

# Laparoscopic Resection of Large Adrenal Ganglioneuroma

G. N. Zografos, MD, K. Kothonidis, MD, C. Ageli, MD, N. Kopanakis, MD, K. Dimitriou, MD, E. Papaliodi, MD, G. Kaltsas, MD, M. Pagoni, MD, G. Papastratis, MD

## ABSTRACT

We report on a 23-year-old woman with a right adrenal tumor 13 cm in diameter who was treated by laparoscopy. The patient was asymptomatic, and the tumor was incidentally diagnosed on abdominal ultrasonography. A subsequent computed tomography (CT) of the abdomen confirmed a 12x7x8-cm homogenous mass of the right adrenal. Magnetic resonance imaging (MRI) showed a solid mass measuring 13x7x7.5 cm arising from the right adrenal. Laparoscopic complete excision of the mass was accomplished through a transabdominal lateral approach. The postoperative period was uneventful, and the patient was discharged on the second postoperative day. Histology was consistent with an adrenal ganglioneuroma. Two years later, there is no evidence of recurrence on abdominal CT scan.

**Key Words:** Ganglioneuroma, Laparoscopic adrenalectomy, Adrenal tumor.

## INTRODUCTION

Ganglioneuroma (GN) is a rare (0% to 6% of incidentalomas), differentiated, benign, slow-growing tumor that commonly arises from sympathetic ganglion cells and is composed of mature Schwann cells, ganglion cells, and nerve fibres.<sup>1,2</sup> The family of tumors originating from ganglion cells includes GN which is a benign lesion, ganglioneuroblastoma which is of intermediate differentiation, and neuroblastoma which is a highly malignant lesion. Neuroblastoma and ganglioneuroblastoma most often occur in infants and children, whereas GN tends to occur in adolescents and young adults.<sup>1-3</sup>

GNs may arise anywhere along the paravertebral sympathetic plexus and occasionally from the adrenal medulla. Their localization is the retroperitoneal (32% to 52%), mediastinal (39% to 43%), or cervical region (8% to 9%).<sup>4-6</sup> Definitive diagnosis is made on histological examination. When such tumors arise from the adrenal medulla, their assessment and management are similar to that of other adrenal tumors. GNs are often asymptomatic even if they are large. Abdominal pain or the palpation of an abdominal mass, or both of these, are among the most common presenting features. Occasionally, symptoms like hypertension, diarrhea, and virilization may develop as a result of mixed hormonally active tumors secreting catecholamines, vasointestinal peptide (VIP), androgens, or all of these.<sup>2,7-10</sup> The prognosis is usually excellent, and recurrence is rare after surgical resection.<sup>11</sup>

The standard treatment suggested for benign adrenal tumors is laparoscopic surgery. At present, relative contraindications to laparoscopic adrenalectomy are a definitive or presumed diagnosis of invasive adrenal cortical carcinoma or circumstances that make a minimally invasive approach technically difficult, such as large tumors.<sup>12</sup>

We present herein a case of a large, incidentally diagnosed adrenal ganglioneuroma that was removed completely by the laparoscopic approach.

## CASE REPORT

A 23-year-old woman was admitted to the endocrine ward of our hospital, for investigation and management of a

Third Department of Surgery, Athens General Hospital, "G. Gennimatas," Greece (Drs Zografos, Kothonidis, Ageli, Kopanakis, Papastratis).

Department of Endocrinology, Athens General Hospital, "G. Gennimatas," Greece (Drs Dimitriou, Kaltsas).

Department of Pathology, Athens General Hospital, "G. Gennimatas," Greece (Dr Papaliodi).

Department of Internal Medicine, Athens General Hospital, "G. Gennimatas," Greece (Dr Pagoni).

Address reprint requests to: G. N. Zografos, MD, 10 K. OURANI str., Athens, 15237, Greece. Telephone: +302106801360, E-mail: gnzografos@yahoo.com

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13-cm mass arising from the right adrenal. This mass was found incidentally following ultrasonography (US) of the abdomen while the patient was being investigated for oligomenorrhea.

On admission, she was in good health and asymptomatic. She had no past medical history of importance. Blood pressure was within the normal range. On physical examination, a mild discomfort at deep palpation of the right quadrant was found. Radiography of the thorax and an electrocardiogram were normal. Routine blood and urine tests as well as tumor markers showed no abnormality.

Endocrinological investigations were within the normal range (**Tables 1–3**).

Computed tomography (CT) of the abdomen showed a 12x7x8-cm homogenous mass of the right adrenal (**Figure 1**). Magnetic resonance imaging (MRI) showed a solid mass measuring 13x7x7.5 cm arising from the right adrenal. On T2-weighted MR images, the tumor was shown as a heterogeneous mass. After intravenous injection of gadolinium, the mass showed a progressive, heterogeneous, and delayed enhancement (**Figure 2**). Preoperative radiological differential diagnoses were those of a GN and

**Table 1.**  
General Endocrinology Data

	Normal Range	Case
Triiodothyronine (T3) (ng/mL)	0.58–1.59	1.00
Free thyroxine (FT4) (pmol/L)	9.01–19.05	15.3
Thyroid-stimulating hormone (TSH) ( $\mu$ IU/mL)	0.35–4.94	1.006
Anti-thyroid peroxidase autoantibody (Anti-TPO)	<12	2.1
Luteinizing Hormone (LH) (mUI/mL)	luteal phase 0.67–23.75	7
Follicle Stimulating Hormone (FSH) (mUI/mL)	luteal phase 1.11–13.99	7.23
Prolactin (PRL)	1.2–29.93	9.88
Adrenocorticotrophic Hormone (ACTH) (pg/mL)	9–52	32.2
17OPG (ng/mL)	0.66–4.95	1.4
Dehydroepiandrosterone Sulfate (DHEA-S) (ng/mL)	1950–5070	2614
D4-androsterone (ng/mL)	0.1–3.0	2.9
Testosterone (ng/mL)	0.1–0.8	1.1
Aldosterone (pmol/L)	decubitus 22.2–477 standing 83–985	456
Renin ( $\mu$ U/mL)	decubitus 5–47 standing 7–76	63.6
Parathyroid Hormone (PTH)	11–62	41.3
Urine* (mL/24h)	–	2310* 4800†
VMA* (mg/24h)	1.8–6.7	1.8
Metanephrine* (mg/24h)	<1	0.36
Adrenaline* ( $\mu$ g/24h)	1.7–22.4	6.4
Noradrenaline* ( $\mu$ g/24h)	12.1–85.5	38.6
Creatinine† (g/24h)	1–2	0.6
Ca/P† (g/24h)	0.11–0.32/0.41–1	0.11/0.4
K/Na† (mEq/24h)	25–125/40–220	120.48/264

\* August 12, 2004.

† August 19, 2004.

**Table 2.**  
Synacthen Test (August 12, 2004)

	Normal Range	0 min	30 min	60 min
Cortisol (nmol/L)	morning 138–690	486.2	1391	1636
Aldosterone (pmol/L)	decubitus 22.2–477 standing 83–985	456	537	772
Active Renin ( $\mu$ U/mL)	decubitus 5–47 standing 7–76	63.6	–	–
17OH-PG (ng/mL)	0.16–3.33	1.4	4.6	5.0
ACTH (pg/mL)	9–52	32.2	–	–

myelolipoma. No evidence was present of calcification or invasion of adjacent tissues or organs.

A laparoscopic approach was decided on, despite the large size of the tumor, because of the low risk of malignancy based on radiology findings.

#### Technical Aspects of the Procedure

We prefer the transperitoneal lateral decubitus approach, as the best for maximal exposure of the gland and major vessels. Using the Hasson technique, we introduced a 12-mm trocar, two 10-mm trocars, and one 5-mm trocar. Laparoscopic exploration of the abdominal cavity was normal, and the tumor was easily identified. The right triangular ligament and the retroperitoneal liver attachments were cauterized and divided to allow liver retraction. After dividing the retroperitoneum, the inferior vena cava (IVC) was identified. The inferior periadrenal fat was carefully dissected from the upper pole of the right kidney and the renal vein identified. The adrenal vein was subsequently identified, dissected, double-clipped, and divided. The inferior and superior adrenal vessels were cauterized or clipped. Ultrasonic scissors were not used. The specimen was extracted through an extension of the

incision done for the Hasson technique. Minimal blood loss occurred, and the patient was not transfused.

The procedure lasted 90 minutes. The surgical specimen was oval and elastic in consistency, measuring 13x9.5x6 cm and weighing 390 g. The patient was discharged on the second postoperative day with bowel function returned to normal and able to resume normal physical activity.

#### Histology Report

By gross inspection, a large encapsulated tumor mass of 13x9.5x6 cm, weighed 390 g. On the surface of the tumor, small foci of flattened adrenal tissue were recognized. By sectioning, the tumor had a solid homogenous appearance and was grayish-white to yellowish on the cut surface. Microscopically, it was composed mostly of spindle cells arranged in fascicles. Mature ganglion cells scattered or arranged in small clusters were also noted (**Figure 3**). The described features were diagnostic of ganglioneuroma. No additional treatment was administered for this benign tumor. Two years after the procedure, there is no evidence of recurrence on abdominal CT scan, and the cosmetic result is satisfactory.

#### DISCUSSION

Ganglioneuromas are rare tumors, and there is a relative paucity of information in the literature concerning their exact incidence.<sup>4,9,10,12–14</sup> This is clinically relevant as the incidence of all adrenal tumors is increasing due to the availability of sophisticated imaging techniques and expertise; approximately, 1% to 10% of abdominal CT scans report incidentally an adrenal tumor.<sup>3</sup> Based on several series, GN accounts from 0% to 6% of incidentalomas. It is encountered rather more frequently in the literature when comparing its incidence among other large tumors of

**Table 3.**

Overnight 1 mg Dexamethasone (DEX) Test (August 14, 2004)

	Normal Range	Case
Cortisol (nmol/L)	morning 138–690	<20
Aldosterone (pmol/L)	decubitus 22.2–477 standing 83–985	79.1
Active Renin ( $\mu$ U/mL)	decubitus 5–47 standing 7–76	31.2
ACTH (pg/mL)	9–52	<5

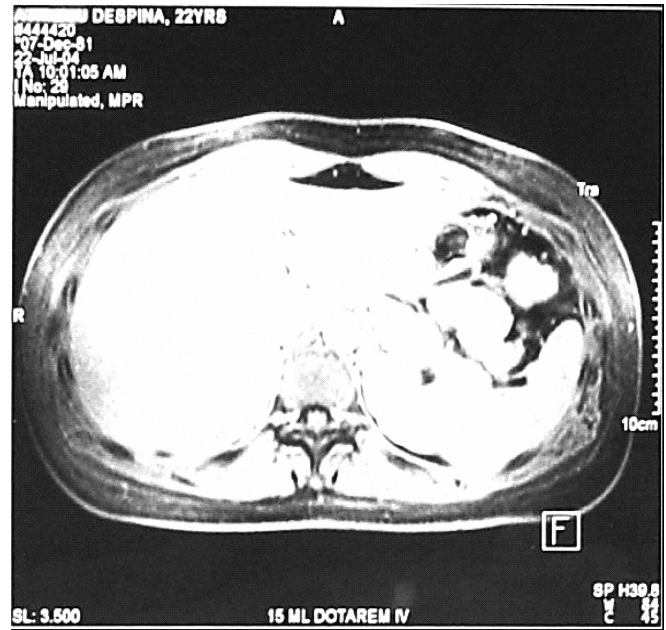


**Figure 1.** Abdominal computed tomographic scan indicating the large benign tumor.

greater than 6 cm in maximum diameter.<sup>3,11,13,14</sup> Approximately 100 cases of adrenal GN and 112 cases of extra adrenal GN have been reported in the Japanese literature.<sup>15</sup>

The imaging based differential diagnosis of an asymptomatic, nonfunctioning giant adrenal mass without evidence of systematic disease is broad and includes besides GN, neuroblastoma, ganglioneuroblastoma, myelolipoma and angiomyolipoma.<sup>8</sup> In the present case, the differential diagnoses suggested on MRI findings were benign tumor of the CNS (GN) and myelolipoma. Both CT scan and MRI are superior to US in detecting and characterizing an adrenal mass. Following the availability of studies comparing radiologically found and histologically proven dimensions of adrenal lesions, it was shown that CT scanning might underestimate the tumor size by 16% to 47%, whereas MRI does so by 20%. However, MRI appearances may correlate better with a histological diagnosis.<sup>11,16–20</sup>

Retroperitoneal GNs appear as well-defined masses that are oval, crescentic, or lobulated. They tend to surround major blood vessels; the result is absent or mild compromise of the lumen.<sup>2,21,22</sup> Intratumoral calcifications are present in 2.4% to 40% of cases; however, no calcifications were observed in our case. Areas of low attenuation that do not enhance are common, although such an enhance-



**Figure 2.** Magnetic resonance imaging. After intravenous injection of gadolinium, the mass showed a progressive, heterogeneous, and delayed enhancement.

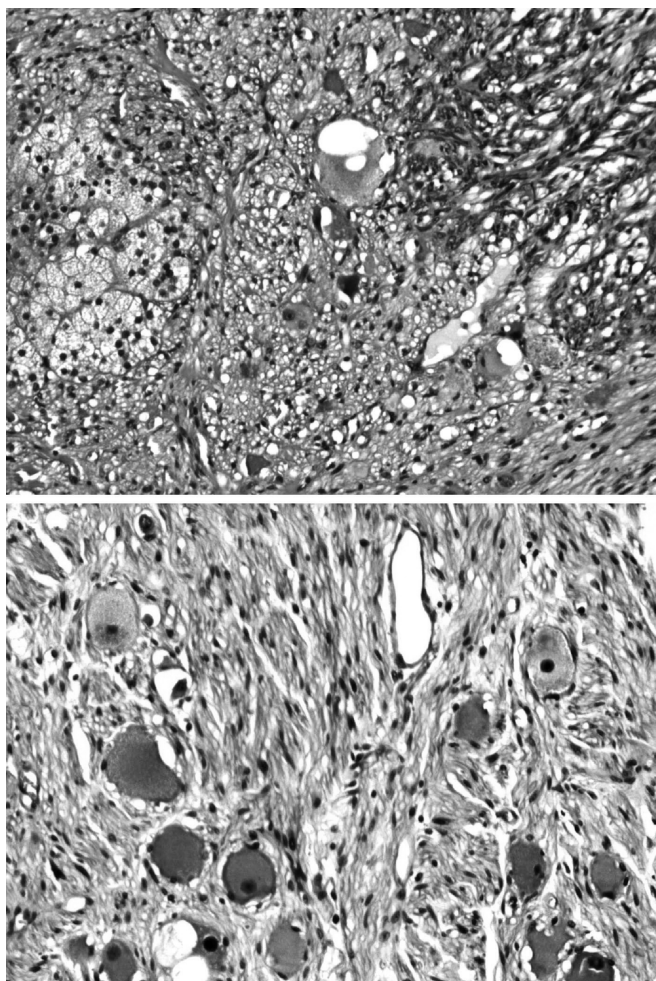
ment is usually of low intensity. An interesting pattern consists of delayed heterogeneous uptake of contrast media, which was observed on the MRI of the present case.<sup>7</sup>

Adrenal myelolipoma is a rare benign tumor composed of mature fat tissue associated with proliferating hematopoietic cells. CT features are characteristic if lipid content is demonstrated, which was not observed in our case.<sup>23,24</sup>

Hemorrhagic adrenal adenoma, adrenocortical carcinoma, metastatic tumors to the adrenal gland, and angiomyolipoma of the upper renal pole may also simulate GN or myelolipoma.<sup>25–29</sup> The other tumors of the CNS (neuroblastoma and ganglioneuroblastoma) develop at the same sites as the GN, but they occur in much younger patients and particularly in children. Such tumors are usually quite aggressive, having developed metastases in bone, liver, and lung in approximately 50% of cases. Intratumoral calcifications are higher in proportion and number.<sup>30,31</sup>

It has recently been demonstrated that scintigraphy and positron emission tomography (PET) can complement CT and MRI in diagnosing GNs.<sup>32,33</sup>

Laparoscopic adrenalectomy has become the gold standard in most patients with adrenal tumors.<sup>34</sup> Laparoscopic procedures are associated with less postoperative discom-



**Figure 3.** Top: Low magnification shows ganglioneuroma (right) with adjacent normal adrenal cortex (left) (H&E x10). Bottom: High magnification demonstrates spindle and mature ganglion cells (H&E x20).

fort, shorter hospital stay, less postoperative disability, and a lower rate of complications. There are few absolute contraindications for laparoscopic adrenalectomy, and most of them are not specific to adrenal surgery. Large but well encapsulated adrenal masses without evidence of local invasion can be removed laparoscopically.<sup>3</sup> The lateral transabdominal approach offers the best visualization of major vessels adjacent to the adrenals.<sup>35</sup> The possibility of malignancy should be appreciated preoperatively (using CT, MRI, or both) as well as intraoperatively. Invasion of periadrenal tissues or organs is not well suited to laparoscopic techniques and needs conversion to an open procedure.

Several authors limit the laparoscopic adrenalectomy to

lesions less than 6 cm in size,<sup>12</sup> whereas others have performed laparoscopic adrenalectomy on tumors up to 13 cm in diameter without any significant morbidity.<sup>5,13</sup> Extensive experience in advanced laparoscopic techniques and open adrenal surgery are mandatory to manipulate and laparoscopically excise large tumors.

## CONCLUSION

In summary, adrenal ganglioneuroma is a rare benign adrenal tumor that needs careful evaluation and surgical treatment. Laparoscopic resection of large tumors is feasible but necessitates experience in advanced laparoscopic surgery.

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