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Analysis of benign retroperitoneal schwannomas: a single-center experience

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Conflict of Interest: The authors declare that they have no conflict of interest.

Abstract:
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Methods Retrospective analysis of 67 patients with retroperitoneal schwannoma admitted to Peking University International Hospital from 2015 to 2021.
Results 67 patients presented with retroperitoneal schwannomas, 37 cases had no obvious clinical symptoms. 62 cases were completely excised, 5 cases were subtotal resection, 7 cases were combined with organ resection. The intraoperative blood loss was 300ml (20-9000ml), the tumor maximum size was 9cm (2.5-26cm), post-complication occurred in 6 cases (9.0%). Compared with abdominal retroperitoneal tumors, pelvic retroperitoneal tumors had larger tumor volume, more bleeding, higher proportion of block resection, and longer postoperative hospitalization time ($P<0.05$). The residual mass progressed slowly in 5 patients with subtotal resection, and no obvious malignant transformation occurred.
Conclusion Complete resection of schwannoma can achieve a good long-term prognosis. Patients with residual tumor after surgery progress slowly and rarely become malignant. We recommend early resection after the discovery of a pelvic retroperitoneal schwannoma.
Keywords Schwannoma; Retroperitoneal neoplasms; Postoperative complications

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Background

Schwannomas are neuroectodermal tumors rich in proliferating Schwann cells. The common sites are the head, neck and extremities. The retroperitoneal site is less common, accounting for about 0.7-3.0% of all schwannomas[1, 2]. Schwannomas are the most common benign soft-tissue

tumors occurring in the retroperitoneum[3], and the complete surgical excision remains the gold standard for the management of these tumors[4]. Due to the large retroperitoneal space, retroperitoneal schwannomas are often detected late, and some tumors grow for a long time and are large in size. In order to improve the clinical understanding, diagnosis and treatment of this disease, we present the clinical, radiological features and clinical efficacy of 67 patients with retroperitoneal schwannoma.

Methods

From April 2015 to October 2021, a total of 67 patients with retroperitoneal schwannoma underwent surgical treatment at Peking University International Hospital. All patients underwent preoperative discussions with a multidisciplinary team (MDT), MDT consists of doctors from surgery, radiology, pathology, and medical oncology, histology was confirmed by two dedicated sarcoma pathologist specialists. The Patient demographics, clinical presentation, imaging examinations, diagnostic assessment, surgical outcomes and long-term outcomes were retrospectively analyzed, and the prognosis was followed up through outpatient clinics and telephone calls until February 15, 2022.

Statistical analysis

SPSS 23.0 statistical software was used for data analysis, normally distributed data was expressed as the Means ± SD and analyzed by independent t-tests. Non-normally distributed data was expressed as the median (range) and analyzed by Mann-Whitney tests, univariate analysis was performed by chi-square test. $P<0.05$ indicated that the difference was statistically significant.

Results

Demographic data and clinical characteristics

From April 2015 to October 2021, 67 patients presented with retroperitoneal schwannomas in our center. There were 24 males and 43 females, with a male-to-female ratio of 1.0:1.8. The median age was 47 years (range, 15-74 y). 37 patients (55.2%) with tumors detected by physical examination or identified incidentally during investigations for unrelated symptoms. 12 patients (17.9%) had abdominal pain and discomfort. 8 patients (11.9%) had low back pain, 5 patients (7.5%) had lower limb pain, 5 patients (7.5%) presented with a palpable mass. None of the patients had a history of neurofibromatosis.

Findings of CT imaging features

All patients in this group underwent enhanced CT and 9 cases underwent MRI examination. Through preoperative discussion, 12 cases of schwannomas were correctly judged□other 10 cases were judged to be neurogenic tumors, 7 cases were misdiagnosed as malignant tumors, the remaining cases were uncertain. CT images showed the tumors were round or oval/irregular, 22 cases of cystic degeneration, 10 cases of necrosis, and 9 cases of calcification (Fig 1). 41 tumors located near the spine, psoas, adrenal region, or kidneys in the retroperitoneal region of the abdomen, and 26 tumors were located in the pelvis. 64 patients had a single tumor, 2 patients had multiple tumors in the same location of the abdominal retroperitoneum, and 1 patient had multiple tumors in both the abdominal and the pelvis retroperitoneum at the same time (Fig 2). Tumors damaged lumbar spine or sacrococcygeal bone in 3 cases (Fig 3,4), encapsulated iliac arteriovenous vessels in 3 cases, and wrapped around the celiac trunk vessels in 2 cases (Fig 5,6). Four cases of interventional contrast were performed, of which 2 were preoperatively invasive embolism.

Fig 1

Fig 2

Fig 3A

Fig 3B

Fig 4A

Fig 4B

Fig 5

Fig 6

Surgical results

All patients underwent surgery, no surgical deaths. There were 59 cases of open surgery, 8 cases of Laparoscopic surgery. 62 patients underwent complete resection, 5 patients underwent subtotal resection. Subtotal resection was performed in 3 cases because of wrapping around the celiac trunk or iliac vessels, and in 2 cases because it extended into the intervertebral foramen or resulted in irregular bone destruction. The median intraoperative blood loss was 300ml (20-7600ml). There were 6 cases of preoperative compression with hydronephrosis and 5 cases of gastrointestinal obstruction. 19 patients were placed preoperatively with ureteral stents, and 7 patients underwent combined multi-organ resection and reconstruction，including 3 cases of partial ureter resection, 2 cases of sigmoid colectomy, 2 cases of partial lumbar cone resection, 2 cases of partial sacral coccyx resection, and 1 case of partial pancreatic resection. Postoperative complications occurred in 6 patients, including 2 cases of intestinal obstruction, 2 cases of paralysis and discomfort in the lower extremities, 1 case of urinary fistula, and 1 case of postoperative bleeding. Due to lack of maneuvering space or proximity to important blood vessels, 9 cases of pelvic retroperitoneal tumors and 3 cases of abdominal retroperitoneal tumors underwent block resection. In this group, 26 tumors occurred in the pelvis retroperitoneal space and 41 occurred in

the abdominal retroperitoneal space, which were divided into two groups for comparison (Table 1). There were statistically significant differences between the two groups in terms of tumor maximum size, blood loss, proportion of block resection, and postoperative hospital stay（P<0.05）.

Table 1

Postoperative pathology

All pathological diagnoses were benign, the range of tumor diameter was 2.5-26cm, the average was 9.56 ± 5.27 cm, HE staining reveals a large number of spindle cells arranged in a cross-striped or beam-like arrangement, manifested as Antoni A region rich in Schwann cells and Antoni B region with low cell content, and positive immunohistochemical S-100 staining.

Follow-up

62 patients (92.5%) were followed up, no tumor recurrence was found from 3 to 68 months, and the survival rate at 1, 3, 5 years was 100%. 5 patients underwent subtotal resection, their follow-up time was 7-43 months, the residual tumors did not have obvious progression, no obvious deterioration was seen, and the close follow-up continued.

Discussion

Schwannomas originating from myelin sheaths are rare benign tumors composed of ordered, alternating cell-rich tracts (Antoni area A) and loose myxoid reticular areas (Antoni area B)[5]. Retroperitoneal schwannoma is more common in female patients, 69% of the patients in this group were female[2, 6]. Retroperitoneal schwannomas are rare, mainly from spinal nerve sheath cells, and tend to occur on both sides of the spine, medial of the psoas or iliopsoas, the anterior pelvic sacral region and other areas[7]. Because of the wide retroperitoneal space, some tumors tend to grow huge, the tumors in this group are up to 26 cm in diameter, and 27 cases with a diameter of more than 10

cm. About 50% of these tumors are clinically silent and discovered incidentally[8], and the symptoms are vague and nonspecific, some tumors are closely related to the nervous system, including low back pain or lower limb pain. Pelvic schwannomas are limited by the bony pelvis and can cause symptomatic compression of the ureters or rectum if they reach significant dimensions[9]. We place the ureteral stent through cystoscope before surgery to prevent damage to the ureter during surgery. Pelvic retroperitoneal tumor surgery was prone to bleeding, and for tumors with rich blood supply, we also used interventional embolization to reduce intraoperative bleeding.

Retroperitoneal tumors are recommended for preoperative discussion by the MDT team, who can improve diagnostic accuracy based on imaging and clinical features of schwannomas. Typical imaging is characterized by low-echo nodules with clear boundaries and uniform echoes, and degenerative changes tend to occur in the tumor center, such as calcification, bleeding, and cystic changes[10, 11]. These typical findings can help distinguish schwannoma from other retroperitoneal tumors. Schwannomas are also easily misdiagnosed as malignant, leading to overtreatment or unnecessarily enlarged range removal. 10.4% of the patients in this group were mistaken for malignancy before surgery, due to the large tumor range, erosion of bone tissue, and wrapping around blood vessels. Although the schwannoma is a benign tumor, it still has a certain degree of erosion and irregular bony destruction, this group has 2 cases of erosion of the lumbar spine (Fig 3), 1 case of erosion of the sacral coccyx (Fig 4), tumors are also easy to grow to the spine or sacrococcygeal foramen. There are also tumors in this group that are wrapped around the celiac trunk or iliac arteriovenous blood vessels (Fig 5,6), which may easily lead to intraoperative bleeding and residual tumor. Surgical design is also an important part of the MDT discussion. When operating retroperitoneal nerve sheath or nerve-associated tumors, monoportal approaches may

have limitations in exposure of tumor and surrounding structures, Christian[12] et al propose an algorithm for a modular strategy based on tumor location, size, spread, and suspicious tumor entity, which can better provide individualized treatment.

Retroperitoneal schwannomas need to be differentiated from neurofibromas, paragangliomas, and ganglioneuromas. Neurofibromas appear as round or irregularly shaped masses along the course of the nerves, with well-defined but lacking capsules, occasional cysts and hemorrhages, and heterogeneous enhancement on contrast-enhanced scans[13]. Paraganglioma is mainly located in the sympathetic chain on both sides of the spine, and may be accompanied by cystic degeneration, calcification and necrosis, and may be accompanied by paroxysmal hypertension[14]. The enhancement after enhanced scan is more obvious than that of schwannoma[15]. The ganglioneuroma tumor has a clear boundary, and the lesions may show pseudopodia-like or embedded growth. The density is mostly uniform. It may be accompanied by calcification or fatty degeneration, less cystic consolidation, and the enhancement scan is not obvious[16]. The indication for biopsy of a retroperitoneal schwannoma is controversial because in most cases the radiologic appearances are characteristic[6]. In this group, only 9 patients underwent preoperative puncture. After the benign tumor was identified, block resection was used to guide the huge tumor that is difficult to be removed in the pelvis. Needle biopsy is difficult to obtain accurate pathology, if it can't affect the comprehensive treatment (E.g, gastrointestinal stromal tumor, Ewing sarcoma, lymphoma, teratoma) or surgical methods, we do not think it is worth recommending.

Postoperative complication rates of retroperitoneal schwannomas are low, prognosis is good. Malignant transformation of schwannomas and local recurrence after resection is extremely rare[6]. Only a very small number of epithelial schwannomas have been analyzed to have malignant

changes[17]. No definitive schwannoma malignancy was found in this group of cases. The location, size, and relationship with surrounding tissues of the schwannoma can significantly affect the course and efficacy of the procedure. Large tumors in the pelvis are difficult to operate, 2 patients in this group had a long clinical observation of pelvic tumors, resulting in tumor enlargement and increased the difficulty of surgery. Due to the small space for pelvic manipulation and the close relationship between the tumor and the sacrococcygeal plexus and the blood vessels in the iliac, it is easy to cause difficult surgical operations and the risk of intraoperative bleeding. For patients with invasion of the lumbar spine and sacrococcygeal bone, we performed simultaneous dorsal approach cone surgery, followed by retroperitoneal tumor surgery to achieve complete resection. Residual tumors are often due to tumor enclosing blood vessels, prone to bleeding in the pelvis, and tumor intrusion into the intervertebral space. All patients with a small amount of residual tumors should be closely followed up, and during the follow-up process of our hospital, no obvious malignant changes have been found, consistent with related reports [18, 19]. In recent years, some schwannomas can also be removed by laparoscopy or robotic laparoscopy [20], the tumor volume of laparoscopic surgery in this group was less than 5cm.

In summary, accurate preoperative diagnosis is very important for surgical selection of schwannoma. Complete resection of schwannoma can achieve a good long-term prognosis. Subtotal resection, which minimizes surgical risk and preserves surrounding vital structures, can also achieve an acceptable prognosis, but requires close follow-up. Long-term observation of pelvic schwannoma requires caution, and we recommend early resection after the discovery of a pelvic retroperitoneal schwannoma.

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Table 1 Clinical data comparison of pelvic group with abdominal group

	pelvis group(n=26)	abdominal group(n=41)	Statistics	p-value
Age（y）	45.50 ± 17.28	44.29 ± 14.49	t=0.18	0.857
Tumor maximum size (cm)	12.24 ± 5.74	7.93 ± 4.27	t=3.24	0.002
Amount of bleeding（ml）	1000 (100-9000)	150 (20-6500)	z=3.06	0.002
Complications, n (%)	4 (15.4)	2 (4.9)	χ^2=2.154	0.142
Block resection, n (%)	9 (34.6)	3 (7.3)	χ^2=8.064	0.005
Postoperative hospital stay (d)	20.1 ± 7.5	14.7 ± 6.8	t=2.561	0.013
Laparoscopic resection, n（%）	2 (7.7)	6 (14.6)	χ^2=0.729	0.393