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Large Asymptomatic Retroperitoneal Ganglioneuroma Displacing Major Abdominal Organs and Vessels in an Adult

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Patient: Female, 29-year-old
Final Diagnosis: Ganglioneuroma
Symptoms: Asymptomatic • incidental finding
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease

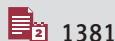
Background: Ganglioneuromas (GNs) are benign neuroblastic tumors. These extra-cranial solid tumors are common in childhood but unusual in adults. Patients with GNs typically do not have any symptoms and the tumors usually are incidental findings. However, if a GN is large enough to compress adjacent organs, complications can occur. Furthermore, even in patients who have incomplete resection of a GN, long-term survival rates are high. After a GN is seen on imaging, the diagnosis usually is made with a biopsy and treatment is with surgery alone.

Case Report: A 29-year-old woman was referred to General Surgery from the Gynecology Clinic for an incidental finding of an abdominal mass on routine ultrasound for secondary infertility and admitted for an investigation. Abdominal and pelvic computed tomography (CT) and magnetic resonance imaging showed a retroperitoneal mass that measured 6.318×22 cm arising from the paravertebral region with intraspinal extension. The mass was displacing the patient's thoracic aorta, abdominal inferior vena cava, and ureters. A CT-guided biopsy revealed a GN. Debulking surgery was performed and a small amount of residual tumor was left in the paravertebral nerve roots. The patient recovered well with no complications. The diagnosis of GN was confirmed with pathology, which was reviewed by the Tumor Board; the Board agreed that only follow-up in the Surgery Clinic was needed. During the patient's last visit, 10 months after surgery, a follow-up CT scan showed that the residual tumor was stable.

Conclusions: GNs are benign abdominal and retroperitoneal tumors that are typically asymptomatic and detected incidentally. Surgical resection is the treatment of choice and even when it is incomplete, the prognosis for patients is excellent.

Keywords: Aorta, Abdominal • Ganglioneuroma • Retroperitoneal Neoplasms

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Background

Ganglioneuromas (GNs) are neuroblastic tumors. Like most neuroblastic tumors, they originate from the neural crest and arise from the sympathetic chain. GNs are one of the most common extra-cranial solid tumors seen in children but they are rare in adults [1]. They are usually found in the posterior mediastinum and retroperitoneum [2]. Patients with GN usually have no symptoms and their tumors are discovered incidentally [3]. A wide variety of symptoms can occur, however, if a GN is large enough to compress adjacent organs. They include back pain associated with compression of paravertebral structures and abdominal pain, nausea, and vomiting

from compression of the bowels [4,5]. In rare cases, GNs can secrete hormones such as catecholamines, vasointestinal peptides, and androgens [6]. GNs are considered benign and long-term survival rates are high, even in patients who undergo incomplete resection [7]. After an incidental radiographic finding, the diagnosis is made based on biopsy, and surgery alone is the usual treatment [1].

Case Report

A 29-year-old woman with no significant medical or surgical history had no symptoms except failure to conceive. She was referred

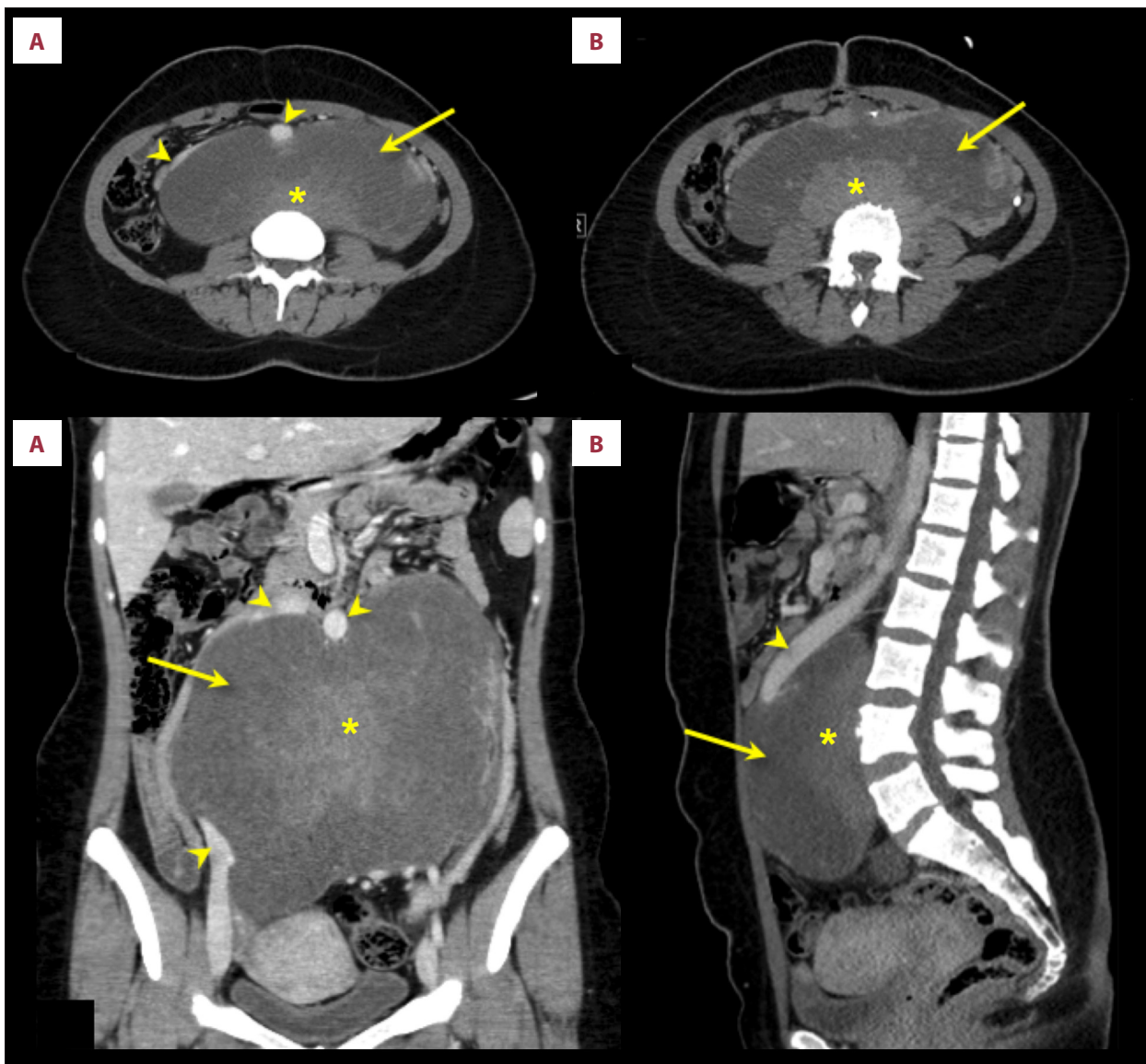


Figure 1. (A, B) Axial, (C) coronal, and (D) sagittal computed tomography scans with intravenous contrast reveal a large retroperitoneal soft tissue mass (arrow) anteriorly displacing the aorta and the inferior vena cava (arrowheads). The mass is predominantly hypodense with enhancing soft tissue components (asterisk).

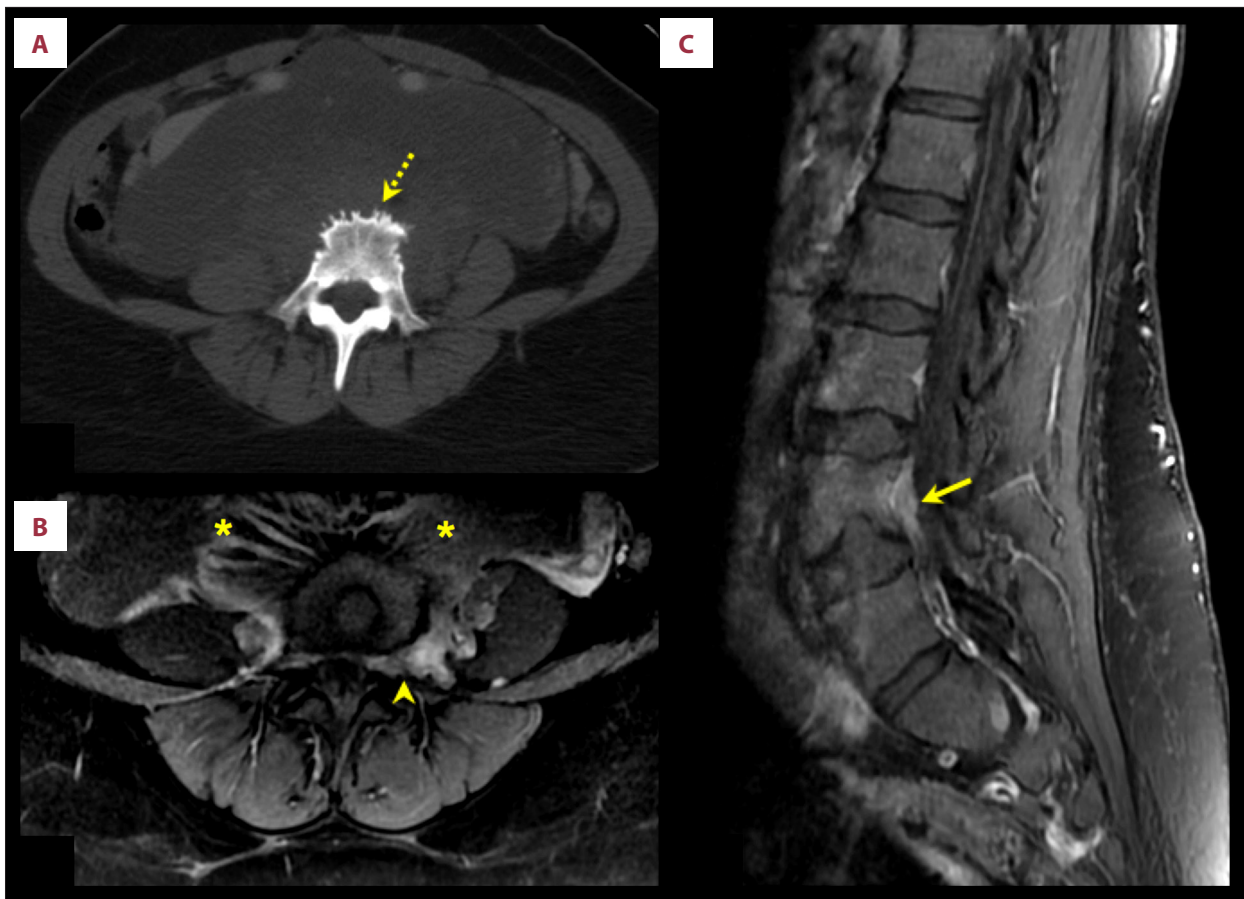


Figure 2. Relation of the mass to the spine. (A) An axial computed tomography scan of the bone shows erosions and remodeling of the L4 vertebral body (dotted arrow). (B) Axial and (C) Sagittal post-contrast T1-weighted magnetic resonance imaging shows a partially visible, heterogeneously enhancing retroperitoneal perivertebral mass (asterisks) surrounding the L4 vertebral body. It extends into the left L4-5 neural foramen (arrowhead) with an anterior epidural enhancing component opposite L4.

to General Surgery from the Gynecology Clinic after an abdominal mass was found incidentally on her routine ultrasound (US) examination. An US performed during the patient's first pregnancy had shown a small mass in her abdomen. The patient was lost to follow-up and when she presented 6 years later for secondary infertility, the mass had increased in size and was more defined.

On admission, the patient was asymptomatic and not taking any medication; her family history was negative for malignancy. Abdominal and pelvic computed tomography (CT) showed a retroperitoneal mass extending from the pancreas to the pelvis that measured 6.31822 cm. It contained small calcifications and papillary soft tissue that was enhanced. The mass displaced the abdominal inferior vena cava (IVC) anteriorly and the ureters laterally, causing mild hydronephrosis without invasion (Figure 1). To confirm the diagnosis, the patient underwent a CT-guided biopsy. The pathology report described a GN.

Magnetic resonance imaging (MRI) of the patient's abdomen was performed to localize the lesion accurately before surgery.

The MRI showed a large, retroperitoneal, prevertebral, heterogeneous soft tissue tumor mass extending from L2 to L5-S1. The lesion was infiltrating the psoas muscles bilaterally, mainly on the left side. Intraspinous extension was visible through the widened left neural foramen at the L4-L5 level, with a ventral epidural enhancing component at L4 that displayed a curtain sign and measured approximately 227 mm. An abnormal marrow signal with ventral vertebral body erosive changes denoting infiltration (sun ray appearance) was visible in the L4 vertebral body (Figure 2).

The patient was counseled and gave consent for a debulking surgical excision. A full midline laparotomy was performed to explore the abdominal cavity. A huge retroperitoneal mass was visible, which pushed the intra-abdominal structures anteriorly. The descending colon was mobilized along the line of Toldt up to the splenic flexure to expose the retroperitoneal structures. The aorta was identified and meticulous mobilization was performed with an infrarenal control using a vessel loop. After mobilization, the right common iliac artery also

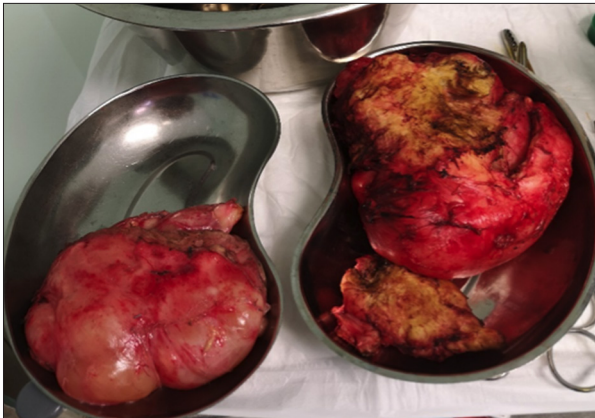


Figure 3. Surgical specimen from the retroperitoneal mass, which was divided down the middle into 2 parts.

was secured with a vessel loop. Then, the mass was carefully separated from the IVC and multiple small feeding vessels were clipped. Because the left common iliac vein had collapsed within the mass, it was sacrificed and ligated. Once the mass was completely separated from the aorta, the small feeding vessels were clipped. We encountered bleeding from the posterior wall of the aorta where the mass was attached, but it was controlled immediately.

We tried to mobilize the mass from the posterior abdominal wall as a unit, but it was difficult, given the anterior position, so we elected to divide it in the middle. Each half was removed separately (Figure 3). A small amount of residual disease was left in the paravertebra at the nerve roots.

The patient tolerated the surgery well and had no complications. She was in the Intensive Care Unit for 1 day before being returned back to the ward. She recovered from the procedure very well and was discharged after 5 days in good

condition. An abdominal CT scan was performed before discharge as a postoperative baseline to assess for residual disease and in anticipation of future evaluation for disease progression (Figure 3).

The histopathology report showed morphology similar to what was found on the percutaneous biopsy. The tumor was predominantly composed of Schwannian stroma embedded in a loose myxoid matrix and associated with scattered mature ganglion cells, which confirmed the diagnosis of GN. Despite extensive sampling, there was no evidence of immature neuroblastic components or neuritic processes (Figure 4). The case was discussed postoperatively by the Tumor Board and the plan was to have the patient follow-up with the Surgery Clinic; systemic therapy was unnecessary.

The patient followed up regularly with the Surgery Clinic. Her last visit was 10 months after surgery. She remained healthy with no symptoms until 1 month ago, when she was admitted and received conservative treatment for an adhesive bowel obstruction. A CT scan done during her admission showed the development of small bowel obstruction with a transition zone at the terminal ileum and stable residual disease (Figures 5, 6).

Discussion

GN, neuroblastomas, and ganglioneuroblastomas are peripheral neuroblastic tumors. Of these, GN is the most differentiated and mature tumor, consisting of nerve fibers and Schwannian cells [8]. Moreover, GN is generally considered benign and diagnosed mostly in children and sometimes in adults. These tumors arise from the sympathetic chain. The most common sites of presentation are the posterior mediastinum, retroperitoneum, and head and neck area [5]. GNs usually produce no

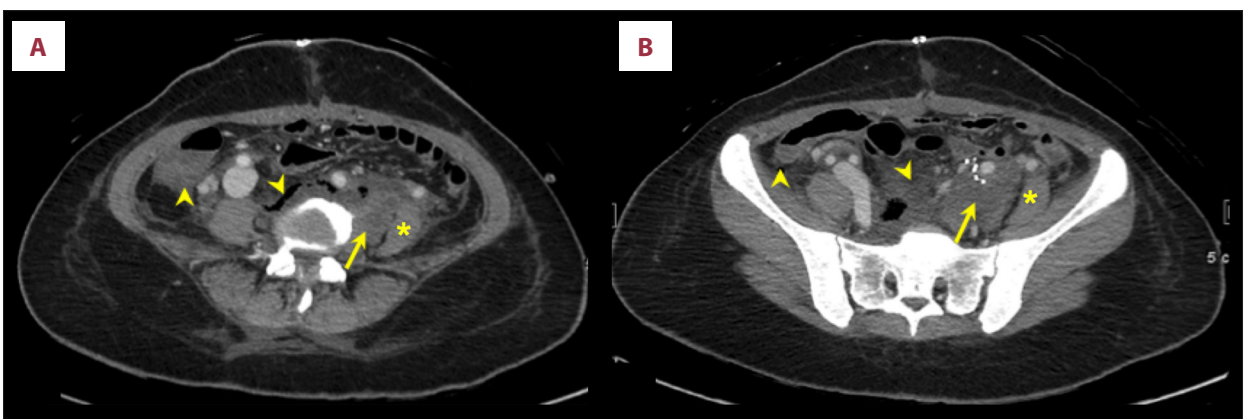


Figure 4. (A, B) Contrast-enhanced axial computed tomography scans of the retroperitoneal mass taken after resection. They show immediate postoperative changes in the form of pneumoperitoneum, pneumoretroperitoneum, and intraperitoneal and retroperitoneal free fluid (arrowheads). There is a hypoenhanced soft tissue residual close to the anterior and medial to the left psoas muscle (asterisk) and posterior to the common left iliac vessels (arrows).

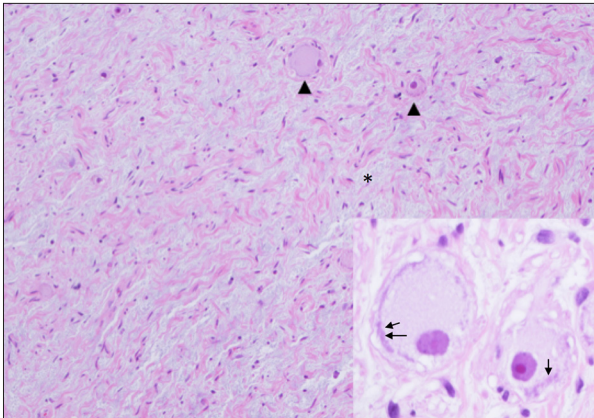


Figure 5. At low magnification, the tumor is predominantly composed of Schwannian stroma with a loose, gray-to-blue, myxoid background (asterisk) and scattered ganglion cells (arrowheads). At higher magnification, the inset shows mature ganglion cells with the prominent nucleoli and basophilic granules at the periphery of the cytoplasm (Nissl substance, arrow).

symptoms and are found incidentally. A wide variety of symptoms of compression can occur, however, depending on the tumor's size and location. Symptoms such as back pain are due to compression of paravertebral structures and abdominal pain, nausea, and vomiting are due to compression of the bowels [5]. Aortic compression syndrome is another symptom that can present if a GN is large enough to compress adjacent vessels.

In rare cases, GNs can secrete hormones such as catecholamines, vasointestinal peptides, and androgens [6]. The imaging modalities of choice are CT and MRI. GNs present as well-defined, oval-shaped solid masses that do not commonly contain cysts, unlike other types of neuroblastic tumors [9]. On MRI, GNs have low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [9], so exact radiological features are lacking for these tumors. The diagnosis, therefore, can be challenging and is more accurately reached with pathology studies after an imaging-guided biopsy.

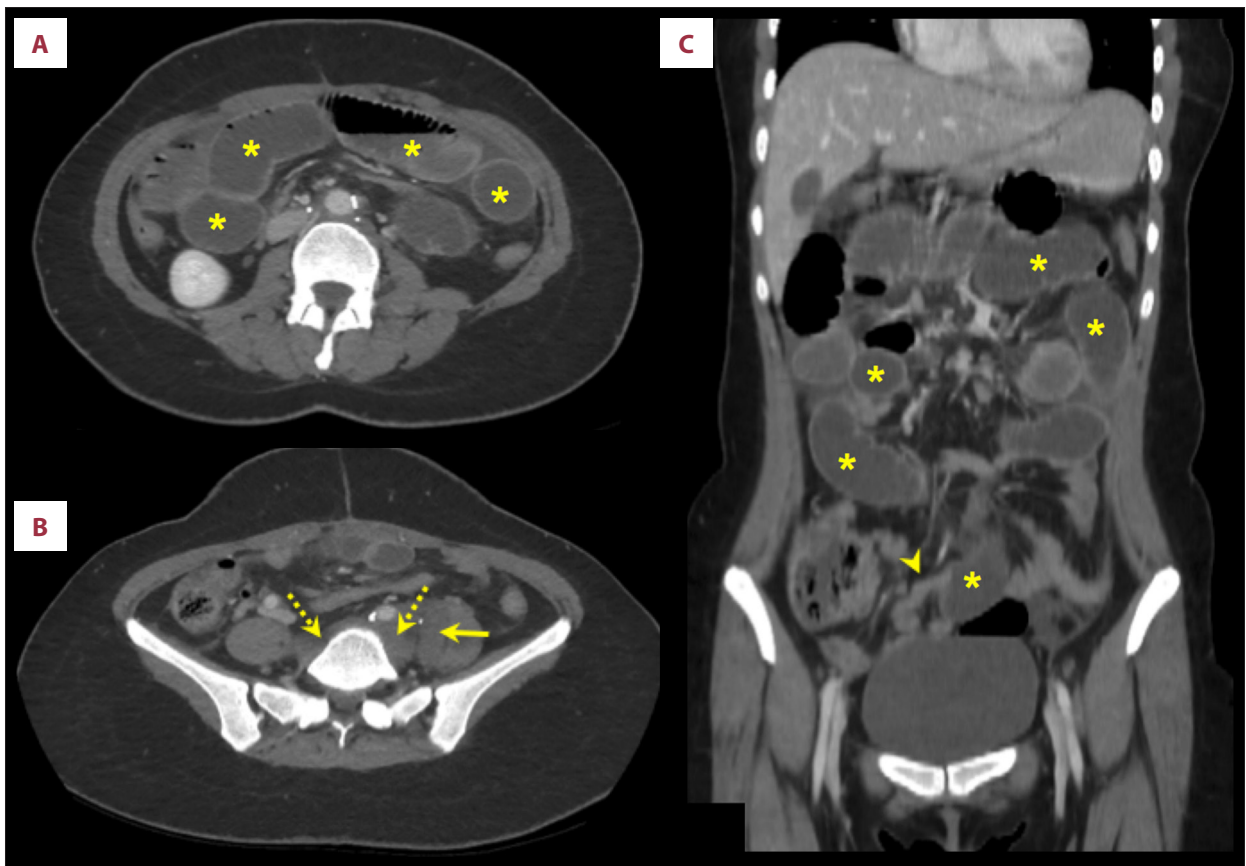


Figure 6. Contrast-enhanced. (A, B) Axial and (C) coronal computed tomography scans of the abdomen performed 9 months after surgery. They show interval development of small bowel obstruction (asterisks) with a transition zone at the distal ileum (arrowhead). A residual retroperitoneal mass is still visible medial to the left psoas muscle (marrow) and in the perivertebral region at the level of L4 and L5 (dotted arrows).

Treatment for GN is with surgical resection alone and the prognosis is excellent even when it is incomplete. These tumors are considered benign and do not commonly progress into malignancy. In rare cases in which ganglioneuroblastoma intermixed (GNBI) is present, however, progression can occur, depending on the stage; therefore, follow-up after resection is recommended for reassurance.

Case reports on GNBI are rare [10]. Open surgery is preferable when operating on a tumor near important blood vessels in the abdominal cavity [11]. When performing surgical resection, however, injury to adjacent organs and structures and postoperative complications are the main concerns. In a retrospective study of 146 children with GN, surgical tumor resection resulted in excellent long-term survival; 22 of the patients (15%) had surgery-related complications, of which 2 were fatal and 7 were severe (eg, Horner syndrome, rupture of the thoracic aorta, and thoracic hemorrhage) [7]. Trans-arterial embolization has been described as a treatment option to avoid surgical complications. It was reported in a small number of cases, with variable efficacy in terms of reduction in size and improvement symptoms [12]. It was not an option in our case because we elected to perform the maximum procedure for a young, fit patient who had a huge mass.

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Conclusions

GNs are benign tumors that present in the abdominal and retroperitoneal cavities. They are usually asymptomatic and discovered incidentally, but patients with them can present with various symptoms, depending on tumor size and location. Surgical resection is the treatment of choice for GNs. The prognosis is excellent, with a low incidence of complications, even when resection is incomplete.

Acknowledgments

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