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Case Report

Giant retroperitoneal schwannoma: a case report

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Abstract: Schwannomas are rare tumors originating from the Schwann sheath of the peripheral or cranial nerves. They are usually benign tumors and in 95% of the cases they originate from the peripheral nerves. Occurrence in the retroperitoneum is rare. We report a case of giant retroperitoneal schwannoma. The patient was a 71-year old female. She was admitted in with the complaints of finding abdominal mass for 8 years and emaciation for 1 year. CT scan showed a 15 cm × 11 cm mass located between the liver and the upper pole of the right kidney. After adequate preparation, the patient underwent surgery. During the procedure, we found a 15 cm × 11 cm × 6 cm mass located between the right liver, right kidney, adrenal gland and vena cava and adhered to them. The pathologic diagnosis was schwannoma.

Keywords: Retroperitoneal schwannoma, neurilemmoma, schwannoma, retroperitoneal tumor, retroperitoneum

Introduction

Schwannomas are nerve sheath tumors that usually affect the head, neck and the flexor surfaces of the extremities. It is rare to find a schwannoma in the retroperitoneal cavity. Schwannomas are usually encountered in young to middle-aged adults. Most of the schwannomas are asymptomatic and found accidently. Microscopically, positive expression of S-100 and negative expression of CD 34 is valuable for diagnosis. Here we report a case of giant retroperitoneal schwannoma and review the literature.

Case report

The patient was a 71-year old female. She was admitted in with the complaints of finding abdominal mass for 8 years and emaciation for 1 year. The abdominal mass was detected 8 years ago by B-ultrasound in local hospital and was 1 cm in diameter. The patient did not received any treatment and didn't pay attention to it. But she lost 4 kg in the last year so she was rechecked the B-ultrasound and found the mass had become very large, up to 15 cm in diameter. She denied abdominal pain, distention, chills, fever, or diarrhea. Her medical and family history was noncontributory. Physical examination: temperature 37.0°C, pulse 78 bpm, respiration 20 bpm, blood pressure 152/81 mmHg. The heart and lungs were normal. The abdomen was flat and soft, with no tenderness or enlarged liver, spleen, or mass was palpable. Murphy's sign was negative. Laboratory and radiology findings: Biochemistry and Routine blood examinations were in normal range. Computed tomography (CT) scan showed a 15 cm × 11 cm mass located between the liver and the upper pole of the right kidney, which was considered from liver, right kidney or right adrenal gland (**Figure 1**). Stones of gallbladder and common bile duct were also found. After adequate preparation, the patient underwent surgery. During the operation, we found a 15 cm × 11 cm × 6 cm mass located between the right liver, right kidney, adrenal gland and vena cava and adhered to them (**Figure 2A**, **2B**). The right renal vessels and inferior vena cava represented transposition and deformation because of the compression of mass. There was a 1 cm stone in the gallbladder and a 0.8 cm stone in the common bile duct. We resected the tumor and did cholecystectomy and remove the stone in the common bile duct. On gross examination, the mass was a 15 cm × 11 cm × 6 cm cucurbit with soft margins and a medium

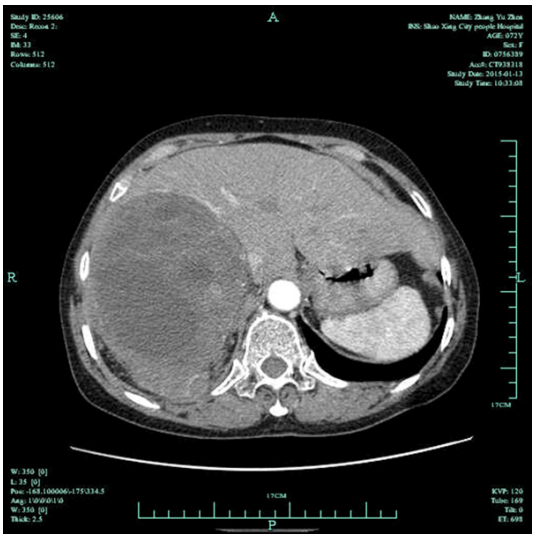

Figure 1. Mass in the CT scan.

texture (**Figure 2C**, **2D**). On microscopic exam, it was confirmed schwannoma with cystic degeneration and calcification. Immunohistochemistry: CD117 (-), CD34 (-), S-100 (+), SMA (-), Vim (+) (**Figure 3A**, **3B**). The pathologic diagnosis was schwannoma. The patient covered well and was discharged.

Discussion

The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumors and malignant neoplasms that can be either primary or metastatic lesions. The occurrence of a retroperitoneal schwannoma is uncommon comprising only 1-3% of all schwannoma and almost 1% of retroperitoneal neoplasms [1]. Schwannoma is tumor arising from Schwan cells of the peripheral nerve sheath. It usually present in cranial and peripheral nerves in the head and neck or in the upper extremites [2, 3]. However schwannomas may appear in the posterior mediastinum, and more rarely in the retroperitoneum. Retroperitoneal region is a rare location for schwannomas. Since the retroperitoneal space is rather large and flexible, most of the patients don't have any symptom. As a result, the diagnosis of retroperitoneal schwannomas is often delayed, and the lesion reaches a significant size and late stage at the time of diagnosis [4]. Some of the patients complaints of abdominal pain and distention. Besides, secondary hypertension, hematuria, and renal colic have also been reported depending on the location of the lesions. In our case, the mass was found early. But due to the absence of symptoms, the treatment was delayed which made the surgery much more complicated and difficult because of the large size of the tumor. It reminds us that close follow-up is very necessary regardless of the tumor size.

Retroperitoneal schwannomas usually affect adult patients aged 20 to 50 years [3, 5]. The preoperative diagnosis of retroperitoneal schwannomas is difficult. Imagelogical examination has certain diagnostic value but lack specificity. In our case, the mass was so large that it occupied much space of the abdominal cavity and closely adhered to the adjacent vessels and organs which made the origination of the mass very difficult to diagnose by CT scan or B-ultrasound. Even during the operation, we thought the mass was from right kidney or adrenal gland when separated from the liver at the beginning. Pathologically, it is a well-encapsulated lesion demonstrating specific Antoni A/B areas. Positive expression of S-100 and negative expression of CD 34 is valuable for diagnosis [6, 7]. Schwannomas are usually benign, slow-growing, encapsulated tumors, and are rarely malignant. Malignant schwannomas are frequently associated with von Recklinghausen syndrome or other types of neurofibromatosis [8].

Complete surgical excision is of first choice for schwannomas since schwannomas are not sensitive to radiotherapy and chemotherapy [7]. Subtotal resection may be performed to minimize surgical risk and preserve surrounding vital structures and some doctors believe that simple enucleation or partial excision is sufficient [2]. In our case, the mass was so large that the right renal vessels and inferior vena cava represented transposition and deformation because of the compression of mass. It was difficult to resect the mass directly for the risk of neighbor vessel and organ injury. Some doctors suggest completed divestion from the capsule. But most of the retroperitoneal schwannomas are large and cystic degenerated. In our case, the capsule was very thin. It was difficult to devest the mass completely and would cause hemorrhage. Besides, malignance could not be exclude only by imaging or frozen section. According to our experience, we con-

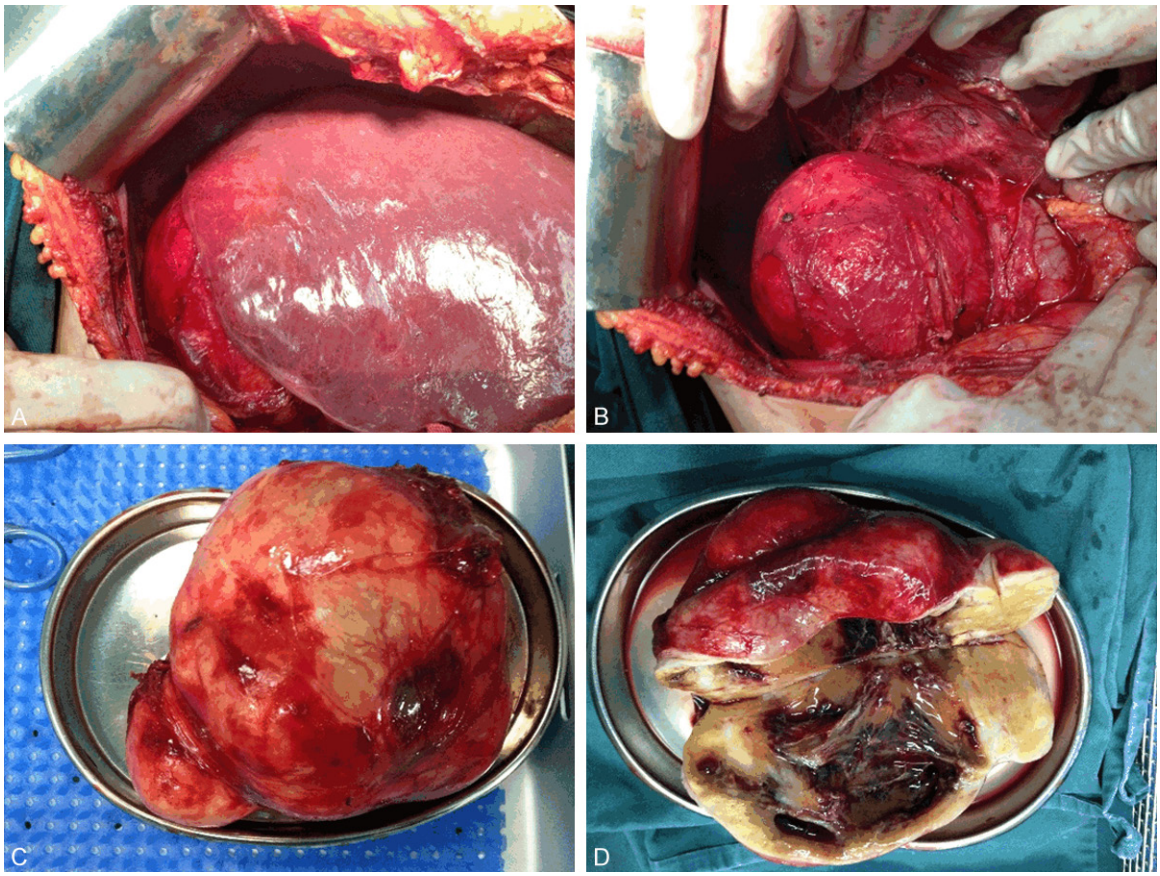

Figure 2. Gross specimen. A, B. Mass during operation. C, D. Mass after resection.

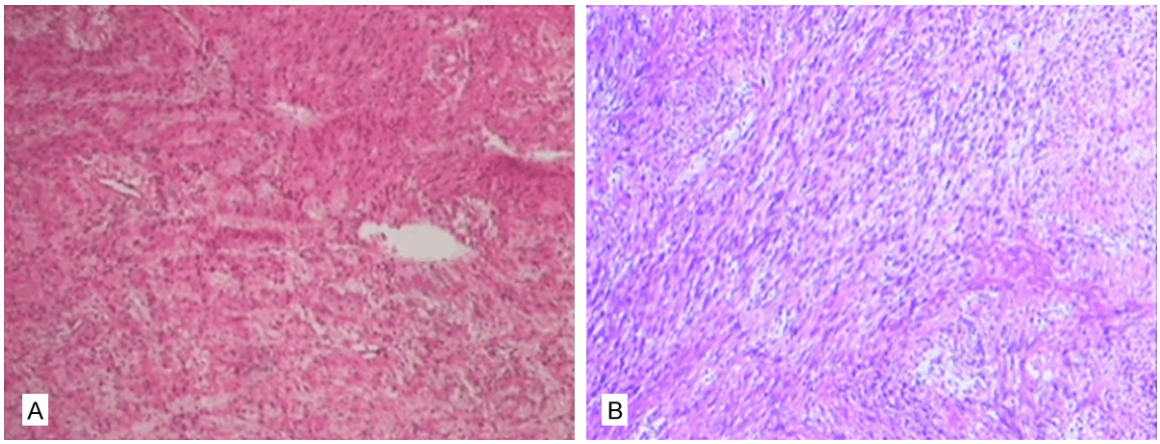

Figure 3. Immunohistochemical examination showed CD117 (-), CD34 (-), S-100 (+), SMA (-), Vim (+).

ducted completed resection with capsule after separation of the mass. We firstly exposed the mass. Then we dissociate the vessels which closely adhered to the mass. Because the right renal vessels and inferior vena cava are displaced and deformated, the process should be very attentive. Traction belt may be very useful because it can separate the vessels from the mass so that accidentally injury could be avoid. To those important nerves adhere to the mass, meticulous anatomy should be made to avoid injury. The surgical difficulty is associated with the size of the mass and neighbor adhesion, so we strongly recommend early operation.

The prognosis of benign schwannomas is good and the most frequent complication is recurrence, probably due to incomplete excision, which is reported in 5-10% cases [9]. In malignant schwannomas, adjuvant chemotherapy or radiotherapy may have marginal added benefit. Some authors advocate induction chemotherapy for this condition [10]. Our case is benign and follow-up was suggested.

Disclosure of conflict of interest

None.

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