

## Laparoscopic Resection of a Retroperitoneal Ganglioneuroma Abutting the Iliac Vein: A Case Report and Review of Literature

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### Abstract

**Background:** Ganglioneuromas (GNs) are rare, benign tumors derived from neural crest cells. They can grow anywhere in the body where sympathetic nerve fibers are found, but most commonly occur in the retroperitoneum and frequently grow near blood vessels. This can make surgical resection challenging. GN should, therefore, be included in the differential diagnosis when evaluating retroperitoneal masses, especially those that are adjacent to major vascular structures.

**Case Presentation:** A 29-year-old male presented to the emergency department with epigastric pain and was incidentally found to have, by computer tomography scan (CT), a 3.7 x 3.3 cm mass in the pelvis abutting the right iliac vessels. The patient underwent diagnostic laparoscopy with resection of this mass. The pathology report showed the mass to be a ganglioneuroma.

In this report we present a review of the literature, discuss the unique characteristics of GN, and provide a detailed description of our surgical approach for the laparoscopic resection of a GN abutting the right iliac vein. To our knowledge, there are only a few case reports on GN that utilize a laparoscopic approach for tumor resection, and there are no recorded reports of a tumor in this location.

**Keywords:** Ganglioneuroma, extra-adrenal ganglioneuroma, neuroblastic tumor, retroperitoneal tumor.

### Introduction

Ganglioneuromas (GNs) are rare, benign tumors that originate from embryonic neural crest cells [1-4]. They are encapsulated, slow-growing neoplasms that arise from sympathetic ganglion and can subsequently occur anywhere along the sympathetic chain [2,4,5]. GNs are most commonly asymptomatic and are found incidentally in the retroperitoneum and around blood vessels [4-6]. As such, they are typically managed by open surgical resection. However, we present a 4.8 cm incidental GN found abutting the right iliac vein that was successfully removed laparoscopically. Our patient subsequently experienced a relatively short operative time, a short length of stay, and a negligible estimated blood loss, pointing to the advantages

of laparoscopy over open surgery in the resection of GN when performed by experienced laparoscopic surgeons.

### Case Report

A 29-year-old morbidly obese male, with a body mass index (BMI) of 45 kg/m<sup>2</sup>, presented to the emergency department with epigastric pain. During his work-up with a CT scan, the patient was incidentally discovered to have a 3.7 x 3.3 cm homogenous, low attenuating mass in the pelvis, abutting the right iliac vessels (Fig. 1). This finding was thought to be due to an enlarged iliac lymph node, raising concern for lymphoma. Given the large size of the lesion and the need to rule out malignancy, the patient underwent a diagnostic laparoscopy with plan for biopsy which was changed to a resection.

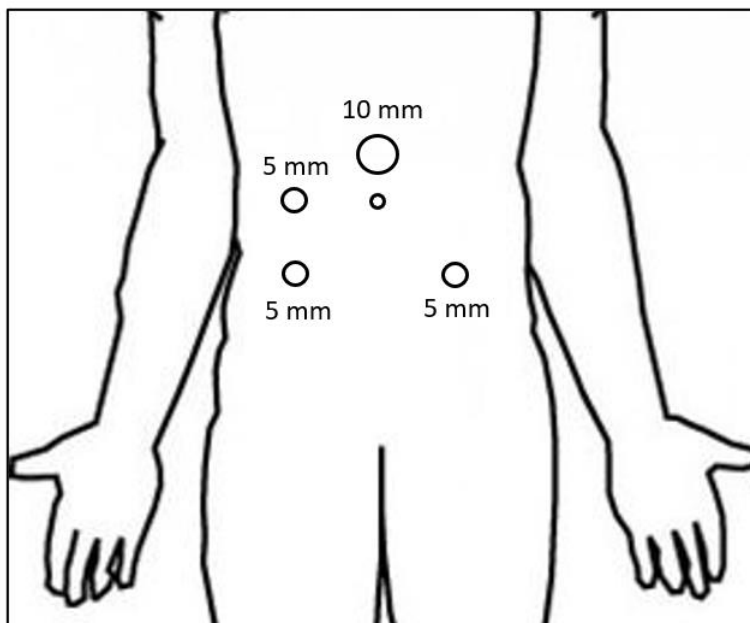


**Figure 1:** CT of the abdomen and pelvis demonstrating a homogenous, low-attenuating, retroperitoneal mass abutting the right iliac vessels, measuring 3.7 x 3.3 cm.

**Surgical Technique:**

The patient was placed in the supine position with both arms tucked to his side. Under general endo-tracheal anesthesia the abdomen was prepped and draped in the usual sterile manner. A Veress needle was used for entry at the supraumbilical site to create the pneumoperitoneum under pre-set pressure of 15 mm Hg. A 10 mm trocar was

then placed at the supra-umbilical location, which was used for insertion of the 10 mm laparoscope, as well as insertion of the specimen retrieval bag at the end of the procedure. Three additional 5 mm working trocars were then placed under direct visualization. One trocar was placed in the right lower quadrant, and one lateral to the right of the umbilicus for the operating surgeon. The third trocar was placed in the left lower quadrant for exposure.



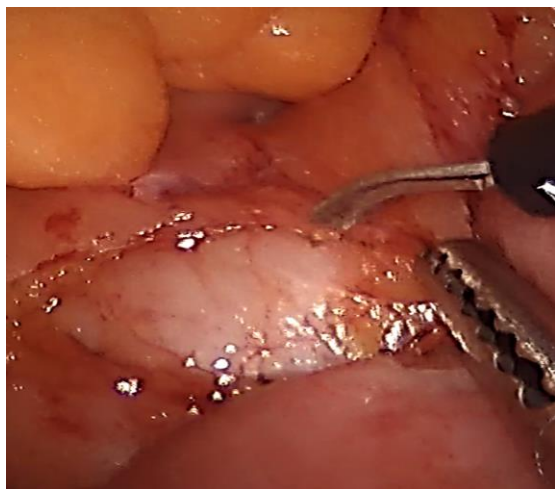
**Figure 2:** Laparoscopic port set-up- 10 mm supraumbilical port used for camera and specimen retrieval bag, 5mm working port in RLQ, 5 mm working port in LLQ, 5 mm working port lateral to the right of the umbilicus

Consistent with the patient’s body habitus, he was noted to have a large amount of thick omentum and intraabdominal fat. The challenge of retracting these structures was relieved by placing the patient in steep Trendelenburg position and using a grasper to retract the redundant sigmoid, allowing for exposure of the retroperitoneum and identification of the pelvic mass over the right iliac vein at the sacral promontory. (Figure 3).

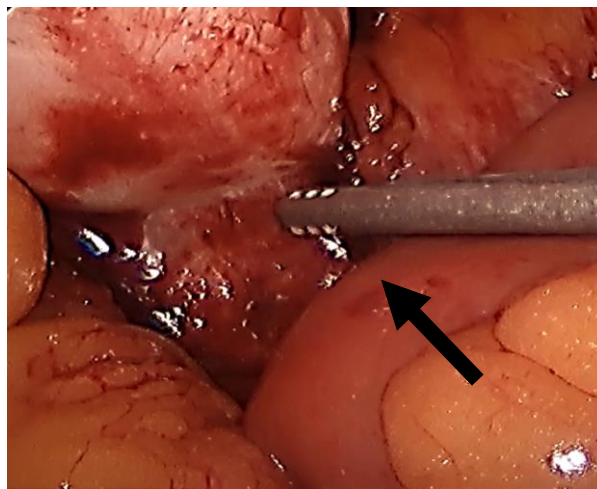
The peritoneum overlying the mass was opened using a laparoscopic scissor attached to cautery (Figure 3). Initially, an attempt was made to excise a piece of the mass for biopsy. However, the tissue was found to be very firm and difficult to cut through, suggesting that the lesion was not a lymph node. We then began to dissect around the

mass circumferentially using the laparoscopic scissor and the suction irrigator interchangeably until the tumor began

to shell out and peel away from the right iliac vein (Figure 4).



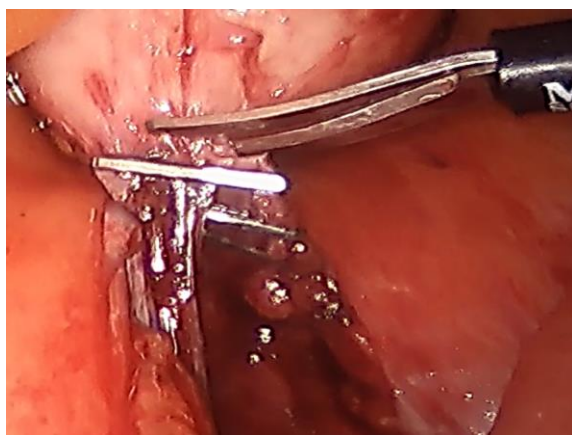
**Figure 3:** Mass exposed



**Figure 4:** Suction dissection of the mass off of the right iliac vein (indicated by arrow) overlying the sacral promontory.

Once the mass was freed from nearly all attachments, it appeared as though there may have been denser tissue adherent to the vein. To prevent potential bleeding, several clips were placed across the remaining attachments, which were divided using the scissor (Figure 5). The

retroperitoneum was then closed using a running absorbable 2-0 V-loc suture [Covidien, Mansfield, MA]. The mass was then placed inside a specimen retrieval bag and brought through the supraumbilical trocar site (Figure 6).



**Figure 5:** Division of final attachments



**Figure 6:** Tumor in retrieval bag

Of note, given the proximity to the iliac vein, we were prepared for conversion to an open procedure immediately, should significant uncontrolled bleeding be encountered at any point in the operation.

Link to surgical video of case- <https://youtu.be/1bYP89aRjck>

The total operative time was 84 minutes and the estimated blood loss was minimal (less than 10 ml) from the retroperitoneal fat, which was easily controlled using electrocautery. The patient's post-operative course was uneventful, and he was discharged home the following day after tolerating a diet.

**Pathology:**

Grossly, the tumor was noted to be a tan, well-circumscribed, encapsulated mass measuring 4.8 cm (Fig. 7). Histology was significant for numerous mature ganglion cells, as well as Schwann cell components, with S100 positive in both cell types (Fig. 8). These findings were consistent with ganglioneuroma.





**Figure 7:** Surgical specimen



**Figure 8:** S100 - S100 immunohistochemical stain at 100x magnification demonstrating positivity within both the Schwann cells (red arrow) and ganglion cells (blue arrow)

## Discussion

GNs belong to a spectrum of peripheral neuroblastic tumors that are divided into 3 groups according to their degree of cellular and extracellular maturation: GN, ganglioneuroblastoma, and neuroblastoma [1,3,4,7,8]. The cell of origin is derived from embryonic neural crest cells, and GNs are thought to represent the final stage of maturation from neuroblastoma [3]. Histologically the ganglion cells are mature with no immature neuroblastic elements present [2]. As such, aside from few rare cases of sarcomatous transformation of GN to a malignant peripheral sheath tumor, GN is a benign neuroblastic tumor, whereas ganglioneuroblastoma is of intermediate differentiation, and neuroblastoma is a highly malignant lesion [9].

In addition to the aggressive nature of immature neuroblastic tumors, they tend to occur in younger patients (median age, just under 2 years), while mature tumors are found in older children and adults [8,9], such as the patient presented in this case. Likewise, GNs can occur in all ages but occur more frequently in individuals between the ages of 10 and 40 years old [6].

GNs are rare tumors and the literature concerning their exact incidence is sparse but is estimated at 1 per million [10]. However, of the cases reported, over half of the tumors were located along the sympathetic ganglion in the retroperitoneum and were primarily found around the adrenal gland [2,5,6,9]. The remainder are located in the mediastinum, the cervical region, and other uncommon sites, such as the skin, pharynx, paratesticular region, and gastrointestinal tract [2]. Gastrointestinal GNs may be associated with MEN 2B, neurofibromatosis-1, Cowden syndrome, tuberous sclerosis, and familial adenomatous polyposis [2]. Of the retroperitoneal GNs, 49% originate in the adrenal gland and 51% are found in extra-adrenal locations [5].

GNs typically follow a benign course, but grow to be very large tumors, with an average size of 7 cm [3]. In their early stage, most GNs are asymptomatic and are typically found incidentally [11,12], such as was the case with our patient. As the tumor grows larger, patients may present with different symptoms resulting from mass effect [5,11].

Much like other retroperitoneal tumors, one of the primary features of a GN is the propensity to grow adjacent to and/or encase major vascular structures [2,13], which can make complete excision both challenging and perilous. Despite the tendency to surround major blood vessels, they typically do not compromise the vessel lumen [9]. We found this to be true in our case, as the tumor abutted the right iliac vein but did not invade or compress it, and it contained no vascular attachments upon careful inspection of the tumor following resection.

Occasionally, a GN may secrete catecholamines, vasointestinal peptide (VIP), androgens, or all of these [9]. In these cases, symptoms like hypertension, abdominal cramping, diarrhea, and virilization may subsequently develop [2,5,9,11].

Ultrasound (US), CT scan, and magnetic resonance imaging (MRI) are the imaging modalities used for assessing GNs. Mediastinal and retroperitoneal tumors are typically detected by US or CT, whereas tumors located at the nerve root are diagnosed using MRI [11]. US of a GN will demonstrate a homogenous, hypoechoic mass, while a CT will show a homogenous, low attenuation mass [14]. Unlike our case, some GNs show extensive calcification, but most are nondescript, homogeneous masses on CT [7]. MRI findings of GN show low signal intensity on T1-weighted images and high signal intensity on T2-weighted images [14].

Overall, CT is the imaging test used most frequently to assess neuroblastic tumors due to the ability to reveal the extent of the tumor, the organ of origin, regional invasion, vascular encasement, and adenopathy [8]. However, CT scanning might underestimate the tumor size by up to 45%,

whereas MRI does so by ~20% [15-17]. We found this to be true in our case, as the maximal dimension of the tumor on CT was measured to be 3.7 cm, whereas its actual size was 4.8 cm in its largest dimension.

GNs grossly appear as well-circumscribed, encapsulated, rubbery tumors [7], with a homogenous, grayish-white cut surface [2]. Histologically, the lesion may have somewhat irregular margins and consists of a proliferation of haphazardly arranged S-100 protein-positive Schwann cells and variably prominent, scattered mature ganglion cells [7], as was seen in our case of GN.

Surgical resection represents the only choice for both definitive diagnosis and treatment of GN (18). Due to the large size of these tumors and the tendency to grow around blood vessels, most cases have historically been resected using open techniques [18,19]. However, with the improvement of laparoscopic surgical skills and equipment, all forms of surgery are increasingly being performed via laparoscopy [19,20]. For example, laparoscopic retroperitoneal surgeries are routinely performed for adrenalectomy, retroperitoneal lymphadenectomy, and nephrectomy [21]. However, the number of retroperitoneal GN excisions approached via laparoscopic surgery is limited by the rarity of the tumor [19]. To our knowledge there are only a handful of case reports on laparoscopic and robotic approaches for resecting these tumors, and most of those cases involved adrenal GNs [12,18-20]. This highlights the importance of reporting the surgical approach of extra-adrenal GNs, especially those that occur in the pelvis near the iliac vessels, which makes our case unique, as a GN at this location has not been previously recorded in the literature.

One small retrospective study comparing open versus laparoscopic resection of GN found that laparoscopic surgery conferred an advantage over open surgery when looking at length of stay (LOS) and estimated blood loss, with no difference in post-operative complications [12]. Laparoscopic surgery was associated with an average LOS of 5.0 vs. 7.3 days for open surgery and an estimated blood loss of 69.5 mL vs. 157.5 mL for open surgery [12]. Comparatively, the LOS in our case was 1 day and blood loss was minimal (less than 10 ml).

Laparoscopic resection via a retroperitoneal approach is also a possibility for surgical resection of GN and has demonstrated good results in the limited literature that exists. One study cited 18 cases of GN resected laparoscopically via a retroperitoneal approach, with an average operative time of 86 minutes, an average estimated blood loss of 85 mL, an average length of stay 5 days, and no post-operative complications [18]. Our transabdominal laparoscopic approach yielded similar results, with no complications and an operative time of 84 minutes, but with less than a third of the blood loss when compared with the retroperitoneal approach, and our patient was discharged on postoperative day 1.

Complete surgical excision is an effective and successful treatment for GN and the prognosis is usually excellent,

even in cases of capsule rupture or incomplete resection [11]. Unless the GNs are associated with ganglioneuroblastoma changes, preoperative or postoperative radiotherapy or chemotherapy is both unnecessary and ineffective [22]. Local recurrence is uncommon, especially in cases of complete resection, but a small incidence has been reported. For this reason, complete surgical resection is recommended and periodic radiologic surveillance is suggested following resection [8], though no protocol for this surveillance has been developed.

## Conclusion

GNs are benign tumors that should be part of the differential diagnosis when a surgeon is faced with an unknown mass in a retroperitoneal location. In their early stage, GNs are asymptomatic and are found incidentally. As these tumors grow, symptoms may ensue, resulting from impingement on adjacent structures. Additionally, the tendency of GN tumors to abut or encase vascular structures can be daunting to a surgeon preparing for resection, and thus, they have been commonly removed using an open surgical approach. However, with the increased experience in laparoscopic surgery, our case demonstrates that laparoscopic resection of GNs can be performed safely and carries with it the benefits of minimally invasive surgery.

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