# Two giant connected retroperitoneal schwannomas: A rare case report

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**Case report**

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# Abstract

**Background** There are few reports of large retroperitoneal schwannomas and few guidelines for the diagnosis and surgical treatment.

**Case presentation** A 40-year-old female came to our department due to abdominal pain for 1 day and there are no other specific symptoms. There were no abnormalities in the patient's tumor markers. Plain computer tomography (CT) scan of the whole abdomen revealed two mass soft tissue density shadows behind the peritoneum, with uneven internal density, cystic low-density shadows and patchy calcification shadows. The larger mass was about 12.0 cm × 12.3 cm in size. Then tumors were completely excised by a reasonable surgical approach while the surrounding organs closely related to the tumor were preserved. Postoperative pathology confirmed that the tumor was benign schwannoma. In the 18-month follow-up, the patient had no recurrences and was asymptomatic.

**Conclusion** We summarize the diagnosis and treatment of a rare giant retroperitoneal schwannoma. Laparotomy for the management of retroperitoneal giant schwannomas may be a safe and effective method.


# 1. Background

Schwannoma is a kind of tumor originating from Schwann cells of peripheral nerve. It is the most common peripheral nerve tumor and is usually benign [1, 2],and some of the tumors have neuroendocrine functions [3]. Schwannoma usually occurs in patients of 20–50 years old, with no significant gender difference. It is common in the head, neck and limbs [4], and can also occur in the larynx, liver, mediastinum, and retroperitoneal space [5]. Retroperitoneal schwannomas account for 0.3% ~ 3.0% of all schwannomas [2, 6], and schwannomas account for 1% of all retroperitoneal tumors [7]. Herein, we report a case of giant retroperitoneal schwannoma, which received satisfactory treatment results by complete resection of the tumor and preservation of surrounding organs.

# 2. Case Presentation

A 40-year-old female (body mass index: 21.9 kg/$m^2$) came to our department with abdominal pain for 1 day. The woman was found to have a mass on her left abdomen two months ago, and no systematic diagnosis and treatment was conducted. The patient came to see the doctor due to the abdominal pain, and there were no other specific symptoms. The patient had jaundice hepatitis and had been cured. She had no history of surgery or hypertension and had a normal menstrual history. Her father and uncle died of stomach cancer. On physical examination, the left abdominal swelling was observed, and the palpation showed that the mass was hard and fixed. Her laboratory tests were abnormal as follows: hemoglobin content was 80 g/L, and C-reactive protein concentration was 74.5 mg/L. There were no abnormalities in her tumor markers. Plain computer tomography (CT) scan of the whole abdomen revealed two mass soft tissue density shadows behind the peritoneum, with uneven internal density,

cystic low-density shadows, and patchy calcification shadows. The larger mass was about 8 cm × 8 cm in size, with CT value of 14–45 Hu, with clear boundaries, displacement of adjacent organs due to compression. No enlarged lymph nodes were observed in retroperitoneum. Further contrast-enhanced CT examination (Fig. 1A) after admission showed that the lesions had obvious space occupying effect and the upper part of the lesion was indistinct from the psoas major muscle. In arterial phase, multiple arterial vessels supplying blood from abdominal aorta can be seen, while in venous phase, there is no enhancement. Multiple cystic low-density shadows were seen in the lesion, and the enhancement degree was weaker than the lesion parenchyma. Three-dimensional reconstruction CT (Fig. 1B, C) of the tumor reveals displacement of the abdominal aorta and left kidney due to compression, with renal arteries and veins spanning the surface of the tumor. Combined with the imaging characteristics of enhanced CT, the possibility of sarcoma was considered. In consideration of the possibility of combined left nephrectomy, she underwent CT urography before surgery, suggesting that both kidneys had good blood perfusion, normal function and unobstructed excretion. After preoperative discussion, we decided to remove the tumor through the left paracolic sulcus into the retroperitoneal space.

Surgery was performed with the patient under general anesthesia in the supine position. The incision of the left rectus abdominis goes into the abdominal cavity layer by layer. To probe the abdominal cavity, no obvious adhesions or effusion were observed. The surface of the liver and spleen was smooth and no masses were seen. No masses were seen in the stomach, small intestine, omentum, ileocecum, colon and upper rectum. No abnormalities were found in bilateral appendages and uterus. The tumors were large and haa abundant blood vessels on its surface (Fig. 2A). The surface of tumors was covered by the omentum majus and left mesentery. It was located on the left side of the spine, behind the left renal arteries and veins, ureters, tail of the pancreas and spleen. The upper end reaches the left diaphragm, the lower end reaches the iliac fossa, the medial edge was attached to the left side of the abdominal aorta, and the lateral edge reached the left paracolic sulcus. Specific surgical procedures: First, the paracolic peritoneum was incised from the left paracolic sulcus and extended upward and downward to dissociate and released the left kidney, tail of pancreas and spleen. Take care to protect the blood vessels and ureters of the spleen and kidney. Then the loosened spleen and left kidney were pushed to the midline of the abdominal cavity to fully expose the left retroperitoneal space, and the tumors were confirmed to be two. Carefully dissociating along the capsule of the tumor and en bloc excision was performed. Ligation hemostasis were performed at the bleeding point of the surgical bed. Finally, confirming that there was no damage to the left colon, kidneys, tail of pancreas, and spleen. After rinsing the abdominal cavity with warm saline, a closed drain was placed in the surgical bed for 10 days. The surgery lasted 180 minutes without intraoperative complications. Intraoperative blood loss was about 200 ml. Examination of surgical specimens (Fig. 2B) showed 2 pale yellow enveloped tumors with smooth surface and hard texture. The larger one was 8.0 cm × 7.0 cm × 5.5 cm, and the smaller one was 4.0 cm × 5.0 cm × 6.0 cm, all of which underwent pathological examination. Histological results showed that the tumor parenchyma was composed of Schwann cells with light to moderate atypia in the nucleus (Fig. 3). Immunohistochemical results showed S-100 (+), SOX10 (+), Desmin (-), CD34 (vessel+), SMA (-), Bcl-2 (-), Ki67 (Positive rate 1%), ER (-), PR (-), NF (-), and CD99 (-) (Fig. 4). Histologic and immunohistochemistry

confirmed the tumor as a benign schwannoma. CT examination of the patient at 18 months after the operation showed no signs of tumor recurrence (Fig. 2C).

# 3. Discussions

Retroperitoneal schwannomas are generally solitary masses with well-defined and intact envelopments [1], most of which are located near the great vessels [2, 4]. The tumor may be solid or multilocular cystic with hemorrhage and necrosis. Under microscopy, the tumor is composed of two distinct forms of Schwann cells [8]. One is the cell-rich Antoni A region, in which the cells are fusiform or oval in shape and arranged into tactile corpuscles or eosinophil corpuscles. The other is the sparsely cellular Antoni B region, in which the cells are loosely arranged in the mucoid matrix and pathological variations are seen as epithelioid or calcified [5]. The typical immunohistochemical features of schwannomas are S100 protein and SOX10 positive but Desmin negative [2, 9–11]. In our case, the expression of S100 protein and SOX10 in the tumor cells was positive, while Desmin was negative, which was consistent with the immunohistochemical characteristics of schwannoma. The positive rate of Ki67 in tumor cells was only 1%, indicating a low degree of malignancy.

Small nonfunctional retroperitoneal schwannomas are often asymptomatic. With the increase of tumor size, patients may have abdominal pain, abdominal distension, constipation and other symptoms, but there is no specificity. At present, preoperative diagnosis of schwannoma mainly relies on CT and MR examination, but the imaging findings are also not specific. In our case, the preoperative CT findings of schwannoma indicated that the tumor boundaries were clear, the density inside the tumor was uneven, and cystic changes and calcification shadows were visible. However, there was no enlarged retroperitoneal lymph node. Contrast-enhanced CT showed heterogeneous enhancement within the tumor, and vascular shadows were seen at the edge of the lesion in the arterial phase. This is consistent with the histological structure and imaging features of schwannoma. MR examination can show the blood supply of the tumor more clearly which indicates its enhancement characteristics. Most of the schwannomas appear as low intensity on T1-weighted images and heterogeneous high intensity on T2-weighted images [5]. The combination of CT and MR examination can improve the diagnostic accuracy of retroperitoneal schwannomas. At present, PET/CT is gradually applied in the diagnosis of schwannoma. PET/CT may be useful in differentiating benign tumors from malignant ones [12]. Although Theodosopoulos *et. al.* [4, 13] believe that puncture biopsy is helpful for preoperative diagnosis, it may cause misdiagnosis due to the heterogeneity of the tumor. Furthermore, it may lead to the spread of tumor cells. Therefore, Maccio *et. al.* [2] do not recommend preoperative puncture biopsy. Sultan *et. al.* [3] suggested that nonfunctional and well-encapsulated heterogeneous retroperitoneal schwannomas could be biopsied.

As far as we know, since only a few cases of large retroperitoneal schwannomas have been reported in the literature, guidelines on the optimal surgical treatment are lacking. According to the research of Ogose *et. al.* [14], the average growth rate of the retroperitoneal schwannomas was 1.9 mm/year (1.9 to 8.7 mm/year). Most of the asymptomatic patients may be suitable for the management with the "wait and see" approach. But Kitagawa *et. al.* [15] argued that cancer growth patterns cannot be accurately

predicted based on patients' symptoms and imaging studies. Because of the obvious space occupying or compression effect of giant retroperitoneal schwannomas, no matter whether the giant retroperitoneal schwannomas are benign or malignant, active treatment should be performed. At present, the surgical procedure for retroperitoneal schwannomas is controversial. First of all, in terms of resection scope, local recurrence and malignant transformation of retroperitoneal schwannomas are extremely rare. As a result, it is suggested that patients with retroperitoneal schwannomas excluded from Von Recklinghausen disease could be treated with local resection, while preserving adjacent important organs [6, 16]. However, prior to the clear diagnosis of schwannoma, it is necessary to appropriately expand the resection boundary or even resect the adjacent organs. Even if the tumor is confirmed to be malignant by postoperative pathology, local recurrence rate can be significantly reduced by extending the resection range [13, 17]. As for the surgical methods, open operation is still the first choice for large retroperitoneal schwannomas. Theodosopoulos *et. al.* [13] reported 3 cases of open resection of retroperitoneal schwannoma, of which 2 cases were combined with partial resection of psoas muscle and 1 case with hysterectomy. During the 48-month follow-up, the patient had no tumor recurrence and was asymptomatic. There were also reports of laparoscopic or robot-assisted resection of retroperitoneal schwannomas [18–23]. However, there are about 10 cases of schwannomas near the great vessels have been reported [2], so laparoscopic surgery cannot be considered to be absolutely safe. In our opinion, we believe that retroperitoneal schwannomas are not suitable for laparoscopic surgery when the size of the tumor is large and the tumor is closely related to the surrounding large vessels or organs. Laparoscopic resection may be considered for tumors which are small, isolated, and away from larger vessels. However, the safety and long-term efficacy of laparoscopic surgery still need to be further evaluated. In this case, the patient's tumors were large in size, deep and fixed in location, and closely related to the abdominal aorta and renal arteries and veins. In order to facilitate the exploration and removal of the tumors, a surgical approach *via* the left paracolic sulcus was performed. Because the capsule of tumor is intact and there was no surrounding invasion, the resection margin was not enlarged. At the 18-month follow-up, the patient had no recurrence and was asymptomatic.

# 4. Conclusion

We summarize the diagnosis and treatment of a rare giant retroperitoneal schwannoma. Preoperative biopsy of the tumor should be considered carefully. The size and location of the tumor are important factors in determining the surgical method and approach. Laparotomy is feasible for larger retroperitoneal schwannomas and laparoscopic surgery may be considered for smaller tumors.

# Declarations


### Acknowledgements

Not applicable


### Authors’ contributions

Tongjun Liu, Jiannan Li and Tao Yang designed the report. Lanqing Cao, Wei Li, Kai Zhang and Yuecheng Zhu collected and assembled the patient data. Tao Yang and Jiannan Li wrote the paper. All authors read and approved the final manuscript.


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### Availability of data and materials

All data generated or analyzed during this study are included in this published article.

### Ethics approval and consent to participate

This study was approved by the Ethics Committee of The Second Hospital of Jilin University (reference number: 2020-159). Informed consent for the use of medical data was obtained from the patient.

### Consent for publication

Written informed consent for publication was obtained from the participant.

### Competing interests

The authors declare that they have no competing interests.

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# Figures

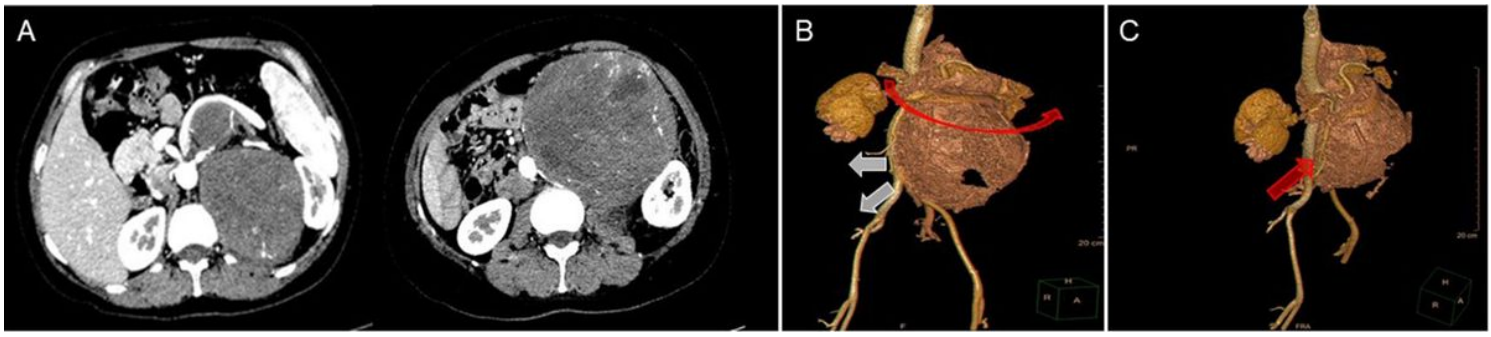



**Figure 1**

Contrast-enhanced CT and three-dimensional reconstruction of the tumor. (A) Contrast-enhanced CT showed that the tumor is located on the left side of the spine, behind the left renal arteries and veins, ureters and spleen. Three-dimensional reconstruction showed that the medial edge is attached to the left side of the abdominal aorta (B) and the tumor compresses the abdominal aorta to the right, and the left renal artery and vein span the tumor surface (C).

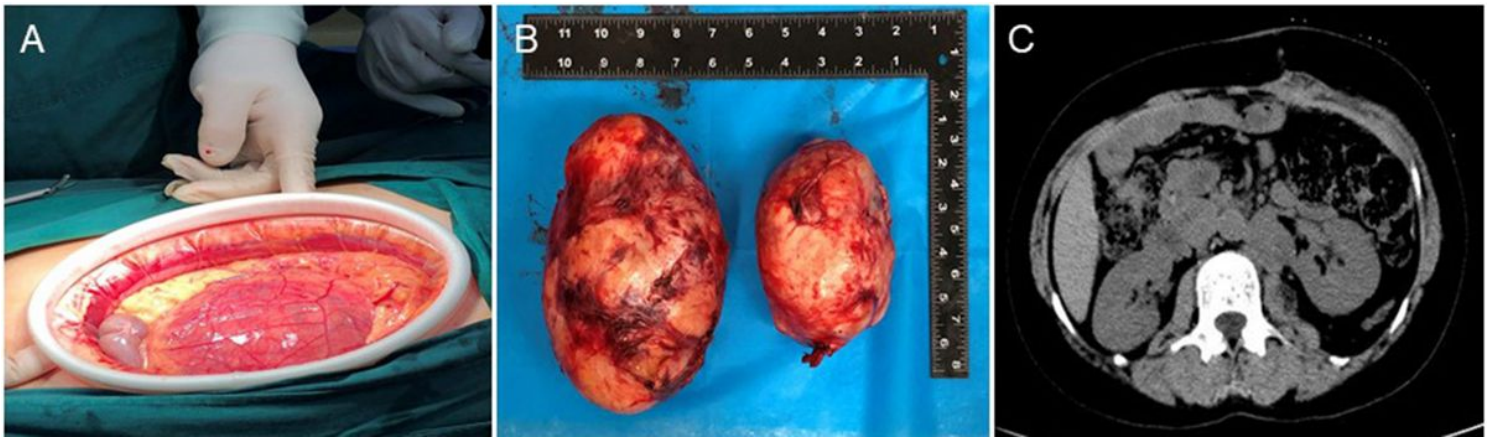
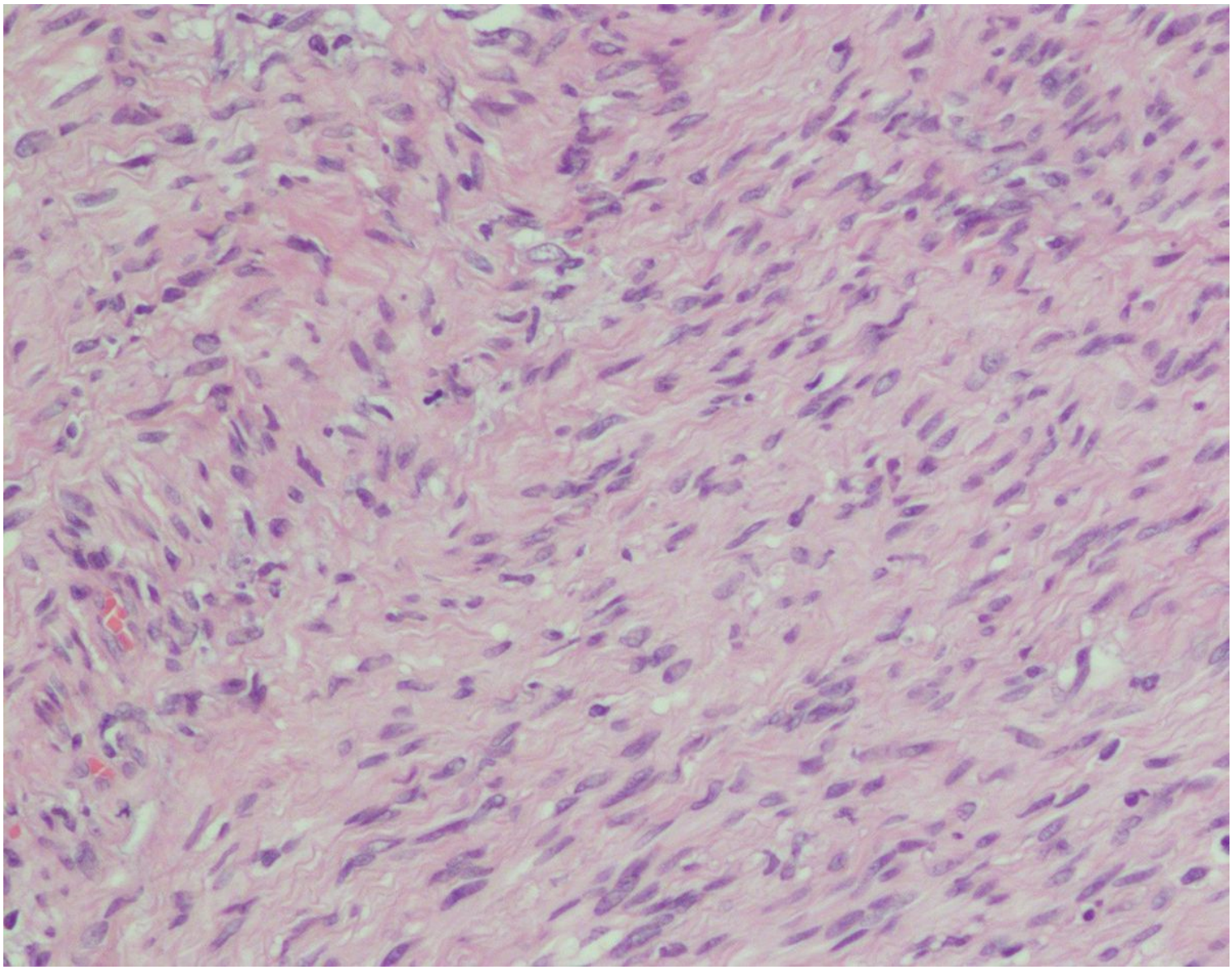


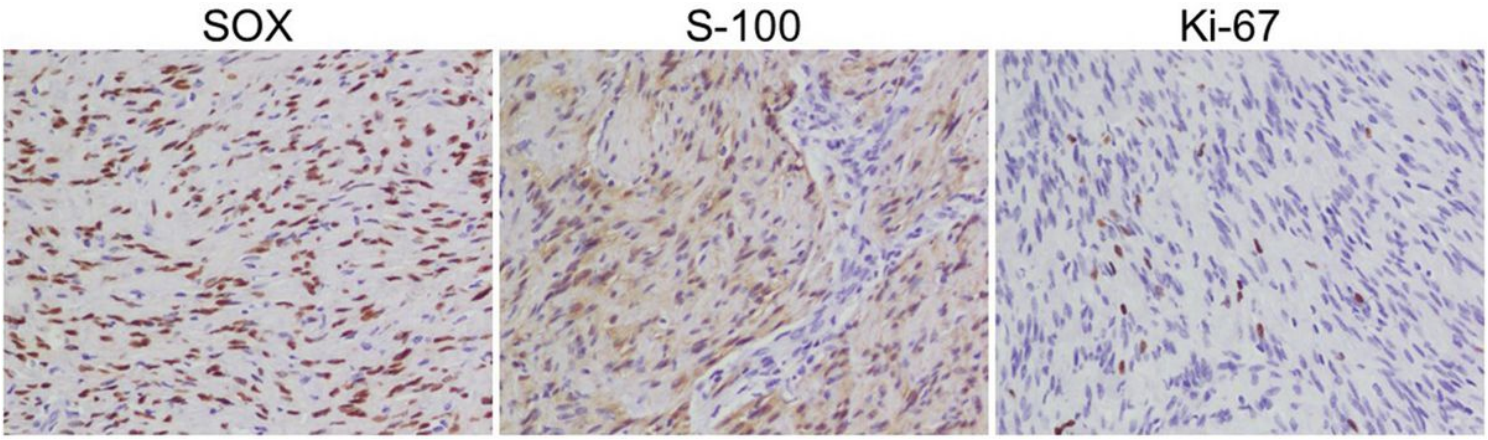

**Figure 2**

Pathological features of the tumor and abdominal CT examination post-surgery. (A) After opening the abdominal cavity, abundant blood vessels can be seen on the tumor surface. (B) The postoperative specimen size is 8.0 cm × 7.0 cm × 5.5 cm and 4.0 cm × 5.0 cm × 6.0 cm. (C) CT scans at 18 months after surgery showed no signs of recurrence.



## Figure 3

Histopathology shows that the tumor parenchyma was composed of Schwann cells with light to moderate atypia in the nucleus (magnification, × 200).




## Figure 4

Immunohistopathology shows S-100 (+), SOX10 (+), and Ki67 (Positive rate 1%) (magnification, × 200).