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Retroperitoneal schwannoma

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Abstract

Background: Retroperitoneal schwannomas are rare, benign tumors. The aim of this study is to present our surgical experience with 7 such tumors.

Methods: Between 1989 and 2004, 7 patients with pathologically proven retroperitoneal schwannomas were reviewed retrospectively. **Results:** There were 6 male patients and 1 female patient, with a mean age of 43 years (range, 23 to 58 years). Two patients were symptomatic and presented with abdominal discomfort, and none of the patients suffered from von Recklinghausen's disease. All the patients underwent computed tomography scanning, which showed a heterogenous retroperitoneal mass, 4 of which were thought to arise from the adrenals. In 2 patients, calcification was seen in the tumors. All 7 of the patients had a preoperative diagnosis of a retroperitoneal tumor including 3 patients who were thought to have adrenal neoplasms (1 patient had a diagnosis of an adrenal neoplasm excluded on magnetic resonance imaging). Laparotomy and complete excision of tumors were performed in all the patients, and there was no morbidity or mortality. The schwannomas had a mean maximum diameter of 7.3 cm (range, 4 to 14 cm), and they were all benign. At a mean follow-up of 17 months (range, 3 to 48 months) postresection, all the patients remained free from recurrence.

Conclusion: Retroperitoneal schwannomas are rare tumors that are difficult to diagnose preoperatively. Radiologic findings are usually nondiagnostic. The treatment of choice is complete surgical excision. © 2006 Excerpta Medica Inc. All rights reserved.

Keywords: Retroperitoneal schwannoma; Neurilemmoma; Schwannoma; Retroperitoneal tumor; Retroperitoneum

Nerve sheath tumors are a subclass of soft-tissue neoplasms that include benign and malignant schwannomas and neurofibromas [1]. Schwannomas are found most commonly in cranial and peripheral nerves, and occurrence in the retroperitoneum is rare, comprising approximately 3% of all schwannomas [1]. Retroperitoneal schwannomas are usually larger and have a higher tendency to undergo spontaneous degeneration and hemorrhage compared with their counterparts arising in the head and neck or extremities, which are usually solid and small [2]. We report our experience with 7 histologically proven cases of retroperitoneal schwannomas with special emphasis on the clinical presentation, preoperative evaluation, and the management.

Methods

Between 1989 to 2004, 96 patients underwent surgical resection for a retroperitoneal tumor or mass at the Department of Surgery, Singapore General Hospital, Singapore, Singapore. Of these patients, 7 (7.3%) had a pathologically proven retroperitoneal schwannoma and were reviewed retrospectively. One of these cases (case 5) has been reported previously [3]. Their case notes, radiologic reports and pathological reports were reviewed restrospectively. The diagnosis of a schwannoma was made based on histological examination showing Schwann cells with Antoni A and B regions with positive staining for S100 protein.

Results

The patients' demographic, preoperative, surgical, and pathological data are summarized in Tables 1 and 2. There

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Table 1
Patients' demographics, presentation, and radiologic findings

Case	Age/sex	Presentation	US	CT	MRI	Preoperative diagnosis
1	46/M	Rt flank discomfort	NA	Heterogenous enhancing retroperitoneal mass with calcification	NA	Retroperitoneal tumor
2	46/M	Incidental	NA	Rt heterogenous adrenal mass displacing IVC. CT-guided biopsy, atypical cells	Round- to oval-shaped suprarenal retroperitoneal tumor not arising from adrenals. Uniform hypointensity on T1 and heterogenous hyperintensity on T2	Retroperitoneal tumor
3	28/M	Incidental (calcification on plain x-ray)	Rt suprarenal mass with calcification	Heterogenous-enhancing solid right adrenal mass with punctate calcification	NA	Rt nonfunctioning adrenal tumor
4	50/F	Incidental palpable mass	NA	Thick-walled retroperitoneal cystic mass with fluid level	NA	Retroperitoneal cyst
5	49/M	Incidental palpable mass	Retroperitoneal cystic mass	Heterogenous enhancing mass with septations arising from adrenal	NA	Rt adrenal tumor
6	58/M	Rt hypochondrial pain	Suprarenal retroperitoneal cystic mass	Heterogenous solid mass with lobulated outline	NA	Rt adrenal tumor
7	23/M	Incidental mass	Lt retroperitoneal echogenic mass	Heterogenous enhancing solid mass, well encapsulated	NA	Lt retroperitoneal tumor

Lt = left; Rt = right; IVC = inferior vena cava; NA = not applicable; M, male; F, female.

were 6 male patients and 1 female patient with a mean age of 43 years (range, 23 to 58 years). Only 2 patients were symptomatic and presented with abdominal discomfort. None of the patients had von Recklinghausen's disease. All the patients underwent computed tomography (CT) scanning, which showed a heterogenous retroperitoneal mass (Fig. 1), and 2 tumors showed calcification. Four of these tumors were thought to arise from the adrenals on CT scan. An adrenal tumor was excluded subsequently in case 2 using magnetic resonance imaging (MRI). In 2 patients, calcification was seen in the tumors. All 7 of the patients had a preoperative diagnosis of a retroperitoneal tumor including 3 patients who were thought to have adrenal neoplasms. However, none were correctly diagnosed preoperatively with a retroperitoneal schwannoma. Twenty-four-hour urinary catecholamines levels were normal in all 7 patients, thus excluding the possibility of a phaeochromocytoma.

Laparotomy and complete excision of tumors were performed in all the patients, and there was no morbidity or mortality. Although the tumors were frequently closely adherent to vital structures in the retroperitoneum such as the inferior vena cava, renal arteries, and veins, complete resection with preservation of these structures could be performed in all patients by careful dissection. None of the 7 tumors encountered involved major retroperitoneal nerves. Frozen section was performed in 3 patients and was diagnostic in 1 patient. The schwannomas had a mean maximum diameter of 7.3 cm (range, 4 to 14 cm), and these

were all benign. Pathological examination showed degenerative changes such as cystic degeneration in 3 tumors, hyalinization in 5, and hemorrhage in 4. At a mean follow-up of 17.0 months (range, 3 to 48 months) postresection, all the patients remained free from recurrence.

Comments

Retroperitoneal schwannomas are rare tumors. In a review of 303 patients with schwannomas, only 2 (0.7%) were found in the retroperitoneum [4], and in another study, 2 (2.6%) cases were found out of a total of 76 schwannomas [5]. Retroperitoneal schwannomas are usually solid encapsulated tumors that arise from the paravertebral region [2]. These are usually asymptomatic and are thought to grow slowly [2,6]. The tumors frequently attain a relatively large size before discovery because the retroperitoneum is flexible and nonrestrictive and may exhibit cystic degeneration, calcification, and hemorrhage [6]. Symptoms are usually nonspecific, and neurologic symptoms are rare as was evident in this series whereby the 2 symptomatic patients had vague abdominal pain and none of them had any neurologic complaints. The observations in this series whereby the tumors ranged in size from 4 to 14 cm in maximum diameter and only 2 (33.3%) were symptomatic also lends support to the hypothesis that these tumors are slow growing.

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