Analysis of Benign Retroperitoneal Schwannomas: A Single-center Experience

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Abstract

Background Retroperitoneal schwannomas are rare. The purpose of this study was to present our experience with the diagnosis and treatment of 67 such tumors.

Methods We retrospectively analyzed 67 patients with retroperitoneal schwannoma admitted to the Peking University International Hospital from 2015 to 2021.

Results Thirty-seven of the 67 patients had no obvious clinical symptoms. Complete excision was obtained in 62 and subtotal excision in 5 cases. In7 cases, tumor resection was combined with organ resection. The intraoperative blood loss was 300 ml (20–9,000 ml), the maximum size of the tumor was 9 cm (2.5–26 cm), and postoperative complication occurred in six 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 five patients with subtotal resection, and no obvious malignant transformation occurred.

Conclusion Complete resection of a retroperitoneal schwannoma can achieve a good long-term prognosis. Residual tumor after surgery progresses slowly and rarely become malignant. We recommend early resection after the discovery of a pelvic retroperitoneal schwannoma.

Keywords
- schwannoma
- retroperitoneal neoplasms
- postoperative complications

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 ~0.7 to 3.0% of all schwannomas.[1,2] Schwannomas are the most common benign soft-tissue tumors occurring in the retroperitoneum,[3] and 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. To improve the clinical understanding, diagnosis, and treatment of this disease, we present the clinical, radiologic features, and surgical results 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; consisting of doctors from surgery, radiology, pathology, and medical oncology). Histology was confirmed by two pathologists with special sarcoma knowledge. The patient

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demographics, clinical presentation, imaging examinations, diagnostic assessment, surgical outcomes, and long-term outcomes were retrospectively analyzed.

Statistical Analysis

SPSS 23.0 statistical software was used for data analysis, normally distributed data were expressed as the means ± SD and analyzed by independent *t*-tests. Non-normally distributed data were expressed as the median (range) and analyzed by the Mann–Whitney *U* test. Univariate analysis was performed by the chi-squared test. A $p < 0.05$ indicated that the difference was statistically significant.

Results

Demographic Data and Clinical Characteristics

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

Findings of CT Imaging Features

All the patients underwent enhanced computed tomography (CT) and nine cases underwent magnetic resonance imaging (MRI) examination. Through preoperative discussion, 12 cases of schwannomas were correctly judged; other 10 cases were judged to be neurogenic tumors and 7 cases were misdiagnosed as malignant tumors. The remaining cases were uncertain. CT images showed the tumors were round or oval/irregular. There were 22 cases of cystic degeneration, 10 cases of necrosis, and 9 cases of calcification (►**Fig. 1**). Forty-one tumors were located near the spine, psoas, adrenal region, or kidneys in the retroperitoneal region of the abdomen, and 26 tumors were located in the pelvis. Sixty-four 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 eroded the lumbar spine or sacrococcygeal bone in three cases (►**Figs. 3**, **4**), encapsulated the iliac arteriovenous vessels in 3 cases, and wrapped around the celiac trunk vessels in 2 cases (►**Figs. 5** and **6**). Four cases underwent vascular interventional examination, and two of them performed interventional embolization before surgery to prevent major bleeding.

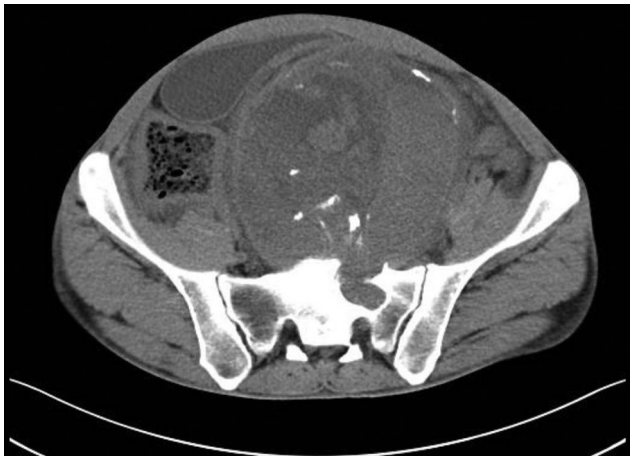

Fig. 1 A pelvic schwannoma containing necrosis and calcifications.

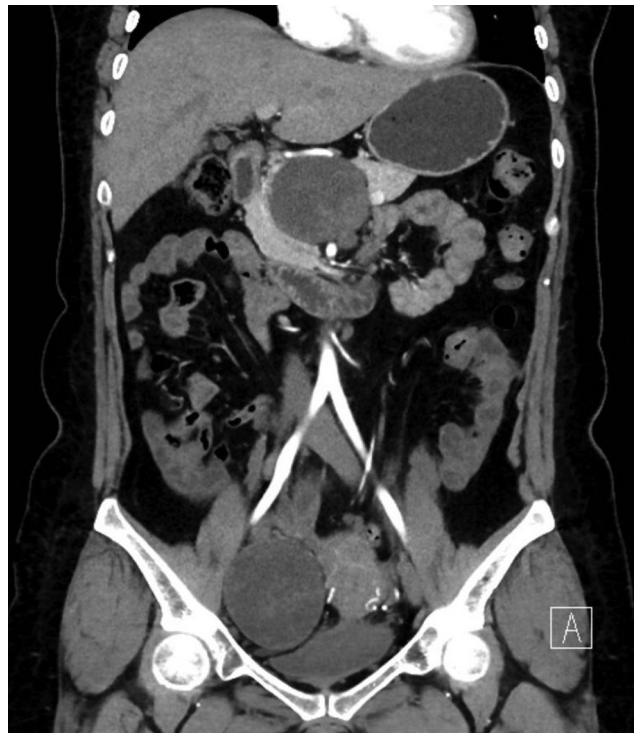

Fig. 2 A case of multiple schwannomas located posterior to the pancreas and to the right pelvis.

Surgical Results

All the patients underwent surgery. There were no surgical deaths. There were 59 cases of open surgery and 8 cases of laparoscopic surgery. Sixty-two patients underwent complete resection and 5 patients underwent subtotal resection. Subtotal resection was performed in three cases because the tumor was wrapped around the celiac trunk or iliac vessels, and in two cases because it extended into the intervertebral foramen or resulted in irregular bone erosion. The median intraoperative blood loss was 300 ml (20–7,600 ml). There were six cases of preoperative ureter compression with hydronephrosis and five cases of ileus. In 19 patients, an ureteral stent was placed preoperatively, 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 vertebrae resection, 2 cases of partial sacral coccyx resection, and 1

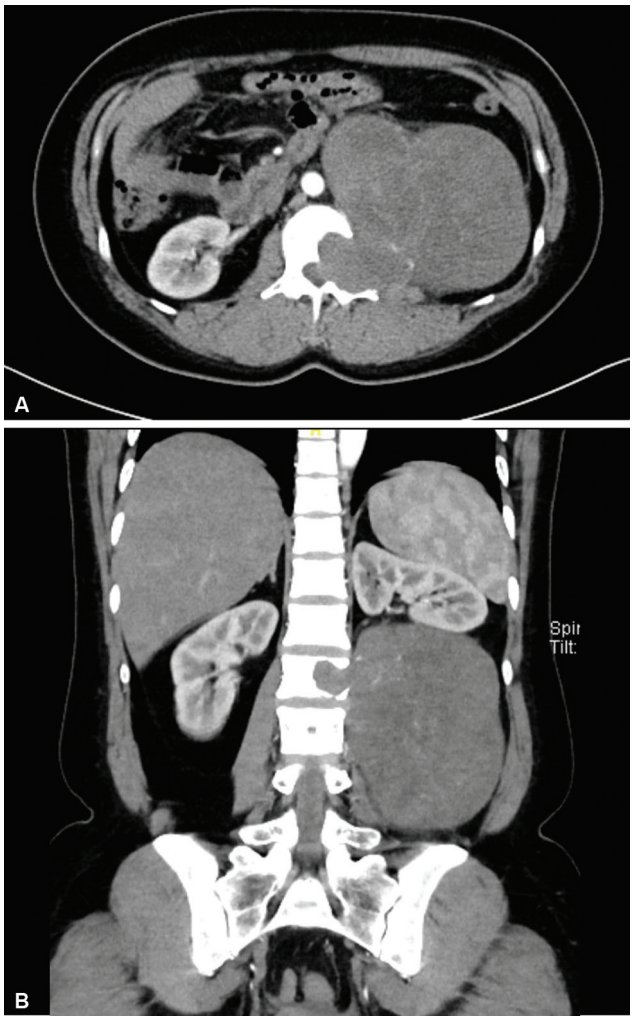

Fig. 3 A left-sided retroperitoneal schwannoma eroded the lumbar vertebral body.

case of partial pancreatic resection. Postoperative complications occurred in six patients, including two cases of ileus, two cases of paralysis and discomfort in the lower extremities, one case of urinary fistula, and one case of postoperative bleeding. Due to lack of maneuvering space or proximity to important blood vessels, nine cases of pelvic retroperitoneal tumors and three cases of abdominal retroperitoneal tumors underwent block resection. Overall, 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 the maximum size of the tumor, blood loss, proportion of block resection, and postoperative hospital stay ($p < 0.05$).

Postoperative Pathology

All tumors were benign. The range of tumor diameter was 2.5 to 26 cm, with the average being 9.56 ± 5.27 cm. Hematoxylin and eosin (H&E) staining revealed a large number of spindle cells arranged in a cross-striped or beamlike arrangement, manifested as Antoni A region rich in Schwann cells

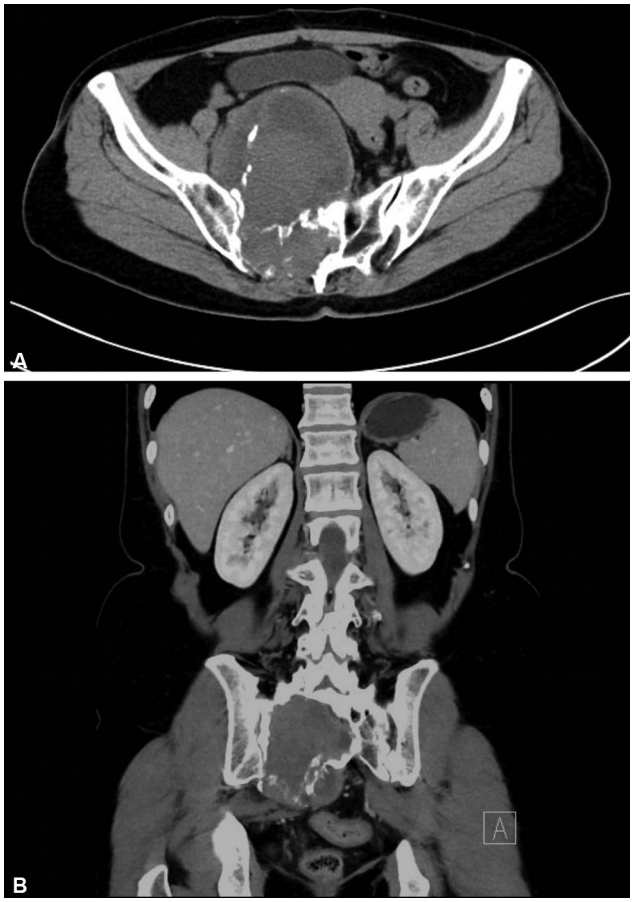

Fig. 4 A pelvic schwannoma eroded the sacrococcygeal bone.

and Antoni B region with low cell content, and positive immunohistochemical *S100* staining.

Follow-up

Sixty-two patients (92.5%) were followed up. No tumor recurrence was found within 3 to 68 months, and the survival rate at 1, 3, and 5 years was 100%. Five patients underwent subtotal resection. Their follow-up time was 7 to 43 months. The residual tumors did not have obvious progression.

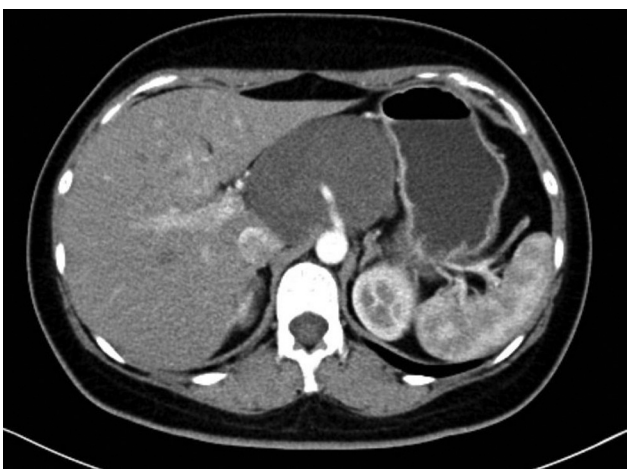

Fig. 5 A retroperitoneal schwannoma wrapped around the celiac trunk.

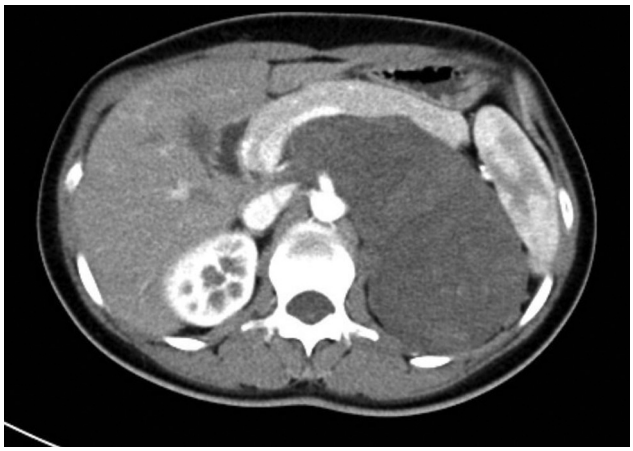

Fig. 6 A retroperitoneal schwannoma wrapped around the celiac trunk and pushed against the left kidney.

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, and 69% of the patients in this group were females.[2,6] Retroperitoneal schwannomas are rare, and originate mainly from the 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 the present study were up to 26 cm in diameter, and in 27 cases the tumors had 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, causing 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 placed an ureteral stent cystoscopically 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 preoperative endovascular embolization to reduce intraoperative bleeding.

Retroperitoneal tumors should be discussed preoperatively by a MDT, which can improve diagnostic accuracy based on imaging and clinical features. 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 schwannomas from other retroperitoneal tumors. Schwannomas are also easily misdiagnosed as malignant, leading to overtreatment with unnecessarily extensive removal. In all, 10.4% of the patients in this group were mistaken for malignancy before surgery, due to the large tumor range, erosion of bone, and the tumor wrapping around the blood vessels. In the study population, the tumor eroded the lumbar spine in two cases (►**Fig. 3**), and the sacral coccyx in 1 patient (►**Fig. 4**). The tumors also can grow to the spinal or sacrococcygeal foramamina. In the study population, there also had been tumors that had wrapped around the celiac trunk or the iliac arteriovenous blood vessels (►**Figs. 5**, **6**), which may easily lead to intraoperative bleeding and residual tumor. The surgical strategy is also an important part of the MDT discussion. When operating on the retroperitoneal nerve sheath or nerve-associated tumors, monoportal approaches may have limitations in exposure of the tumor and surrounding structures. Heinen et al[12] proposed an algorithm for a modular strategy based on tumor location, size, spread, and assumed 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] Paragangliomas are mainly located in the sympathetic chain on both sides of the spine, and may be accompanied by cystic degeneration, calcification, and necrosis; paroxysmal hypertension can be occasionally found.[14] The contrast enhancement is more obvious than that of schwannoma.[15] The ganglioneuroma has a clear boundary, and the lesions may show pseudopodialike or embedded growth. The density is mostly uniform. It may be accompanied by calcification or fatty degeneration: cysts are less frequently seen. Contrast enhancement is not obvious.[16]

Table 1 Clinical data comparison of the pelvic group with the 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)	1,000 (100–9,000)	150 (20–6,500)	$z = 3.06$	0.002
Complications, n (%)	4 (15.4)	2 (4.9)	$\chi^2 = 2.154$	0.142
Rn block resection, n (%)	9 (34.6)	3 (7.3)	$\chi^2 = 8.064$	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)	$\chi^2 = 0.729$	0.393

The indication for biopsy of a retroperitoneal tumor is discussed controversially because in most cases the radiologic appearances are characteristic.[6] Furthermore, it is difficult to obtain the accurate diagnosis by needle biopsy. In this group, only nine patients underwent preoperative biopsy. We do not think that biopsy is worth recommending.

Postoperative complication rates of retroperitoneal schwannomas are low. Prognosis is good. Malignant transformation of schwannomas and local recurrence after resection are extremely rare.[6] Only a very small number of epithelial schwannomas show 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 with increased risk of intraoperative bleeding. due to the small space for surgical maneuvers and the close relationship between the tumor, the sacrococcygeal plexus and the iliac vessels. In patients with invasion of the lumbar spine and sacrococcygeal bone, we worked with orthopedic surgeons at the same time, followed by retroperitoneal tumor surgery to achieve complete resection. Incomplete tumor resection often is due to blood vessels enclosed by the tumor, and tumor growth into the intervertebral space. All patients with a small residual tumor should be closely followed up. In our series, no obvious malignant changes were found during follow-up consistent with other reports.[6,18] In the recent years, schwannomas have also been removed by laparoscopy or robotic laparoscopy.[19] The tumor volume of our patients undergoing laparoscopic surgery was less than 5 cm.

In summary, accurate preoperative diagnosis is helpful to guide the surgical treatment of schwannomas. Complete resection is associated with a good long-term prognosis and should be performed early after detection. Subtotal resection, which minimizes surgical risk and preserves surrounding vital structures, can also achieve an acceptable prognosis, but requires close follow-up.

Conflict of Interest
None declared.

References

1 Vijayan SKL. Retroperitoneal schwannoma: an atypical presentation. J Clin Diagn Res 2014;8(10):ND22–ND23
2 Li Q, Gao C, Juzi JT, Hao X. Analysis of 82 cases of retroperitoneal schwannoma. ANZ J Surg 2007;77(04):237–240
3 Improta L, Tzanis D, Bouhadiba T, Abdelhafidh K, Bonvalot S. Overview of primary adult retroperitoneal tumours. Eur J Surg Oncol 2020;46(09):1573–1579
4 Harhar M, Ramdani A, Bouhout T, Serji B, El Harroudi T. Retroperitoneal schwannoma: two rare case reports. Cureus 2021;13 (02):e13456
5 Tortorelli AP, Rosa F, Papa V, et al. Retroperitoneal schwannomas: diagnostic and therapeutic implications. Tumori 2007;93(03): 312–315
6 Strauss DC, Qureshi YA, Hayes AJ, Thomas JM. Management of benign retroperitoneal schwannomas: a single-center experience. Am J Surg 2011;202(02):194–198
7 Transatlantic Australasian Retroperitoneal Sarcoma Working Group. Intercontinental collaborative experience with abdominal, retroperitoneal and pelvic schwannomas. Br J Surg 2020;107 (04):452–463
8 Ogose A, Kawashima H, Hatano H, et al. The natural history of incidental retroperitoneal schwannomas. PLoS One 2019;14(04): e0215336
9 Theodosopoulos T, Stafyla VK, Tsiantoula P, et al. Special problems encountering surgical management of large retroperitoneal schwannomas. World J Surg Oncol 2008;6(107):107
10 Hughes MJ, Thomas JM, Fisher C, Moskovic EC. Imaging features of retroperitoneal and pelvic schwannomas. Clin Radiol 2005;60 (08):886–893
11 Mastoraki A, Toska F, Tsiverdis I, et al. Retroperitoneal schwannomas: dilemmas in diagnostic approach and therapeutic management. J Gastrointest Cancer 2013;44(04):371–374
12 Heinen C, Schmidt T, Kretschmer T. Decision making in retroperitoneal nerve sheath and nerve-associated tumors: a modular approach. Neurosurgery 2020;87(03):E359–E369
13 Ahlawat S, Blakeley JO, Langmead S, Belzberg AJ, Fayad LM. Current status and recommendations for imaging in neurofibromatosis type 1, neurofibromatosis type 2, and schwannomatosis. Skeletal Radiol 2020;49(02):199–219
14 Jain A, Baracco R, Kapur G. Pheochromocytoma and paraganglioma-an update on diagnosis, evaluation, and management. Pediatr Nephrol 2020;35(04):581–594
15 Shen Y, Zhong Y, Wang H, et al. MR imaging features of benign retroperitoneal paragangliomas and schwannomas. BMC Neurol 2018;18(01):1
16 Xiao J, Zhao Z, Li B, Zhang T. Primary retroperitoneal ganglioneuroma: a retrospective cohort study of 32 patients. Front Surg 2021; 8:642451
17 McMenamin ME, Fletcher CDM. Expanding the spectrum of malignant change in schwannomas: epithelioid malignant change, epithelioid malignant peripheral nerve sheath tumor, and epithelioid angiosarcoma: a study of 17 cases. Am J Surg Pathol 2001;25(01):13–25
18 Song JY, Kim SY, Park EG, et al. Schwannoma in the retroperitoneum. J Obstet Gynaecol Res 2007;33(03):371–375
19 Ohsawa M, Miguchi M, Yoshimitsu M, et al. Laparoscopic excision of a retroperitoneal schwannoma: a case report. Asian J Endosc Surg 2019;12(02):192–196