RETROPERITONEAL SCHWANNOMA — A CASE REPORT AND REVIEW OF THE LITERATURE—

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Abstract: Reported herein is a rare case of retroperitoneal schwannoma in a patient without von Recklinghausen's disease. This patient was free of symptoms, and the tumor was found by chance during a periodical physical examination. Complete excision of the tumor was performed, and as of this writing, 36 months after the operation, the patient is free of the disease.

In addition, a review was carried out on the 121 cases of retroperitoneal schwannoma reported in the Japanese literature from 1981 to 1992. In this series, malignant tumors were seen in 26.4% of all cases, and when this tumor was found in association with von Recklinghausen's disease, the malignancy rate increased to 52.9%. Thirty seven percent of the patients with malignant tumors had died at the time of writing of their case report, and their mean survival period was 18 months after the first surgical treatment. All the patients with benign tumors underwent complete resection and had a good prognosis.

Index Terms

schwannoma, retroperitoneal tumor

INTRODUCTION

Schwannoma is generally believed to arise from the peripheral nerves of the extremities. It is commonly locate in the head and upper or lower extremities[1),2)]. Schwannomas occurring in the retroperitoneal space excite surgical interest in view of their rarity(1.7-3% of all schwannomas), lack of a definitive diagnostic test and poor prognosis in malignant cases[1),2)].

We report herein a case of benign retroperitoneal schwannoma. A literature survey of this rare retroperitoneal neoplasm is provided, and its clinical characteristics, diagnosis and treatment are also discussed.

CASE REPORT

A 53-year-old Japanese female was admitted in April 1990, because a non-tender abdominal mass was incidentally found during a periodical physical examination. The medical history and family history were negative for malignancy or stigmas of von Recklinghausenes disease. The physical examination revealed a firm, immovable, and well-defined round mass in the right upper quadrant of the abdomen.

Laboratory evaluation of this patient revealed normal blood chemistries. An abdominal X-ray film showed a soft tissue mass density on the upper pole of the right kidney. Ultrasound

examination(US)revealed a well defined, hypoechoic and solid, 5 cm in diameter, mass located between the right kidney and inferior vena cava. Computed tomography(CT)scan disclosed a huge retroperitoneal mass and its cystic nature. Magnetic resonance imaging(MRI)scan delineated the mass to be low-intensity signals on T1 weighted images and high intensity signals on T2 weighted images(Fig. 1). Aortography and selective L2 lumbar arteriography demonstrated a large hypervascvlar mass fed by the L2 to L5 lumbar arteries. Venacavography showed displacement of the vena cava to the left and extraluminal compression.

Under the diagnosis of retroperitoneal neurogenic tumor, laparotomy was performed. An oval, firm, well-encapsulated and dull whitish smooth tumor was found occupying in the retroperitoneal space. Although the tumor adhered slightly to the psoas muscle and compressed the right ureter, it was completely extirpated. There was no gross evidence of invasion to

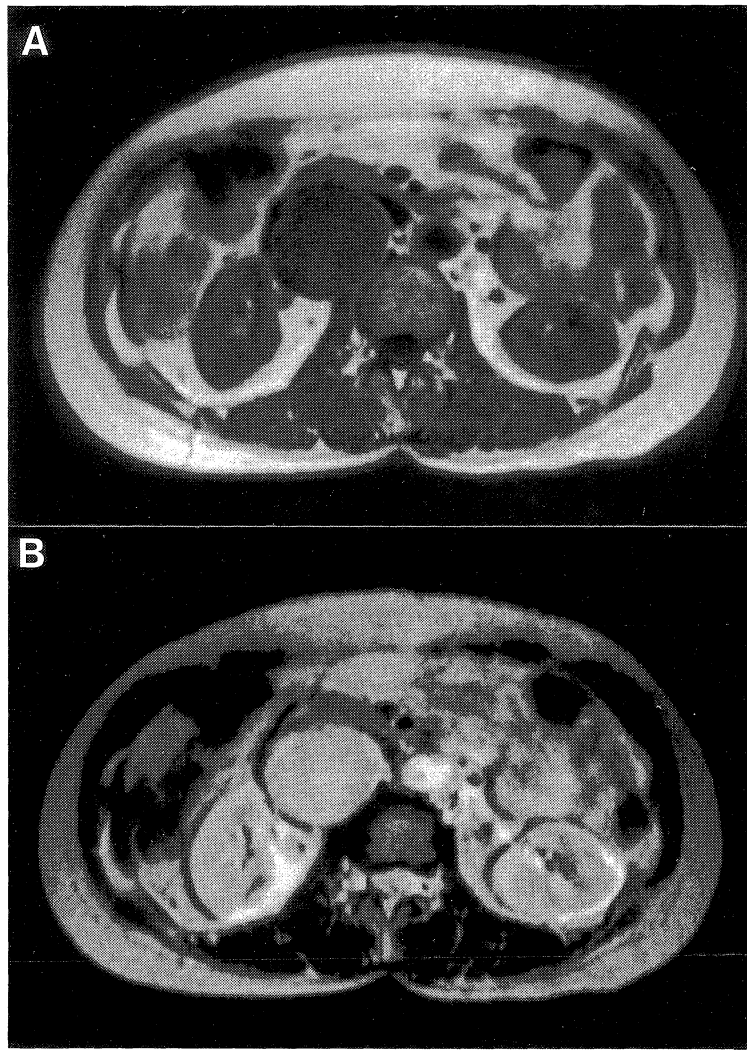

Fig. 1. MRI scan displayed the low intensiy mass on T1 weighted image (A) and high intensity mass on T2 weighted image (B).

adjacent organs or regional lymph-node swelling. The resected specimen measured 5.5x5x5 cm and weighted 120 g. On cross section, the mass contained a predominantly solid area with multicystic changes but there was no evidence of hemorrhage or necrosis.

Microscopically, the tumor tissue was composed of spindle-shaped cells with oval nuclei. Mitoses were rare. In most places the cells formed typical palisading arrangement with their nuclei in a well-organized pattern suggestive benign schwannoma, Antoni A type(Fig. 2). The post-operative course was uneventful, and the patient has been free of the disease for 36 months.

DISCUSSION

Schwannoma, also known as neurilemmoma, is a primary nerve sheath tumor that usually arises from the peripheral nerves of the extremities. The most common location is the head, followed by in descending order, the upper extremities, the lower extremities and trunk[1),2)]. The retroperitoneal space is a rare site with a reported incidence of 1.7-3% of all schwannomas[1),2)]. Furthermore, schwannomas constitute approximately 4% of all retroperitoneal tumors[3)]. Although several cases have been reported in the Japanese literature, review articles of this neoplasm are very scarce. We reviewed the 121 cases, including the present case, of retroperitoneal schwannomas reported in the Japanese literature from 1981 to 1992.

This series comprised 58 men and 63 women. At the time of diagnosis, the patients ranged in age from 11 to 81 years with a median age of 53 years(Fig. 3). Of 112 cases in which the histopathological findings were clearly described, 80 cases(71.4%)were benign and 32(28.6%) were malignant. Association with von Recklinghausen neurofibromatosis(NF1), which is a common hereditary disorder, was reported in 14(11.6%)cases. They comprised 7 men and 7 women with a median age of 34(ranging from 11 to 59 years). Heterotopic presentation of

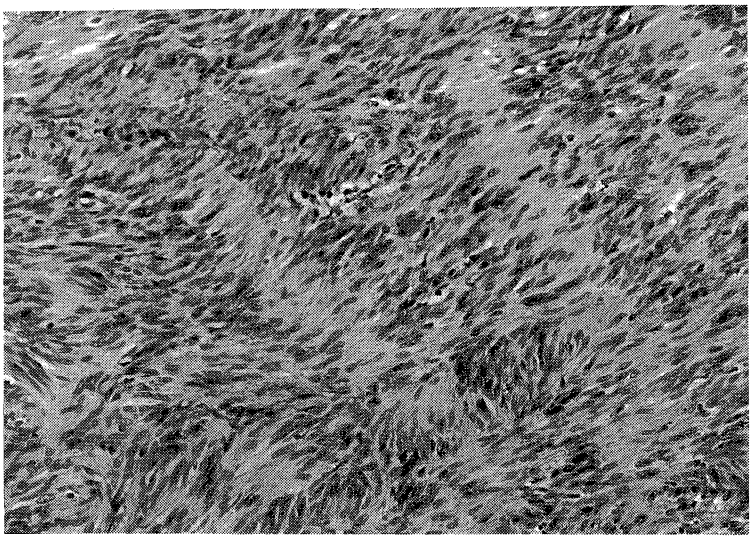

Fig. 2. Histopathologic examination of the resected tumor shows a typical palisading arrangement with their nuclei in a well-organized pattern, Antoni A type.

schwannoma in the head, neck, mediastinum or extremities occurred in 6(43.0%)and 7(6.5%) cases in with or without NFl, respectively. The incidence of malignancy was higher(64.2%) in the cases with NFl than in those without NFl(23.5%). It has been unclear why schwannomas in NFl-positive patients are clinically more aggressive than those in NFl-negative patients[4]. Recently this is considered partly due to the result of mutation of the NFl gene, although the precise role of the NFl gene in the tumorigenesis as yet remaines uncertain[5].

The most common initial symptom was palpable abdominal mass(38.0%)(Table 1). Abdominal pain or loin pain, which may be vague or colicky and is sometimes difficult to localize, was noted in 22.3%. Peripheral nerve symptoms such as pain in the legs occurred more frequently (9.2%)than in other types of retroperitoneal tumors(0.3%)[6]. In 19 patients(15.7%)the tumor was detected incidentally during periodical medical examinations as in our case. Genitourinary symptoms, changes in bowel habits and gross hematuria were rare. Ascites, lower extremity edema and intestinal obstruction or ileus could but are late manifestations of the disease.

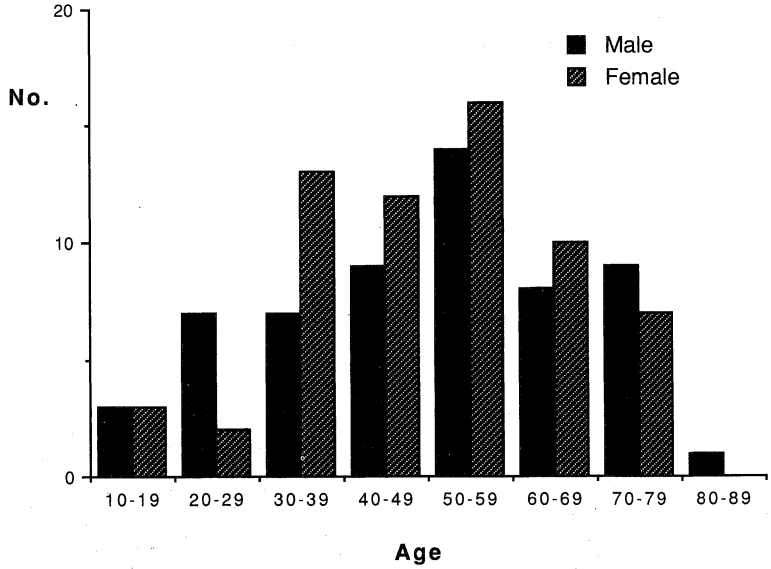

Fig. 3. Age and sex distribution of retroperitoneal schwannoma.

Table 1. Initial symoptoms of retroperitoneal schwannoma. Numbers in parenthesis are percentages.

Palpable abdominal mass	46	(38.0)
Abdominal pain	27	(22.3)
Leg pain	11	(9.1)
Genitourinary symoptom	5	(4.1)
General fatigue	5	(4.1)
Leg edema	3	(2.5)
Gross hematuria	2	(1.7)
Others	3	(2.5)
Symptom free	19	(15.8)
total	121	

On physical examination, an abdominal mass can be palpated in the majority of the cases (60.6%), otherwise a pelvic mass is rare(4.2%). Laboratory findings were all within the normal range, except for an increase of CEA in one case.

As for radiological examinations, US, CT and angiography should be performed when retroperitoneal tumor is suspected. In this series these examinations were perfored in 59.5%, 80.1%, and 73.5% of the cases, respectively. In our series, US examination showed a cystic mass with some solid foci in 86.1% of the cases and CT showed a well delineated low density and heterogenous soft tissue mass in 77.3% of the cases, with calcification as a rare finding(8.6%). Although these cystic changes may be important distinct diagnostic features in schwannoma, they are not a sign of malignancy. They were present in 68.2% of malignant schwannomas, but were also found in 72.5% of the benign ones. This was also the experience of Takatera et al[7]. These cystic changes are considered due to certain vascular wall changes. MRl, which was performed in 15.4%, is a new promising modality in the preoperative assessment. Multiple axial, sagittal, and coronal planes that are not available by CT, can be imaged[6),8)].

Gross inspection of this tumor showed a well encapusulated solid tumor with some degree of cystic changes in 84.0%, those were hemorrage or necrosis. The tumor diameters ranged from 4 to 31 cm(mean 10.5 cm).

Regarding the treatment of retroperitoneal schwannoma, including metastatic area, most authors agree that complete surgical resection with tumor-free margins is a prerequisite for potential long-term survival. Adjacent organs including the kidney, pancreas, psoas muscle, or vertebra attached to the tumor should be considered potentially invaded and should be resected *en bloc* with the specimen. Among the 32 malignant cases, data were sufficient for analysis in 30 cases. At the time of surgery, 4 patients had metastatic disease. Each of the patients had intra-abdominal metastasis with liver metastasis, liver metastasis, left atrial metastasis and brain metastasis. Twenty-five patients underwent complete resection and 3 patients underwent partial resection or debulking. Two patients were subjected to diagnositic laparotomy. In the complete resection group, 12 patients(48.0%)required adjacent organ resection to avod leaving gross residual disease. In general, schwannomas are relatively benign tumors ; however, some may become malignant and metastasize distantly through the bloodstream or lymphatics, or both[3),4)]. Unfortunately, 6 patients undergoing successful removal of all gross tumor, developed a local disease by 3 years(1 month-3 years). Thirty seven percent of the patients with malignant tumors had died at the time of writing of their case report, and their mean survial period was 18 months after the first surgical treatment. All the patients with benign tumors underwent complete resection and had a good prognosis.

Because of the rarity of retroperitoneal malignant schwannoma, there is still no generally accepted mode of adjuvant treatment. Only 9 of 80 patients with malignant tumors received adjuvant chemotherapy which was a regimen consisting mainly of adriamycin, vincristin and cyclophosphamide. Adjuvant radiation therapy was employed in 2 cases after complete resection. They had no impact on survival, and a meaningful statintical analysis was very

difficult because of the small number of patients, different modalities and lack of controls. Similarly, world-wide experience with adjuvant therapy is still very limited, and further studies on more patients are necessary.

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