

## Case Report

# Complete resection of a gigantic ganglioneuroma in the retroperitoneal space

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Received June 19, 2016; Accepted July 23, 2017; Epub September 15, 2017; Published September 30, 2017

**Abstract:** Retroperitoneal ganglioneuroma (GN) is a type of benign neurogenic tumor that originates from the retroperitoneal sympathetic ganglia, with an extremely low morbidity rate. The retroperitoneal space is a suitable environment for the growth of this indolent tumor. Confined by the narrow space, GNs usually invade adjacent vessels and organs, thus making complete tumor resection challenging. Herein, we describe a case of a gigantic GN within the retroperitoneal space, along with a literature review. A 28-year-old woman was diagnosed with a neurogenic tumor that originated from the retroperitoneum based on imaging examinations. She was subsequently admitted to our hospital for resection. Intraoperatively, a grayish-yellow mass of 10.5×6×8.5 cm with a well-defined margin was detected near many important vessels and organs. The tumor was completely resected. The pathological diagnosis of the resected specimen was GN. The postoperative period was uneventful, and the patient was recurrence-free 8 months post-surgery. In this case, the main goals of excision were the identification of the neoplasm boundary and the determination of a possible gap between the neoplasm and adjacent tissues to ensure accurate resection of the tumor.

**Keywords:** Retroperitoneal ganglioneuroma, computed tomography, magnetic resonance imaging

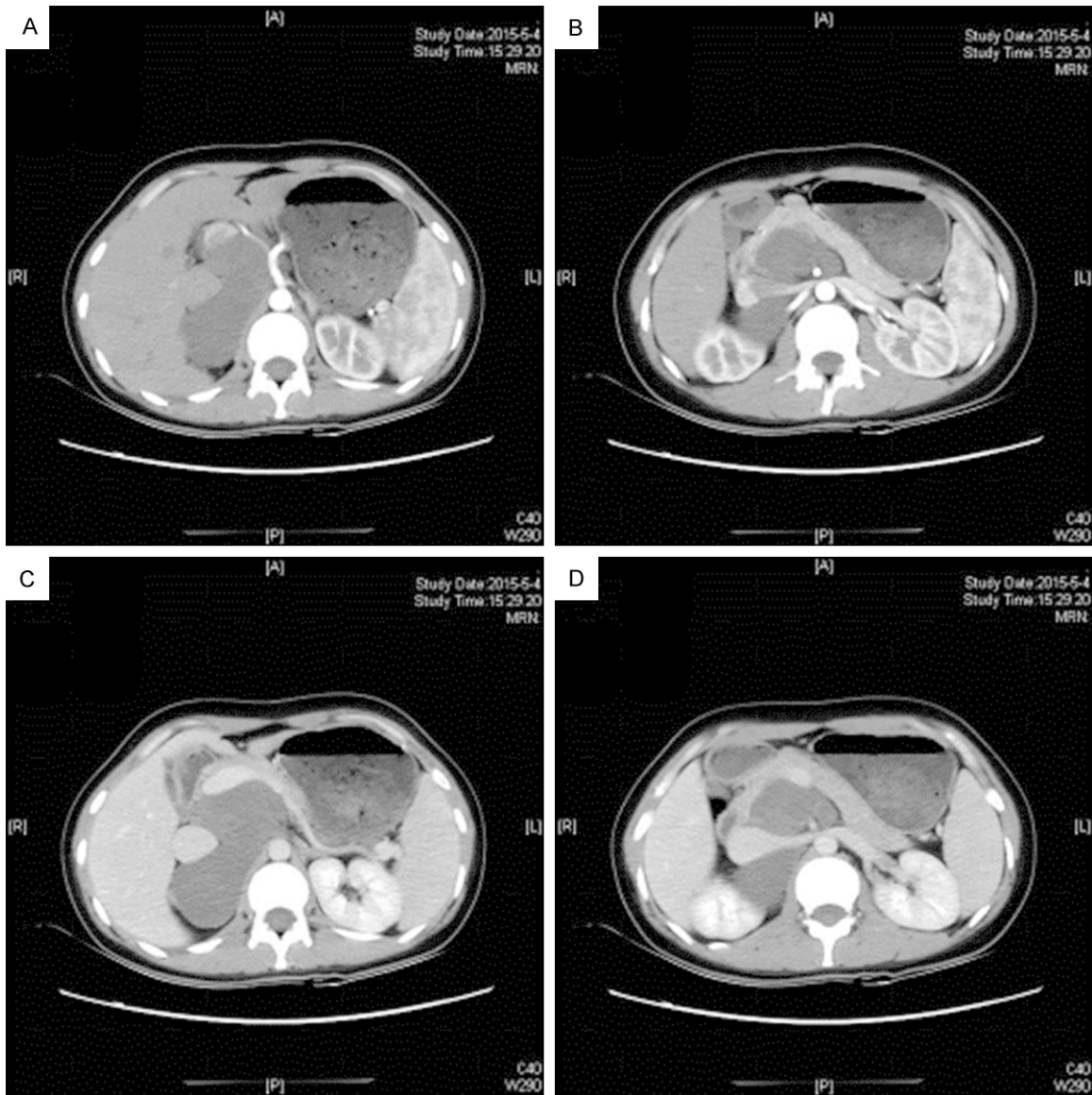
## Introduction

Retroperitoneal ganglioneuroma (GN) is a type of benign neurogenic tumor that originates from the retroperitoneal sympathetic ganglia, with an extremely low morbidity rate [1, 2]. GN occurs primarily in patients aged 20-35 years, and the incidence rate is similar between the sexes. The clinical presentation of GN is typically asymptomatic. The retroperitoneal space is a suitable environment for the growth of this indolent tumor. Confined by the narrow space, GNs usually invade the adjacent vessels and organs, thus making complete tumor resection challenging. The definitive diagnosis can be made only on the basis of the pathological examination findings of the resected specimen, and the prognosis is usually favorable after resection [1, 3]. Herein, we describe a case of a gigantic GN in the retroperitoneal space, along with a literature review.

## Case report

A 28-year-old woman complaining of intermittent abdominal pain was admitted to our hospi-

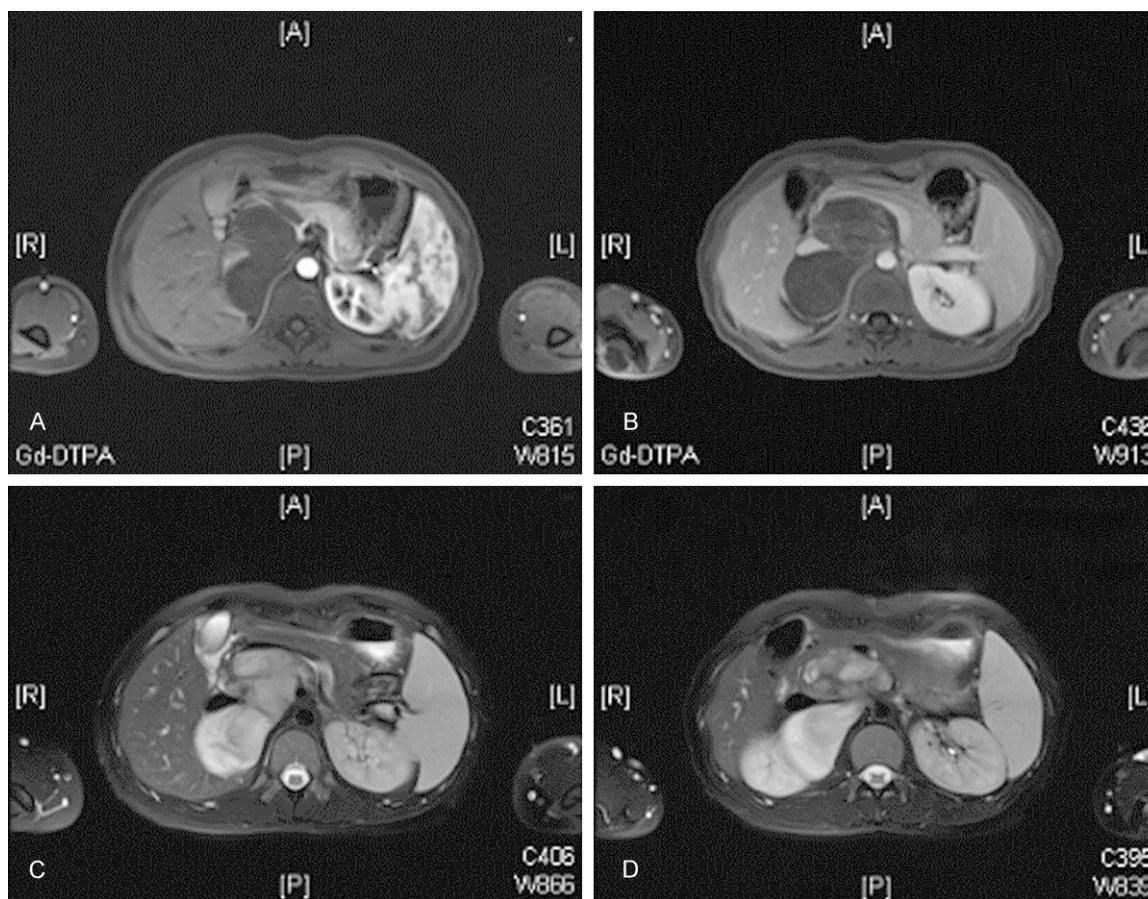
tal on May 6, 2015, owing to a retroperitoneal tumor diagnosed via ultrasonography evaluation. No significant findings were noted on physical examination or in the medical history. The laboratory blood test results were all within the normal ranges. Ultrasonography revealed an irregularly shaped hypoechoic mass with low Doppler signals in the right flank of the retroperitoneal space. On abdominal computed tomography (CT) (**Figure 1**), the retroperitoneal mass was observed as a 10.2×5.8×8.5-cm dumbbell-shaped, homogeneous, and hypodense neoplasm (CT value, 34HU) with a well-defined margin, and was located between the pancreas and the right adrenal gland, exerting a compressive effect on the postcava, pancreas, right renal vein, and adrenal gland. The neoplasm demonstrated slight contrast enhancement. Low and heterogeneous high signal intensities were observed on T1-weighted (**Figure 2**) and T2-weighted magnetic resonance imaging (MRI) scans, respectively, while slight contrast enhancement was noted on contrast-enhanced scans. Based on these findings, a neurogenic neoplasm was suspected. Laparo-



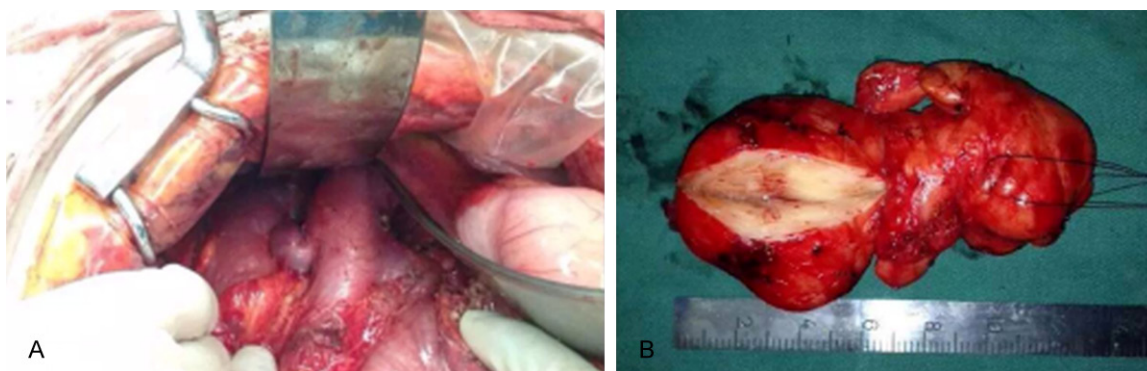
**Figure 1.** Abdominal computed tomography (CT) findings. Arterial-phase CT showing that the tumor was contiguous to (A) the postcava and (B) the right renal vein. Delayed-phase CT showing that the tumor was contiguous to (C) the postcava and (D) the right renal vein.

tomy confirmed a well-encapsulated dumbbell-shaped retroperitoneal neoplasm (**Figure 3A**), measuring 10.5×6.0×8.5 cm, that compressed and partially surrounded the postcava and the right renal vein and that was tightly adhered to the hepatic portal vein, superior mesenteric and splenic veins, celiac trunk, superior mesenteric artery, pancreas, and right adrenal gland. The neoplasm was carefully separated from the adjacent organs and vessels after identifying its margins and making an incision. Complete resection of the neoplasm was performed, with

little hemorrhage and no injury to important organs or vessels. The incised specimen had a grayish-yellow fish-meat-like appearance, without hemorrhage or necrosis (**Figure 3B**). The postoperative pathological diagnosis of the resected specimen was GN (maturing) (**Figure 4**). Immunohistochemical staining revealed positive staining for Vim (**Figure 5A**), S-100 (**Figure 5B**), NF (**Figure 5C**), and CD34 (**Figure 5D**), but negative staining for Bcl-2 and SMA. The Ki67 labeling index was 1% (**Figure 6**). The postoperative period was uneventful, and at the latest



**Figure 2.** Magnetic resonance imaging (MRI) findings. Arterial phase T1-weighted MRI showing that the tumor was contiguous to (A) the postcava and (B) the right renal vein. T2-weighted MRI showing that the tumor was contiguous to (C) the postcava and (D) the right renal vein.



**Figure 3.** A. Intraoperative findings. B. The resected specimen.

follow-up, 8 months postoperatively, the patient was recurrence-free, as confirmed by CT.

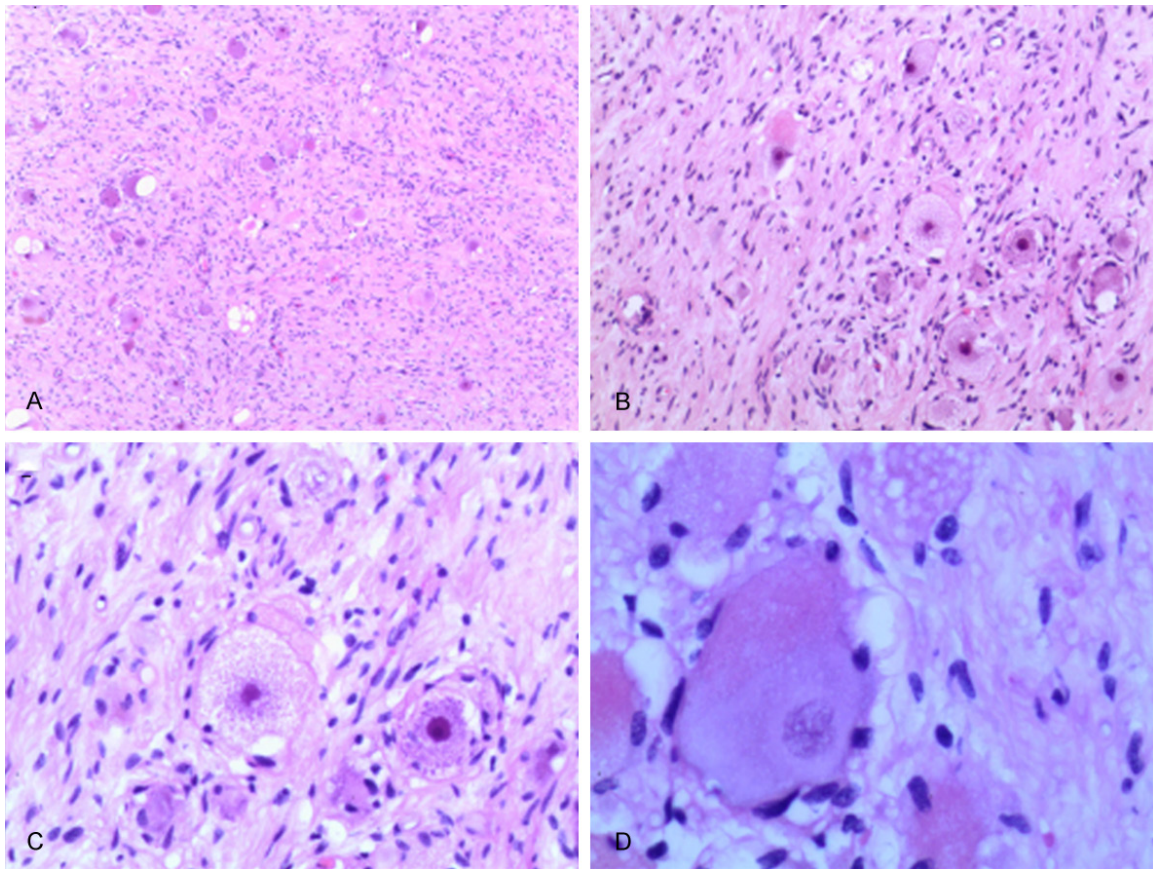
### Discussion

GNs are benign neurogenic tumors that originate from the neural crest cells of the sympa-

thetic ganglia, with an extremely low morbidity rate [1, 2]. Neuroblastoma and ganglioneuroblastoma may progress to GN; however, this is very rare [4, 5].

GNs primarily occur in patients aged 20-35 years and the incidence rate is equal in men





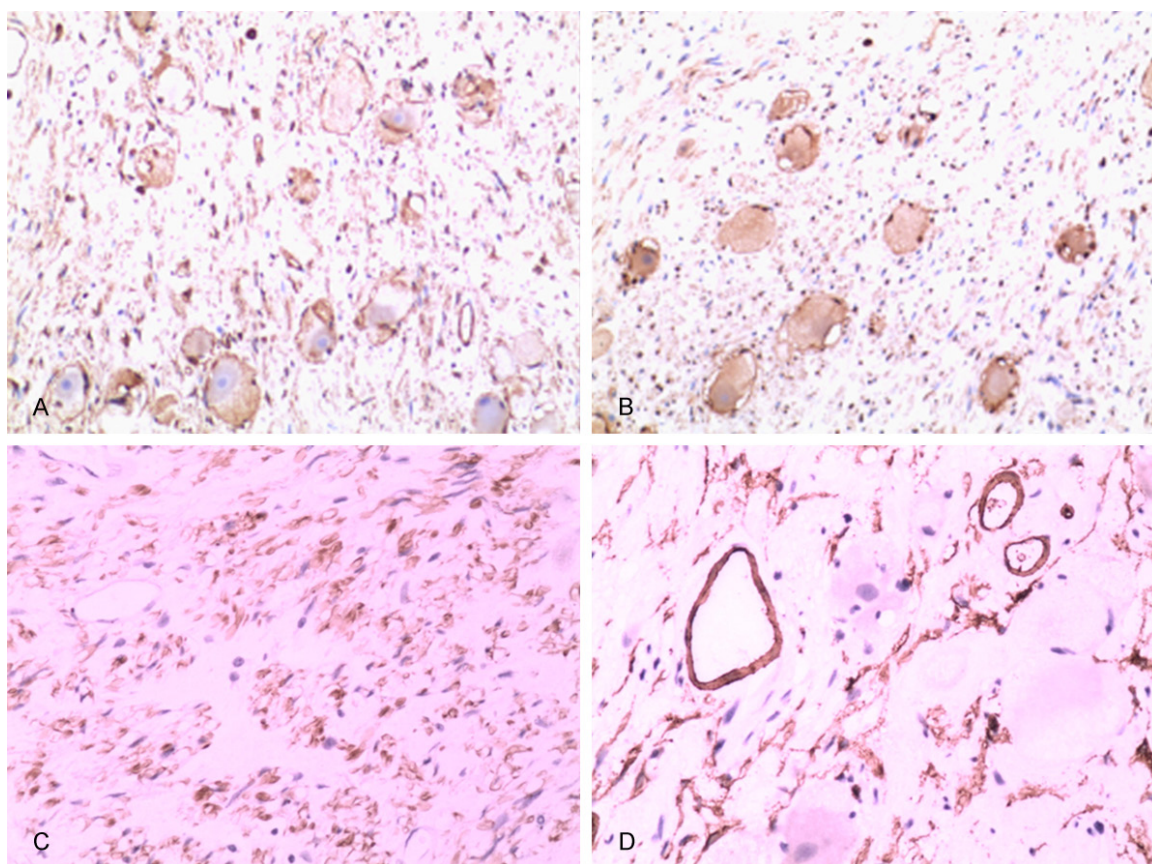
**Figure 4.** Postoperative pathological findings of the resected specimen. A. Hematoxylin and eosin staining (HE), ×40; B. HE, ×100; C. HE, ×200; D. HE, ×400.

and women. It usually arises in the retroperitoneum (32-52%), posterior mediastinum (39-43%), and neck (8-9%) adjacent to the spinal cord [5-7].

Retroperitoneal GNs usually present without any significant symptoms. A minority of patients may present with fever or headache, weight loss, arthralgia, or paralysis of the legs, which could be associated with malfunction of the compressed nerves or inflammatory factors secreted by the tumor [8]. Occasional hormonal secretions of catecholamines, vasoactive intestinal peptides, and testosterone have been reported to cause hypertension, diarrhea, and virilization [5, 9]. In some cases, retroperitoneal GNs might progress to malignant cancer over several months or years [10] by invading the bones directly or by metastasizing diffusely [11].

GNs present low Doppler signals on ultrasonography. In this case, the Doppler signals of the

tumor confirmed its solid texture. On CT scanning, GNs present as voluminous, well-defined, encapsulated masses with a round, oval, or dumbbell shape. The tumors are homogeneous and hypodense due to the massive amounts of mucus present [12]. GNs usually grow throughout vessels and tissues and form pseudopodium-like lesions; they may partially cause obstruction of the vessels but rarely completely obstructs the blood flow [13, 14]. The GN in this case deeply compressed the postcava but did not obviously obstruct the blood flow. The neoplasm showed gradual slight-to-intermediate heterogeneous contrast enhancement in the venous and delayed phases. This postponing manifestation is owing to the massive amounts of mucus in the intercellular space obstructing the contrast instillation [15]. Further, the tumor in our case showed low signal intensity on T1-weighted MRI scans, homogeneous or heterogeneous high signal intensity on T2-weighted scans, and slight contrast enhancement on



**Figure 5.** A. The tumor cytoplasm showed positive staining for Vim. B. The tumor cytoplasm and nuclei showed positive staining for S-100. C. The tumor cytoplasm showed positive staining for NF. D. The tumor cytoplasm showed positive staining for CD34.

contrast-enhanced scans. A whorled appearance on T2-weighted images corresponds to interlacing bundles of Schwann cells and collagen fibers on histology [16].

AsGNs are benign, well-differentiated, slow-growing tumors, complete resection is the preferred management approach [3]. A nephrectomy or Whipple's operation could be executed if there is difficulty separating the tumor from the kidneys or pancreas. Recurrence is a possibility when the tumor is partially resected or spreads due to incorrect or inappropriate technical procedures during operation [17]. The malignancy rate increases with recurrence, and radiotherapy or chemotherapy should not be administered unless the GN becomes malignant [18]. Laparoscopic resection is a suitable procedure that causes less trauma in selected GNs [19, 20]. In the present case, as the neo-

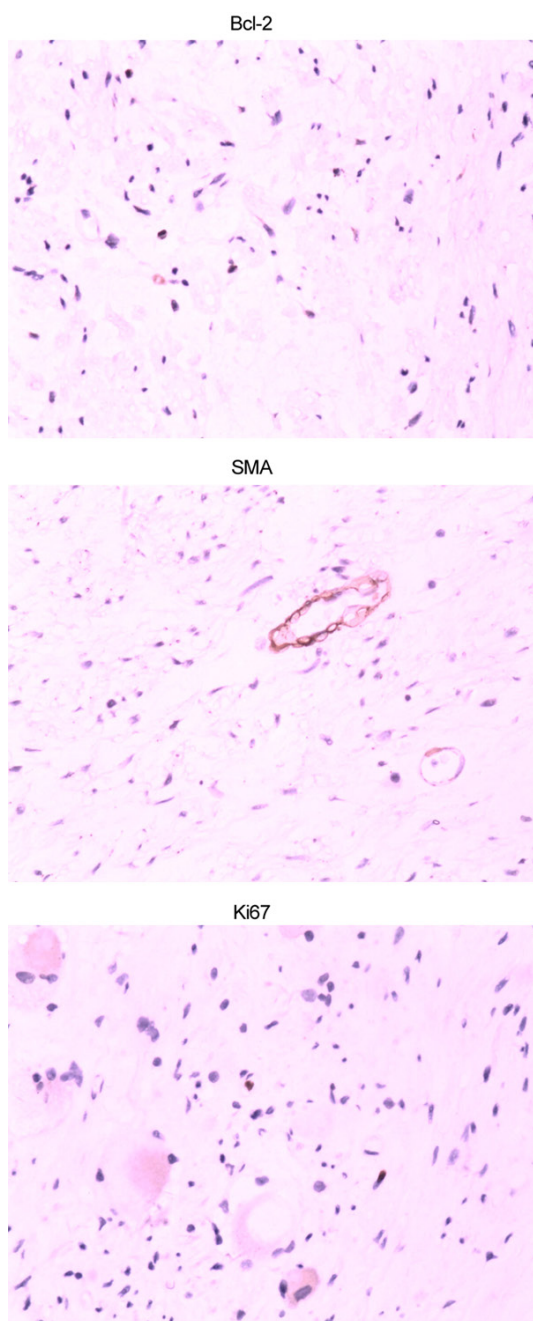
plasm obviously compressed and partially surrounded the postcava and right renal vein and was intimately associated with many important vessels and organs, it seemed difficult to completely resect the neoplasm without injuring other vessels and organs. Therefore, the main goal of the excision was to identify the margins of the neoplasm and a possible gap between the neoplasm and adjacent tissues to ensure accurate resection.

## Disclosure of conflict of interest

None.

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**Figure 6.** The tumor cytoplasm showed negative staining for Bcl-2 and SMA, and the Ki67 labeling index of the tumor cytoplasm was 1%.

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