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A Case of Primary Retroperitoneal Schwannoma Arising in the Hilus Renalis

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A 35-year-old-woman was admitted to hospital after a medical check-up revealed signs of liver dysfunction. On physical examination, a palpable mass was found on the left side of her upper abdomen. Abdominal ultrasound (US) and computerized tomography (CT) showed a 10 cm cystic mass compressing the left renal vein adjacent to the mesenterium and inferior to the pancreas. Magnetic resonance imaging (MRI) demonstrated a retroperitoneal mass with solid and cystic components. The surgical and pathohistological findings enabled a final diagnosis of benign retroperitoneal schwannoma. Nerve sheath tumors in the retroperitoneum are infrequent. Here, a very rare case of a benign retroperitoneal schwannoma arising in the hilus of the left kidney is presented.

Key words: retroperitoneal schwannoma, retroperitoneal tumor, benign schwannoma, ancient schwannoma, neurilemoma

Introduction

The most frequent sites for benign solitary schwannomas are the extremities or the head and neck region. Occurrence in the retroperitoneal region is rare, and a qualitative diagnosis is difficult because of this rarity. This report describes a very rare case of a retroperitoneal schwannoma arising in the hilus of the left kidney.

Case

A 35-year-old-woman was admitted to hospital after a medical check-up revealed signs of liver dysfunction in May 2003. On physical examination, a large round mass was palpable in her left upper abdominal quadrant. Her past medical history and family history suggested no concerns of malignancy. An abdominal ultrasound (US) examination showed a large cystic mass with multiple separated septa (Fig. 1). A computerized tomography (CT) examination revealed a well-defined, round, 10 cm mass with calcification compressing the left renal vein adjacent to the mesenterium and inferior to the pancreas (Fig. 2). Magnetic resonance imaging (MRI) demonstrated a large retroperitoneal mass with solid and cystic components (Fig. 3). No other abdominal abnormalities were evident. An arteriogram demonstrated tumor vascularity arising from the branches of the superior mesenteric artery (Fig. 4). Serum tumor marker (carcinoembry-

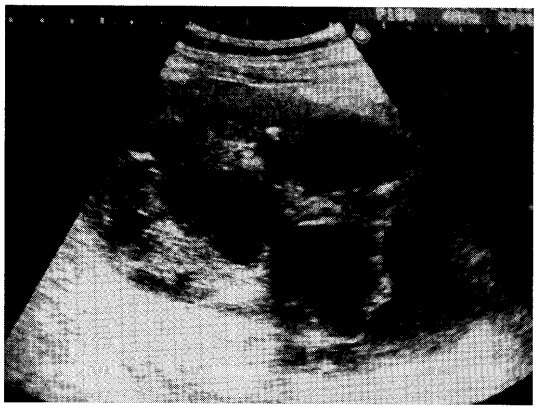

Fig. 1 Ultrasound image showing a large cystic mass with multiple separated septa

onic antigen, carbohydrate antigen 19-9) levels were all within the normal limits.

The patient underwent a laparotomy, and a well-encapsulated mass was found anterior to the left renal vein and fixed to the mesenterium; the mass was located inferior to the pancreas and anterior to the left kidney. The excised mass, 7 × 8 × 6 cm in size, was encapsulated, and the cut surface showed some irregular yellow areas and contained multicystic lesions with central areas of hemorrhage and necrosis (Fig. 5).

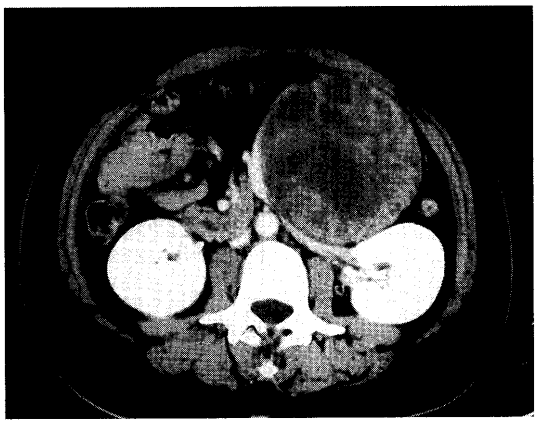

Fig. 2 Abdominal CT image showing a well-defined, round, 10 cm mass with calcification compressing the left renal vein

Histopathologically, a thick connective tissue capsule surrounding mixed solid and loose tumor tissue was observed. The texture of the solid tissue was composed of interwoven bundles of long bipolar cells. In some tissue masses, the cells had a palisading arrangement, with their nuclei in a well-organized pattern. The loose area contained pleomorphic cells. This finding corresponds to mixed-type Antoni A and B tissue, which is characteristic of benign schwannomas (Fig. 6).

The patient had an uneventful postoperative course and has not experienced any other symptoms for 20 postoperative months.

Discussion

The retroperitoneum is flexible and not limited structurally; therefore, large, deeply situated tumors can develop asymptomatically. Such abdominal tumors or masses are often found only upon a physical examination. The most frequent initial complaint of patients with retroperitoneal tumors is abdominal pain. The second most common complaint is the discovery of a definite abdominal mass

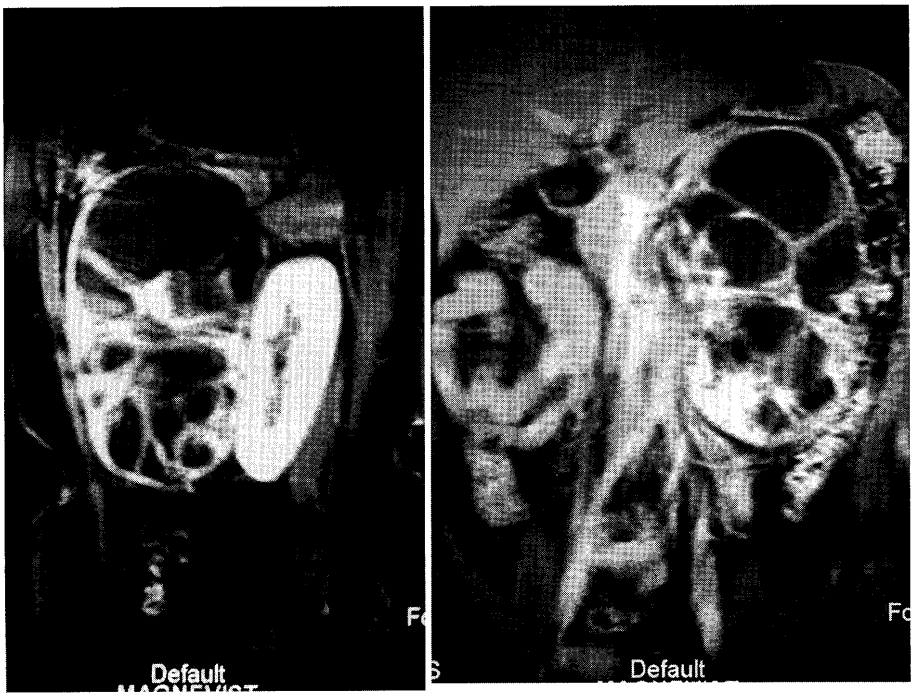

Fig. 3 MRI image showing a large retroperitoneal mass with solid and cystic components

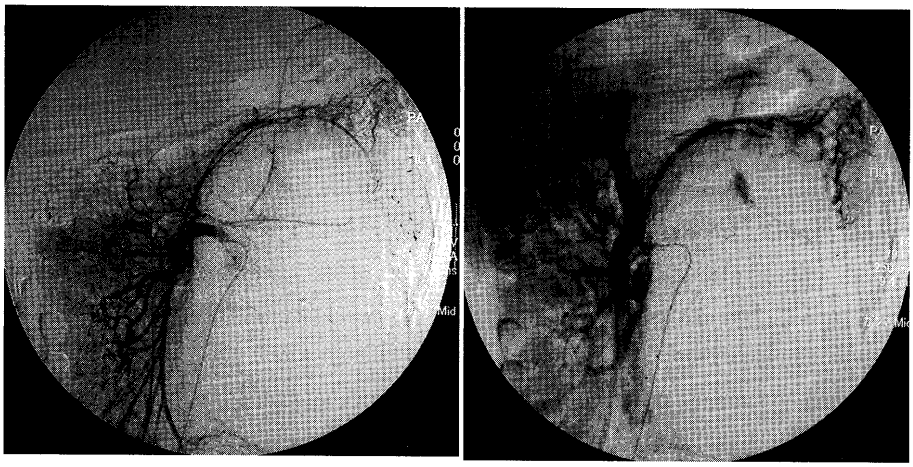

Fig. 4 Arteriogram showing tumor vascularity arising from the branches of the superior mesenteric artery

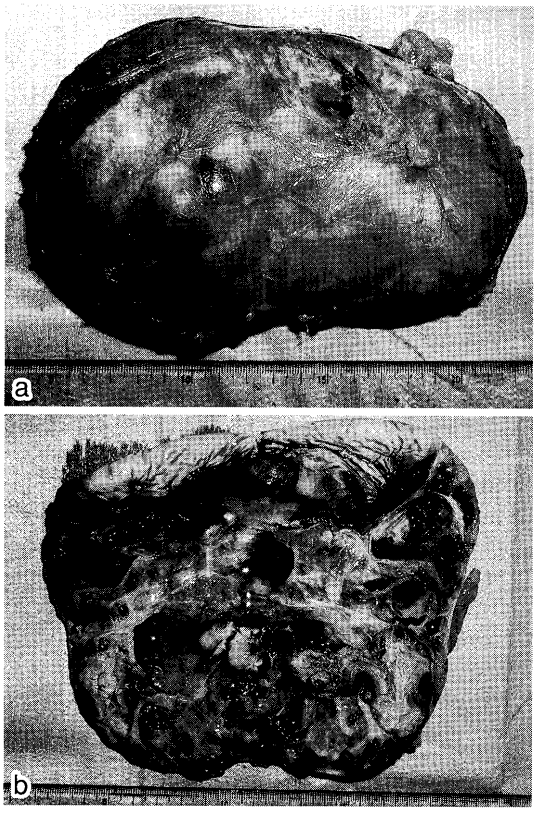

Fig. 5
a: The large retroperitoneal solitary schwannoma that was removed from the patient
b: Cut surface showing multicystic lesions with central areas of hemorrhage and necrosis

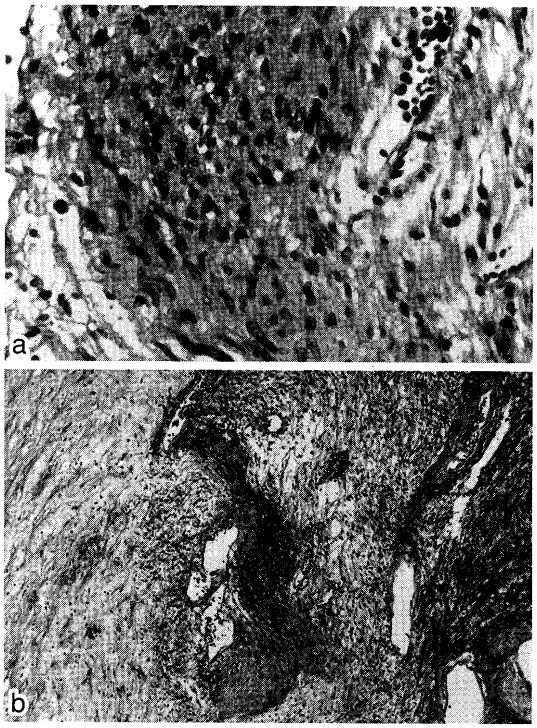

Fig. 6
a: Photomicrograph of a section showing an area of Antoni A tissue with palisading cells (HE×100)
b: Areas of Antoni A and B tissue (HE×40)

by the patients or an awareness that the patient's abdomen is becoming progressively larger. The most common genitourinary complaints are hematuria, dysuria, urgency, and frequency. The majority of these tumors are not tender upon palpation[1].

The most common sites for benign solitary schwannomas are the extremities or the head and neck region. These tumors are usually associated with von Recklinghausen's neurofibromatosis. Das Gupta reported that only 0.6% of these tumors are observed in the retroperitoneal region[2]. Further-

more, occurrence in the kidney region is extremely rare.

Schwannomas are rare tumors that originate from nerve sheaths of cranial or extremity nerves and are slow-growing benign tumors with a favorable prognosis. Small tumors are usually solid, but larger ones may become cystic[3]. Schwannomas often display degenerative changes, such as cystic formation, calcification, hemorrhage, and hyalinization[4]. Histologically, these tumors are composed of two types of tissue. In type A of Antoni tissue, the tissue is compact and composed of interwoven bundles of long bipolar cells. In some tissue masses, the cells have a palisading arrangement, with their nuclei in a well-organized pattern. In type B tissue, the tissue is loose-textured and the tumor cells are pleomorphic[3].

Although schwannomas constitute a small percentage of all retroperitoneal tumors, they should be included in the differential diagnosis of retroperitoneal masses. Imaging principles, including ultrasound and CT, are helpful in approximating the size, location, presence of invasion, and involvement of surrounding organs[5]. Both ultrasound and CT examinations are likely to show similar findings, consisting of a well-circumscribed complex mass with or without septa. MRI seems to provide more information for larger masses. On MRI, benign schwannomas have smooth margins and are isointense with muscle on T1-weighted images and hyperintense on T2-weighted images[6]. These modalities can help to characterize the lesion and may predict the presence of a primary retroperitoneal tumor but do not provide enough information to enable a specific preoperative diagnosis. CT-guided biopsy or fine needle aspiration may be helpful for a diagnosis only if the sample contains enough Schwann cells to be visualized macroscopically[7].

The preferred treatment for retroperitoneal schwannomas is surgery. Recently, a minimally invasive approach has been described for the excision of these tumors using an endoscope-assisted minilaparotomy[8]. The best management for retroperitoneal schwannomas is complete surgical excision because malignant transformation of schwannomas, although extremely rare, has been reported[9]. Postoperative follow-up using careful imaging studies is necessary to monitor the possible recurrence of this benign tumor[10].

The patient's postoperative course was uneventful, and she was discharged from hospital 15 days after surgery. She is doing well 20 months after surgery, with no evidence of recurrence. A longer follow-up period may be needed to assess the possible recurrence of these tumors[6].

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腎門部に発生した後腹膜神経鞘腫の1例

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症例は35歳女性．健診で肝機能障害を指摘され当院を受診し，触診で左季肋下に腫瘤を触知し病変を指摘された．US，CTで左腹部，膵の下方と腸間膜に接し約10cm大の石灰化と隔壁構造を伴う腫瘤を認め，左腎静脈を圧排していた．血管造影で上腸間膜動脈の分枝に栄養血管を認めた．以上の画像所見から左腎静脈近傍に発生した後腹膜腫瘍と診断し，開腹手術を施行した．腫瘍は腎静脈の前面に接し，結腸間膜，膵下面，左腎前面に癒着していたが，腎静脈を温存し腫瘍を摘出した．摘出標本は7×8×6cmで厚い線維性の被膜に覆われ，出血と壊死を伴った．病理組織は紡錘型の核を有する紡錘形細胞が粗に増殖し，部分的には棚状配列を呈し密に増生する部分が混在していた．腫瘍細胞に明らかな異型はなく核分裂像は認めず，良性の後腹膜神経鞘腫と診断した．後腹膜の神経鞘腫は稀であり，若干の文献的考察を加えて報告する．