

Rare retroperitoneal giant sacral schwannoma: A case report

SHUNCHANG ZHOU^{1*}, SHENYI WAN^{1*}, LIANG LI², WEI DONG¹,
XUHUI MA¹, HAIBO CHU¹ and YUXU ZHONG³

Departments of ¹General Surgery and ²Radiology, Jiaozhou Branch of Shanghai East Hospital, Tongji University, Qingdao, Shandong 266300; ³State Key Laboratory of Toxicology and Medical Countermeasures, Beijing Institute of Pharmacology and Toxicology, Beijing 100850, P.R. China

Received November 10, 2023; Accepted February 15, 2024

DOI: 10.3892/ol.2024.14394

Abstract. Schwannomas localized in the sacrum are relatively infrequent, accounting for 1-5% of all spinal axis schwannomas; they present with vague symptoms or are symptomless, so often grow to a considerable size before detection. Sacral schwannomas occasionally present with enormous dimensions, and these tumors are termed giant sacral schwannomas. However, their surgical removal is challenging owing to an abundant vascularity. The present study retrospectively analyzed the clinical and follow-up data of a patient with a giant sacral schwannoma. The patient experienced numbness in the left buttock and lower extremity, with radiating pain in the sole of the foot that had persisted for 3 years. A presacral mass was found by computed tomography examination 6 months after the stool had become thin. A tumor resection was performed using the anterior abdominal approach. A schwannoma was diagnosed by postoperative pathology. The postoperative course was uneventful, with the complete resolution of symptoms during the 21-month clinical follow-up. Overall, the present study reports the case of a giant sacral schwannoma with pelvic pain that was resected without complications and also discusses its successful management. Additionally, the study presents a systematic review of the literature. We consider that the surgical treatment of giant sacral schwannomas with piecemeal subtotal excision can achieve good outcomes, avoiding unnecessary neurological deficits.

Introduction

Sacral schwannomas are infrequent tumors accounting for 1-5% of all spinal schwannomas (1-3). The tumors occur most commonly at 50-60 years of age (median, 56 years) (4). In 1932, Masson revealed that Schwann cells were derived from the nerve sheaths of myelinated nerves (5). Schwannomas can occur in any Schwann cell-containing body part, such as the head, neck and extremities, but retroperitoneal schwannomas are rare. Retroperitoneal schwannomas are solid or cystic benign tumors encapsulated within a membrane; they are often located in the paraspinal and presacral areas. Latent symptomatic retroperitoneal schwannomas account for 47.7-66.6% of all schwannomas (6). In addition, 5-8% of schwannomas are accompanied by neurofibromatosis (7). Sacral schwannomas have a relatively slow growth rate, along with a large retroperitoneal space, excellent ductility and lack of pain transmission. Consequently, when detected, sacral schwannomas have a large volume and present with numerous degenerative pathological changes, such as cystic degeneration, calcification and hemorrhage, with cystic degeneration and calcification occurring in 60 and 23% of individuals, respectively (8). Computed tomography (CT) or magnetic resonance imaging (MRI) results exhibiting these features allow clinicians to consider the possibility of sacral schwannomas.

The incidence of malignant schwannomas, in which the nerve sheath membrane tends to undergo malignant differentiation, is 0.001% and accounts for 3-10% of soft-tissue sarcomas (9). Malignant schwannomas are considered high-grade sarcomas that are highly malignant, strongly invasive, and prone to recurrence and metastasis, as well as having a low long-term survival rate; therefore, an early diagnosis is particularly critical. According to the literature, 15-70% of patients with malignant schwannomas have combined neurofibromatosis type 1, which may be associated with the malignancy development of peripheral neurofibromas (10,11). The World Health Organization (WHO) has used the terms 'malignant Schwann cell tumor', 'neurofibrosarcoma' and 'malignant schwannoma' to indicate the origin and malignant behavior of schwannomas. In 2000, schwannomas were formally defined by the WHO as any malignant tumor originating in the peripheral nerves or showing nerve sheath differentiation, except for tumors originating in the nerve periphery or peripheral neurovascular system. Malignant

Correspondence to: Professor Haibo Chu, Department of General Surgery, Jiaozhou Branch of Shanghai East Hospital, Tongji University, 89 Zhanqian Road, Jiaozhou, Qingdao, Shandong 266300, P.R. China
E-mail: haibochuwf@163.com

Professor Yuxu Zhong, State Key Laboratory of Toxicology and Medical Countermeasures, Beijing Institute of Pharmacology and Toxicology, 27 Taiping Road, Beijing 100850, P.R. China
E-mail: yuxuzhong2008@aliyun.com

*Contributed equally

Key words: sacral schwannoma, pre-sacral, operation

schwannomas, including two specific subtypes, epithelioid malignant schwannomas and malignant salamander tumors, were classified as soft-tissue tumors in 2013 (12).

As sacral schwannomas usually exhibit slow growth and unspecific symptoms, diagnosis and treatment are often delayed until the tumor enlarges (1). Most patients do not show specific clinical manifestations and characteristic imaging features; thus, the preoperative diagnosis of sacral schwannomas is usually difficult (13). Benign schwannomas are differentiated from malignant ones based on the duration of the disease, clinical neuroerosive corrosive symptoms, capsule integrity on CT or MRI, and pathological examination results. The present study describes the surgical management of a giant sacral schwannoma and reviews the relevant literature, particularly associated case reports.

Case report

A 54-year-old man complaining of paraesthesia and radiating pain in the lower left extremity visited Jiaozhou Branch of Shanghai East Hospital, Tongji University (Qingdao, China) in February 2022. Radiating pain in the sole of the foot had increased in intensity gradually during the last 3 years, and the stool had become thin in size over the last 6 months. The patient also had a history of mild back pain and sciatica over the last 3 years and had been diagnosed previously as having L_{3,4}, L_{4,5}, L₅ and S₁ lumbar disc herniation on the basis of CT images. Left foot CT and MRI revealed osteochondral injury with degenerative cysts in the inner side of the trochlea in the left talus and a small effusion in the left ankle. The patient had undergone some massage therapy and physiotherapy in a number of the larger hospitals in Qingdao. The diagnosis was delayed, as an abdominal CT had not been performed. A history of trauma was not observed, and the patient was otherwise healthy. The physical examination revealed mild limitation in the movement of the left lower extremity, atrophy in the left buttock, thigh and calf muscles, and skin hypoesthesia. The circumference of the left lower extremity was 34 cm at 10 cm above the knee, 30 cm at 10 cm below the knee and 19.4 cm at 5 cm above the ankle. The circumference of the right lower extremity was 35.5 cm at 10 cm above the knee, 31.8 cm at 10 cm below the knee and 20.3 cm at 5 cm above the ankle. Contrast-enhanced CT of the pelvis demonstrated a cystic-solid mass on the left side of the pelvis. Three-dimensional CT reconstruction technology revealed that the feeding artery that originated from the internal iliac artery flowed into the tumor. There were more branches of the left internal iliac artery than usual, and tortuous branches were observed along the tumor margin (Fig. 1A-C). The diagnosis at the time of hospital admission was of a pelvic tumor with nerve compression syndrome in the left lower extremity. After the patient underwent blood routine, coagulation function, liver and kidney function, blood glucose, blood fat, electrolyte, carcinoembryonic antigen, α -fetoprotein, cancer antigen (CA)19-9, CA125 and CA153 tests, abdominal ultrasound and lower extremity electromyogram examinations, and was administered preoperative multi-disciplinary treatment, the pelvic tumor was resected using the transabdominal anterior approach in March 2022. After opening the abdomen, a large encapsulated retroperitoneal mass of ~5 cm in diameter was detected by the anterior approach in the left side of

the pelvis region, displacing the urinary bladder upwards and rectosigmoid colon to the right side. The anterior approach has the advantage of not causing spinal instability. The lesion was immobile, without evidence of local invasion. The excision of the mass was challenging. Due to its size, the posterior peritoneum was opened over the mass. The sigmoid colon was mobilized from the presacral space to expose the tumor mass. The ureters were identified bilaterally and protected. A plane was then established between the tumor mass and the presacral alar tissue. As the tumor's blood supply originated from the left internal iliac artery, and thick, tortuous veins were observed on the tumor surface, the left internal iliac artery was first ligated and severed. The tumor was firmly attached by fibrous tissue to the presacral fascia and left iliac vein. A tissue block was collected from the tumor surface and subjected to rapid frozen pathology. The pathology revealed a benign schwannoma. Using an ultrasonic knife, in circumferential fashion, the tumor was completely mobilized from the sigmoid colon and upper rectum, the left iliac vein and the other surrounding anatomical structures with great care for exact hemostasis. The tumor envelope was then incised along the longitudinal axis of the nerve, and using sharp dissection. Finally, the remaining attachment of the tumor tissue to the presacral nerve root(s) was identified and carefully dissected free with ligasure device. The tumor was then completely resected by an intralesional resection (the indications of intralesional resection included the large size of the tumor and the clinical manifestations of nerve bundle erosion). For giant schwannomas, we believe it is not possible to lift the peritoneal sac as the tumor mass is so large. In addition, this mass had to be broken up to safely dissect the iliac vessels and ureters. Moreover, it was a multidisciplinary event, especially for the digestive surgeon, who had a perfect knowledge about this type of surgical approach while being an expert in laparoscopic surgery. The digestive surgeon performed the first operating phase with the surgical approach and the identification of vessels and ureters, and the neurosurgeon, by virtue of having knowledge of the lumbosacral plexus, helped with the identification of the sacral roots and the excision of the lesion. This allowed the association of two complementary specialists and reduced both operating time and surgical risks.

The central region of the tumor was cystic in appearance and the tumor measured 5x4x4 cm³ (Fig. 2A and B). A schwannoma was diagnosed on the basis of the postoperative pathology. Specimens were fixed in 10% neutral formalin solution at room temperature for 24 h, dehydrated, embedded and serially sliced into 4- μ m sections. One slice of the sample was used for H&E staining. The sections were baked at 60°C for 30 min, and the sections were viewed under an optical microscope. For the immunohistochemical analysis, specimens were fixed in 10% neutral formalin solution at room temperature for 24 h, dehydrated, embedded and serially sliced into 4- μ m sections. Antigen retrieval was performed using endogenous peroxidase blocker at room temperature for 10 min, and the non-specific antigens were blocked. The immunohistochemical SP method was performed. Sections were then incubated overnight at 4°C with primary monoclonal antibodies (ready-to-use; rabbit anti-human S-100, cat. no. ZM-0224; Vimentin, cat. no. ZM-0260; and Ki-67, cat. no. ZM-0224; OriGene Technologies, Inc.), followed by incubation at room temperature for 30 min with enzyme-labeled

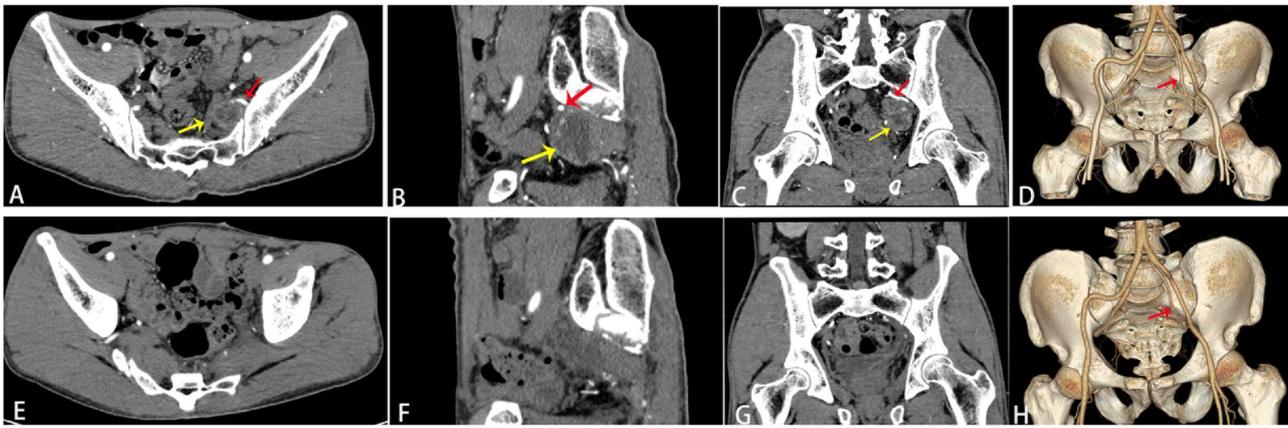


Figure 1. Images of sacral schwannoma. (A) Horizontal view of enhanced CT on the left side of the pelvic region showing a round, 5.1x4.3-cm cystic-solid mass. (B) Sagittal view of enhanced CT on the left side of the pelvis region showing the tumor partially adjacent to the sacrum and had abundant blood vessels. (C) Coronal enhanced CT on the left side of the pelvic region showing delayed slight enhancement of the tumor capsule wall. (D) Three-dimensional CT reconstruction showing that the feeding artery originating from the internal iliac artery cranially flows into the tumor, the left internal iliac artery branches are increased in number, and tortuous branches are present along the tumor margin. (E-G) Postoperative enhanced CT showing that the tumor was completely resected, and the blood vessels were significantly reduced in the surgical area. (E) Horizontal, (F) sagittal and (G) coronal enhanced CT views, and (H) three-dimensional CT reconstruction showing the severed left internal iliac artery and the remaining stump of the artery. The red arrow indicates the internal iliac artery and the yellow arrow indicates the tumor. CT, computed tomography.

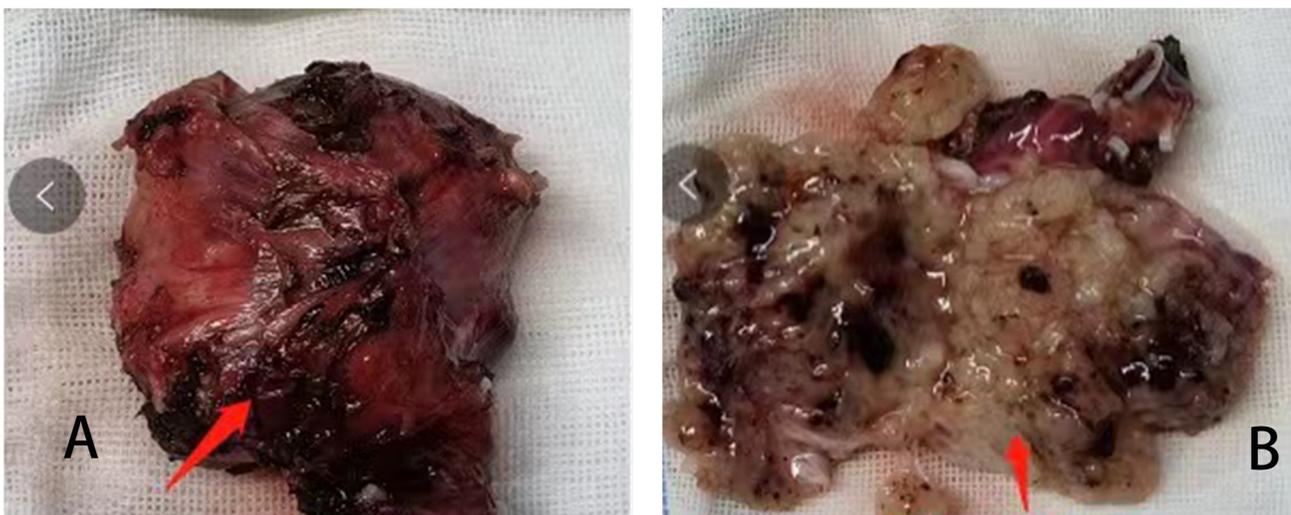


Figure 2. Resected sacral schwannoma. (A) The tumor was quasi-circular and cyst-solid, with thick veins on the surface. (B) The dissected tumor showed cystic changes and intracapsular bleeding.

sheep anti-mouse/rabbit IgG polymer secondary antibody (ready-to-use; cat. nos. PV-6000D and IB000084; OriGene Technologies, Inc.) with diaminobenzidine (DAB) substrate buffer solution and DAB concentrated colour reagent, and counterstained with hematoxylin at room temperature for 5 min. The negative control group was studied using the same steps, but phosphate-buffered saline was used instead of primary antibody. The sections were viewed under a BX51 optical microscope (Olympus Corporation) at x200 and x400 magnification.

Immunohistochemical observations were as follows: S-100-positive, vimentin-positive and Ki-67-positive diffuse tumor cells were observed, and the Ki-67 index of the original tumor was 1% (Fig. 3A-D). Postoperative three-dimensional CT reconstruction technology revealed that the tumor was completely resected, with a significant reduction in blood

vessels in the surgical area. The left internal iliac artery was severed and only a stump of the artery remained (Fig. 1D-F). Follow-up of the patient was performed every 6 months, including EMG of the left lower extremity, enhanced CT and MRI. No recurrence was observed during the 21-month follow-up, and the patient did not receive any further treatment.

Discussion

Although the pathogenesis of schwannomas is unclear, their occurrence may be related to relevant gene mutations. Peripheral nerve schwannomas are well demarcated, encapsulated tumors, typically round and attached to the nerve; they arise from the Schwann cells and are eccentric in location, involving one or two fascicles, and sparing and displacing the other neural fascicles of the involved nerve. The tumors occur

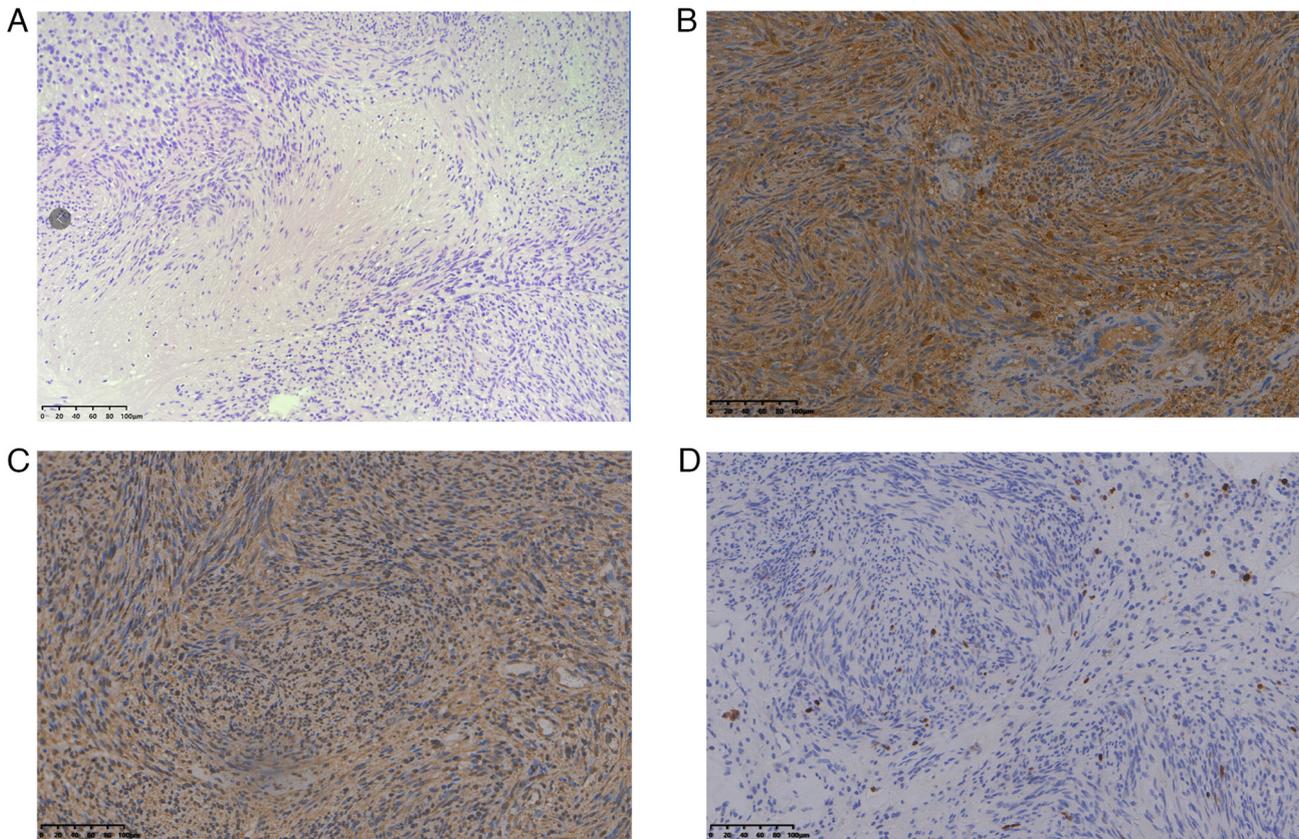


Figure 3. Pathology of sacral schwannoma. (A) Microscopically, a large number of densely spindle cells can be seen in the Antoni A area and sparse tumor cells rich in mucous matrix in the Antoni B area (hematoxylin and eosin; magnification, x200). (B) S-100-positive expression (magnification, x200). (C) Vimentin-positive expression (magnification, x200). (D) Ki-67-positive expression (magnification, x200).

within the endoneurium and are surrounded by perineurium and fibrous epineurium that encapsulate the tumor (14). In retroperitoneal schwannomas, the mean tumor growth rate is 1.9 mm/year. The majority of peripheral tumors are solitary (96%), while only 4% are plexiform (15). According to their anatomical locations, schwannomas are divided into intraosseous, dumbbell and retroperitoneal types. The intraosseous type refers to tumors located in or invading the spinal canal. Dumbbell-type schwannomas are tumors extending outside the spinal canal through the intervertebral foramen. Retroperitoneal-type schwannoma refers to a tumor located in the posterior peritoneum outside the spinal canal (16). The tumor size is generally based on length, but the size of the dumbbell type is based on the largest part of the tumor. Sacral schwannomas have a slow growth rate. Moreover, schwannomas display strong concealment abilities due to the large pelvic space. Various clinical symptoms, including pelvic organ or nerve tissue compression, bone destruction, sciatica, lower back pain, bladder and rectum compression due to difficulty in urination and defecation, and weakness in lower limbs, are associated with sacral schwannomas (17-19). Feldenzer *et al* (20) found that 67% of patients with sacral schwannomas exhibited rectal irritation symptoms due to tumor compression. Therefore, a delayed diagnosis is a clinical feature of sacral schwannomas. In the present case, the patient had suffered from numbness in the left buttock and lower extremity, with persistent radiating pain in the foot sole for 3 years. The patient had visited several hospitals in Qingdao

previously, but only the presence of lumbar and foot lesions was determined by the treating physicians, who ignored the results of the abdominal CT examination. The tumor remained undetected until the patient developed rectal compression symptoms and underwent an abdominal CT, which is an informative lesson for clinicians. Clinically, the numbness of the skin of the lower limbs on the affected side and the presence of muscle atrophy suggested that the tumor had eroded nerve bundles. According to one previous study, the mean time for a sacral schwannoma diagnosis was 5.2 years, and the longest time was 11 years (21). In general, sacral malignant tumors are common in chordoma and chondrosarcoma, and are metastatic, whereas sacral benign tumors are common in giant osteocytoma, aneurysmal bone cysts and osteoblastoma (22). However, sacral schwannomas are rare and need to be differentiated from fibrosarcomas, liposarcomas, astrocytomas, hydatid cysts, hematomas and connective tissue diseases (23).

On enhanced CT scans, the cystic wall presents with delayed slight enhancement, and parenchymal components exhibit plaque-like progressive but irregular enhancement. MRI T2-weighted imaging (T2WI) sequencing displays the following pathological characteristics of the internal tumor: The Antoni A area with dense cells showing an equal or slightly high signal, and the Antoni B area rich in stroma and mucus showing a higher signal (24). Verstraete *et al* (13) and Cuesta *et al* (4) summarized the imaging data of schwannomas and found tumors with low-density changes on enhanced CT, whereas unique changes were present in the schwannomas on MRI (a low or equal

signal on T1WI and a high or heterogeneous signal on T2WI). Pan *et al* (25) reported the following diversified CT and MRI changes: Hemorrhage, degeneration, necrosis and liquefaction. On MRI, the low- and high-density signals on T1WI and T2WI, respectively, helped surgeons to provide a reference for a schwannoma diagnosis before the operation. Makni *et al* (26) suggested that the CT density helped to evaluate fluid and adipose tissues, determine how tumors and the sacrum or the coccyx and rectum are anatomically related, and determine the presence or absence of calcification (excluding chordoma). MRI can help estimate the tumor size, exact location and relationship of the tumor with adjacent nerves. Preoperative CT-based 3D reconstruction technology can more clearly reveal the relationship between tumors and adjacent blood vessels and nerves (27). Large volumes are characteristic of retroperitoneal schwannomas. In a previous study, CT revealed that degeneration and hemorrhage often occur in the tumor body, and calcification was observed (28). Additionally, a single benign schwannoma can be detected using fluorodeoxyglucose positron emission tomography/CT (29). The standard agreed diagnostic criterion for giant schwannomas is a tumor size of ≥ 5 cm on CT or MRI (1,30). In the present case, enhanced CT and three-dimensional reconstruction of the pelvis unveiled that the tumor was round, cystic-solid, with a thin cystic wall and smooth inner and outer walls. The tumor exhibited uniform enhancement, predominantly hypointense, with delayed and mild enhancement of the cystic wall. The tumor was partially adjacent to the sacrum, and the branches of the left internal iliac artery were increased in number and traveled along the tumor margin. The venous phase exhibited further mild progressive enhancement, with cloudy flocculent. The demarcation between high and low densities was more clearly defined. The aforementioned imaging changes were consistent with the imaging features of schwannomas. Enhanced CT and three-dimensional reconstruction of the pelvis revealed that the tumor was completely resected, and the blood vessels in the surgical area were reduced in number. The left internal iliac artery was severed and only a stump of the artery remained. With sacral schwannoma, accurate preoperative imaging evaluation is important for choosing a rational surgical approach (both to completely resect the tumor and to avoid nerve trunk damage). In the case, preoperative three-dimensional CT reconstruction revealed that the tumor originated from the left internal iliac artery, suggesting that surgeons should make clear the relationship between tumor and blood vessels, and choose a reasonable surgical approach, so as to avoid accidental massive haemorrhage during the operation. Moreover, a MRI view that shows the continuity to a nerve root may help the diagnosis. In the present case, MRI was not performed before the operation, which is a limitation of the study.

In the present study, based on the gross pathological examination, the schwannoma was white/yellow with a hard texture. Under the microscope, several densely arranged spindle cells were observed in the Antoni A area, and fewer tumor cells with the mucinous matrix were observed in the Antoni B area. Xu *et al* (23) and Yu *et al* (29) reported that schwannoma pathology is based on degenerative changes and nuclear atypia. Immunohistochemical analyses, such as analysis of levels of S-100, neuron-specific enolase or vimentin, are crucial for the pathological diagnosis. Ki-67 refers to the nuclear proliferation index. This index reflects cell proliferation and simultaneously

indicates the probability of tumor recurrence after operation. Ogose *et al* (30) reported that the Ki-67 index of presacral schwannomas was generally $<1\%$. In the present case, the gross specimen view revealed that the tumor was quasi-circular, cystic-solid, and thick veins were observed on the surface. On dissecting the tumor, cystic degeneration and intracapsular hemorrhagic changes were found. Under the microscope, numerous densely arranged spindle cells were noted in the Antoni A area, and sparse tumor cells rich in mucus stroma were observed in the Antoni B area. Positive expression of S-100, vimentin and Ki-67 was also observed. Schwannoma is a tumor originating from the mesenchymal tissues. The positive expression of S-100, a neuroendocrine marker, indicates that the tumor originates from neural tissues (31,32). Vimentin is a fibroblast component that belongs to the cytoskeleton; it is mainly distributed among cytoskeletal elements, as well as in tumors originating from mesenchymal cells, such as schwannomas or neurofibromas. Vimentin-positive expression also suggests a tumor originating from neural tissues (33). Ki-67 is a nuclear antigen that reflects cell proliferative activity (34,35). In the present case, the Ki-67 index of the original tumor was 1% , which indicated a benign tumor, and there was a low recurrence rate. Ozdemir *et al* (36) found that 50% of patients with sacral schwannomas exhibited nerve erosion, and emphasized that invasion and erosion are two different concepts. Invasion is tissue destruction by the tumor, whereas erosion is tissue replacement by the tumor. These concepts suggest that surgeons should consider both the thoroughness of tumor resection and the preservation of the nerve function when opting for surgical procedures. Considering the pathological characteristics of giant sacral schwannomas, surgeons must perform a reasonable operation rather than a perfect operation.

The envelopes of schwannomas are normal nerve sheath tissues that are not a part of the tumor; thus, the encapsulated tumor should be completely removed. A potential risk of nerve injury may arise when approaches such as extracapsular tumor resection are used to deliberately reduce the recurrence rate. Therefore, the complete resection of the enveloped tumor is sufficient for preventing its recurrence, protecting nerve functions, reducing nerve damage and reducing the probability of surgical complications. Studies have highlighted the use of 3D imaging technology-based preoperative needle biopsy for designing surgical protocols, as well as the need for multidisciplinary team collaboration and careful discussion, to avoid any possible complications (3,37). The use of a preoperative needle biopsy remains controversial. The advocates of this technique believe that a biopsy can help in making a clear diagnosis of the tumor, excluding lymphoma, and in the selection of reasonable surgical procedures. The opponents argue that a risk of infection and bleeding exists; nonetheless, most scholars have advocated for a preoperative hollow-needle biopsy (2,22,38). No consensus exists on the surgical management of sacral schwannomas. Surgical procedures may include a total or subtotal resection or a partial resection of the schwannoma. In the so-called subtotal resection, the tumor is resected to $\geq 90\%$, while $<90\%$ of the tumor is resected in a partial resection (1,18). Handa *et al* (16) reported that giant sacral schwannomas were surgically removed in 11 patients who were followed up for a mean time of 50 months, and that no recurrence was observed in 8 patients who underwent a

total or partial resection, whereas recurrence was observed in 2 out of the 3 patients who underwent a partial resection. Using curettage and radiotherapy approaches for giant sacral schwannomas in 4 patients maximally protect the sacral nerve bundle in a study by Chandhanayingyong *et al* (39); however, the recurrence rate was 54%. Some scholars have proposed that nerve monitoring should be performed during the removal of giant sacral schwannomas to achieve maximal tumor resection while avoiding nerve injury and reducing tumor recurrence (19,40). The diagnosis must be reliable before surgical treatment for giant sacral schwannoma, avoiding the intralesional resection of retroperitoneal malignancies (41). In this present case, the abdomen was explored using the anterior approach, and the posterior peritoneum was opened. The tumor's blood supply originated from the left internal iliac artery, and thick, tortuous veins were observed on the tumor surface. The left internal iliac artery was first ligated and severed, and the nerve trunk was completely exposed at the tumor's base. Using an ultrasonic knife, the tumor envelope was separated, and the nerve trunk at the tumor's base was completely exposed to determine the tumor-nerve association. A tissue block was collected from the tumor surface and subjected to rapid frozen pathology. The pathology revealed a benign schwannoma. The tumor envelope was then incised along the longitudinal axis of the nerve, and using sharp dissection, the tumor was completely resected (intralesional resection indications include the large size of the tumor and the clinical manifestations of nerve bundle erosion). While resecting the schwannoma, nerve bundle damage was avoided as much as possible. Most studies have recommended careful intracapsular excision of the tumor to minimize postoperative neural deficits, since most schwannomas are encapsulated. If nerve fibers surround the tumor surface, intracapsular enucleation can be carried out while preserving nerve fibers by making a small longitudinal incision in the capsule (14,23,42,43). A number of studies have used laparoscopy or robotic assistance to clearly observe and enlarge the characteristics of tissue structures in a narrow space, remove tumors completely and protect nerve structure integrity, in alignment with the concept of precision and minimally invasive surgeries (2,44).

Adjuvant therapies for sacral schwannomas are embolization, chemotherapy, hyperthermia, Gamma Knife[®] therapy and cryosurgery (30,45). Different postoperative complications and prognoses of sacral schwannomas have been reported. Based on a review of the literature, Paulo *et al* (21) reported that of the 68 patients with sacral schwannomas, 10% had movement disorders, 6% had incontinence, 18% developed recurrence and 9% underwent reoperation after the initial surgery. Muallem *et al* (46) reported that, in 27 patients in whom sacral schwannomas were resected, 59.3% experienced a complete recovery from symptoms, 33.3% displayed a partial recovery from symptoms and 11.1% showed no symptom recovery. Yu *et al* (29) reported 12 cases of sacral schwannoma, 6 of which exhibited Ki-67 levels of >2% and 4 of which displayed postoperative recurrence. Thus, the postoperative recurrence of schwannomas is associated with the surgical procedure and biological tumor characteristics. The recurrence of schwannoma is affected by tumor size and the degree of resection. Therefore, laparoscopic or robotic surgery is recommended for giant sacral schwannomas, as the tumor location is deep and microscopic surgery cannot be performed. The magnification and clarity of image technology can facilitate

minimally invasive and complete removal of the tumor, and prevent nerve damage while reducing the recurrence rate. In the present case, no recurrence was observed during the 21-month follow-up period and the patient was asymptomatic.

In conclusion, sacral schwannomas are rare, and their clinical diagnosis is frequently delayed. Furthermore, surgical procedures for removing sacral schwannomas are diverse. Problems such as postoperative nerve injuries and tumor recurrence may be a challenge for surgeons when choosing appropriate treatment strategies for sacral schwannomas. Therefore, future studies should concentrate on measures for avoiding nerve injuries and reducing the postoperative recurrence rate.

Acknowledgements

The authors would like to thank Professor Yang Qing (Department of Pathology, Jiaozhou Branch of Shanghai East Hospital, Tongji University, Qingdao, Shandong, China) for guidance and help with the pathology in this study.

Funding

This study was funded by the Military Logistic Project of China (grant no. AWS17J008).

Availability of data and materials

The data generated in the present study may be requested from the corresponding author.

Authors' contributions

SZ and SW conceived the report and participated in data acquisition and the drafting of the manuscript. HC and YZ participated in the conception and design of the report, and critically revised the manuscript. LL and WD collected imaging data and performed study revisions. XM was responsible for the pathological analysis. SZ and HC confirm the authenticity of all the raw data. All authors read and approved the final manuscript.

Ethics approval and consent to participate

The reporting of this study conforms to the CARE guidelines. This study was exempt from ethical approval by the ethics committee of the Jiaozhou Branch of Shanghai East Hospital, Tongji University.

Patient consent for publication

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

Competing interests

The authors declare that they have no competing interests.

Authors' information

ORCID: Professor Haibo Chu, <https://orcid.org/0000-0002-3568-6431>.

References

- Macciò A, Kotsonis P, Aste L, Voicu MA, Madeddu C, Conti C and Camparini S: An interdisciplinary approach for laparoscopic removal of a large retroperitoneal pelvic schwannoma attached to vital vessels: A case report. *Medicine (Baltimore)* 98: e181492020, 2019.
- Ohsawa M, Miguchi M, Yoshimitsu M, Oishi K, Kohashi T, Hihara J, Mukaida H, Kaneko M, Egi H, Ohdan H and Hirabayashi N: Laparoscopic excision of a retroperitoneal schwannoma: A case report. *Asian J Endosc Surg* 12: 192-196, 2019.
- Korduke O, Omar A, Viner W and Kododa K: Pre-sacral schwannoma. *ANZ J Surg* 90: 1805-1807, 2020.
- Cuesta JP, Rodríguez LC, Bastidas N and Hernández M: Sacral schwannoma a case report and review of the literature. *Neuroradiol J* 36: 371-374, 2023.
- Masson P: Experimental and spontaneous schwannomas (peripheral gliomas): IL spontaneous schwannomas. *Am J Pathol* 8: 389-416, 1932.
- Brian KPG, Tan YM, Chuang YFA, Pierce KHC, London LPJO and Wong WK: Retroperitoneal schwannoma. *Am J Surg* 192: 14-18, 2006.
- Liu QY, Lin XF, Zhang WD, Li HG and Gao M: Retroperitoneal schwannomas in the anterior pararenal space: Dynamic enhanced multi-slice CT and MR findings. *Abdom Imaging* 38: 201-210, 2013.
- Kudo T, Kawakami H, Kuwatani M, Ehira N, Yamato H, Eto K, Kubota K and Asaka M: Three cases of retroperitoneal schwannoma diagnosed by EUS-FNA. *World J Gastroenterol* 17: 3459-3464, 2011.
- Yun JS, Lee MH, Lee SM, Lee JS, Kim HJ, Lee SJ, Chung HW, Lee SH and Shin MJ: Peripheral nerve sheath tumor: differentiation of malignant from benign tumors with conventional and diffusion-weighted MRI. *Eur Radio* 31:1548-57,2020.
- Stramare R, Beltrame V, Gazzola M, Gerardi M, Scattolin G, Coran A, Faccineto A, Rastrelli M and Rossi CR: Imaging of soft-tissue tumors. *J Magn Reson Imaging* 37: 791-804, 2013.
- Baheti AD, O'Malley RB, Kim S, Keraliya AR, Tirumani SH, Ramaiya NH and Wang CL: Soft-tissue sarcomas: An update for radiologists based on the revised 2013 world health organization classification. *AJR* 206: 924-932, 2016.
- Doyle LA: Sarcoma classification: an update based on the 2013 world health organization classification of tumors of soft tissue and bone. *Cancer* 120: 1763-1774, 2014.
- Verstraete KL, Achten E, De Schepper A, Ramon F, Parizel P, Degryse H and Dierick AM: Nerve sheath tumors: Evaluation with CT and MR imaging. *J Belge Radiol* 75: 311-320, 1992.
- Suárez C, López F, Rodrigo JP, Mendenhall WM, de Bree R, Mäkitie AA, Vander Poorten V, Takes RP, Bondi S, Kowalski LP, *et al*: Benign peripheral non-cranial nerve sheath tumors of the neck. *Adv Ther* 39: 3449-3471, 2022.
- El Sayed L, Masmejean EH, Parfait B, Kalamarides M, Biau D and Peyre M: Natural history of peripheral nerve schwannomas. *Acta Neurochir (Wien)* 162: 1883-1889, 2020.
- Handa K, Ozawa H, Aizawa T, Hashimoto K, Kanno H, Tateda S and Itoi E: Surgical management of giant sacral schwannoma: A case series and literature review. *World Neurosurg* 129: e216-e223, 2019.
- Khan UA, Ismayl G and Malik I: Giant sacral schwannoma treated with a 360 approach: A rare case and systematic review of the literature. *World Neurosurg* 115: 65-72, 2018.
- Tahta A, Dinc C, Ozdenkaya Y and Cakir A: Giant sacral schwannoma causing bilateral hydronephrosis: Case report and review of the literature. *World Neurosurg* 142: 184-187, 2020.
- Ragurajakash K, Hanakita J, Takahashi T, Ueno M, Minami M, Tomita Y, Tsujimoto Y and Kanematsu R: Giant invasive sacral schwannoma with aortic bifurcation compression and hydronephrosis. *World Neurosurg* 135: 267-272, 2020.
- Feldenzer JA, McGauley JL and McGillicuddy JE: Sacral and presacral tumors: Problems in diagnosis and management. *Neurosurgery* 25: 884-891, 1989.
- Paulo D, Semonche A and Tyagi R: Surgical management of lumbosacral giant invasive spinal schwannoma: A case report and literature review. *World Neurosurg* 114: 13-21, 2018.
- Turner ML, Mulhern CB and Dalinka MK: Lesions of the sacrum differential diagnosis and radiological evaluation. *JAMA* 245: 275-277, 1981.
- Xu H, Sha N, Li HW, Bai M, Chen X, Hu HL and Wu CL: A giant pelvic malignant schwannoma: A case report and literature review. *Int J Clin Exp Pathol* 8: 15363-15368, 2015.
- Leclerc A, Lebreton G, Huet A, Alves A and Emery E: Management of giant presacral schwannoma. Clinical series and literature review. *Clin Neurol Neurosurg* 200: 106409, 2021.
- Pan W, Wang Z, Lin N, Huang X, Liu M, Yan X and Ye Z: Clinical features and surgical treatment of sacral schwannomas. *Oncotarget* 8: 38061-38068, 2017.
- Makni A, Fetirich F, Mbarek M and Ben Safta Z: Presacral schwannoma. *J Visc Surg* 149: 426-427, 2012.
- Kinoshita T, Naganuma H, Ishii K and Itoh H: CT features of retroperitoneal neurilemmoma. *Eur J Radiol* 27: 67-71, 2015.
- Bai X and Wang XM: Solitary benign schwannoma mimics residual malignancy on FDG PET/CT. *Clin Nucl Med* 43: 782-784, 2018.
- Yu NH, Lee SE, Jahng TA and Chung CK: Giant invasive spinal schwannoma: Its clinical features and surgical management. *Neurosurgery* 71: 58-66, 2012.
- Ogose A, Hotta T, Hatano H, Kawashima H, Umezumi H, Higuchi T and Endo N: Presacral multiple cellular schwannomas. *Spine (Phila Pa 1976)* 28: E426-E429, 2003.
- Mokhtari M, Iranpour P, Golbahar Haghighi A and Ghahramani L: Schwannoma of the rectosigmoid colon. *Adv Biomed Res* 11: 5, 2022.
- Doan L and Cassarino DS: A rare case of cutaneous pseudoglandular schwannoma. *J Cutan Pathol* 50: 798-800, 2023.
- Bohlok A, El Khoury M, Bormans A, Galdon MG, Vouche M, El Nakadi I, Donckier V and Liberale G: Schwannoma of the colon and rectum: A systematic literature review. *World J Surg Oncol* 16: 125, 2018.
- Prueter J, Norvell D and Backous D: Ki-67 index as a predictor of vestibular schwannoma regrowth or recurrence. *J Laryngol Otol* 133: 205-207, 2019.
- Singh A, Aggarwal M, Chadalavada P, Siddiqui MT, Garg R, Lai K and Chahal P: Natural history of gastrointestinal schwannomas. *Endosc Int Open* 10: E801-E808, 2022.
- Ozdemir N, Bezircioğlu H and Akar O: Giant erosive spinal schwannomas: Surgical management. *Br J Neurosurg* 24: 526-531, 2010.
- Lin CL, Fang JJ and Lin RM: Resection of giant invasive sacral schwannoma using image-based customized osteotomy tools. *Eur Spine J* 25:4103-4107,2016.
- Çağlı S, Işık HS, Yıldırım U, Akıntürk N and Zileli M: Giant sacral schwannomas. *J Neurooncol* 110: 105-110, 2012.
- Chandhanayingyong C, Asavamongkolkul A, Lektrakul N and Muangsomboon S: The management of sacral schwannoma: Report of four cases and review of literature. *Sarcoma* 2008: 845132, 2008.
- Colonna MR, Costa AL, Mastrojeni C, Rizzo V, Nirta G, Angileri FF, Ieni A, Milone E and Macri A: Giant sacral schwannoma excised under intraoperative neuromonitoring in an elderly patient: case report. *J Surg Case Rep* 2021: rjab460, 2021.
- Berner EA, Hung YP, Nielsen GP and Lozano-Calderón SA: Malignant peripheral nerve sheath tumors arising from schwannomas: Case series and literature review. *APMIS* 129: 524-532, 2021.
- Ram S, Vivek V, Shekhar R, Gabbita AC and Ganesh K: Giant Cervicodorsal Schwannoma. *J Exp Ther Oncol* 13: 155-158, 2019.
- Ozturk C, Mirzanli C, Karatoprak O, Tezer M, Aydogan M and Hamzaoglu A: Giant sacral schwannoma: A case report and review of the literature. *Acta Orthop Belg* 75: 705-710, 2009.
- Yin J, Wu H, Tu J, Zou C, Huang G, Xie X, He Y and Shen J: Robot-assisted sacral tumor resection: A preliminary study. *BMC Musculoskelet Disord* 19: 186, 2018.
- Yang CC, Chen HC and Chen CM: Endoscopic resection of a presacral schwannoma: Case report. *J Neurosurg Spine* 7: 86-89, 2007.
- Mualem W, Ghaith AK, Rush D, Jarrar R, Alexander Y, Zamanian C, Atkinson JLD, Yaszemski MJ, Krauss WE, Spinner RJ and Bydon M: Surgical management of sacral schwannomas: A 21-year mayo clinic experience and comparative literature analysis. *J Neurooncol* 159: 1-14, 2022.

