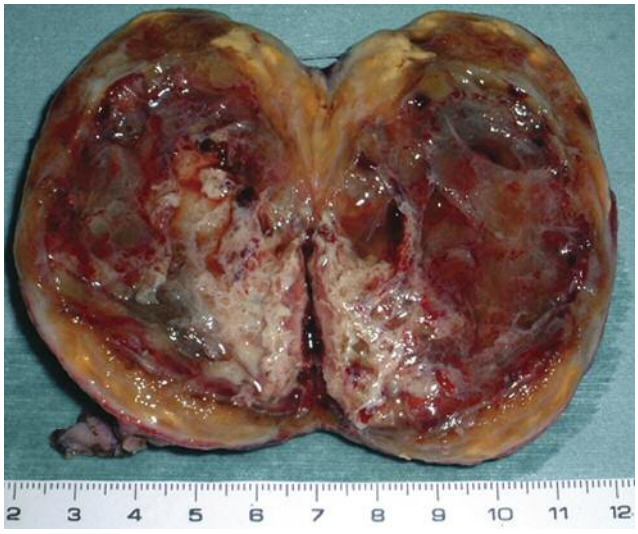

Figure 2. *The cut surface of the excised retroperitoneal mass showed red-brownish cyst formation and a yellowish-white rim.*

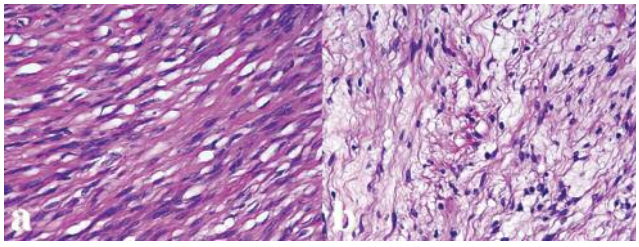

Figure 3. *Pathological findings (×400) of the retroperitoneal mass revealed compact cellular lesion (a: Antoni type A tissue) and loose, hypocellular, myxoid lesions with microcystic spaces (b: Antoni type B tissue).*

Antoni-A and -B patterns with areas of hyalinization and fibrinoid degeneration (Figure 3). Some of the tumor cells were nuclear atypical, but lacked mitotic figures. Immunohistochemical analysis for S-100 was positive. These findings were consistent with the diagnosis of benign ancient schwannoma. Furthermore, no adrenal tissue was found in the specimen, which suggested that the tumor had an extraadrenal origin.

Discussion

Schwannomas are tumors of the nerve sheath of Schwann cells and may arise along the course of any myelinated nerve, with the vestibulocochlear nerve being the most frequent site. These tumors are usually encapsulated, present as a solitary mass and have a benign course (4). These tumors can vary from firm, solid masses to fluctuant cysts. The occurrence of a retroperitoneal schwannoma is rare, comprising only 1-3% of all schwannomas (1) and only 1% of all retroperitoneal tumors (5). Retroperitoneal schwannomas occur most commonly in patients between forty and sixty years of age, with a male:female ratio of 2:3 (6). Because they are usually slow-growing and asymptomatic, retroperitoneal schwannomas are often found incidentally, as in the present case (7). Although patients with a retroperitoneal schwannoma are usually asymptomatic, some present with vague and nonspecific abdominal or back pain.

Laboratory data for schwannoma are generally not remarkable or contributory to its diagnosis. Histologically, schwannomas consist of compact cellular lesions (Antoni type A tissue) and loose, hypocellular, myxoid lesions with microcystic spaces (Antoni type B tissue). In addition, almost all schwannomas show intense immunohistochemical staining for S-100 protein, confirming the neuroectodermal origin of the tumor cells (8).

An ancient schwannoma is a rare variant first described by Ackerman and Taylor (2) and is characterized by degenerative changes and diffuse hypocellularity. A benign schwannoma is depicted as a well-defined and inhomogeneous low-density mass on CT scan images, while MRI is able to demonstrate the tumor location and its relationship to surrounding structures. MRI of benign schwannomas typically shows hypointensity on T1-weighted images and hyperintensity on T2-weighted images; although this was noted in only 57% of previous cases (5).

A definitive diagnosis is based on pathological, histological and immunohistochemical findings. A preoperative diagnosis is very difficult to make because of the lack of typical imaging features (US, CT and MRI). However, radiological imaging is helpful in treatment planning because it provides information about tumor size, location and possible invasion of other structures. Fine-needle aspiration biopsy may be useful if Schwann cells are found in the sample, but tissues for diagnosis are often inadequate and may be misleading because of cellular pleomorphism in degenerated areas that can be interpreted as malignancy (9). Surgical excision is considered the

treatment of choice for these tumors, which respond poorly to radiation and chemotherapy. Furthermore, there is the possibility of local recurrence and malignant change in benign schwannomas despite prior benign diagnosis (10). Therefore, it is very important to remove the tumor completely. Resection of the tumor under laparoscopic surgery may be performed more completely and easily than under open surgery, because the surgical view under laparoscopy is magnified. To our knowledge, to date, only eleven reports of retroperitoneal benign schwannomas resected by laparoscopic surgery have been reported in the English literature. No cases were definitively diagnosed as schwannoma during preoperative examinations. From available data, the average size of tumors, operation time and intraoperative blood loss including our case were 45 mm (19-80) in maximum diameter, 174 minutes (90-300) and 150 ml (15-310), respectively. There were no severe operative or postoperative complications in almost all cases. Our case had an uneventful postoperative course and was discharged from our hospital on the tenth postoperative day.

In accordance with previous reports, our report shows that laparoscopic surgery is very useful, safe and feasible for the removal of retroperitoneal schwannoma for diagnosis and treatment with minimal invasiveness and rapid postoperative recovery.

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