Laparoscopic Approach in Large Rare Adrenal Ganglioneuroma

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INTRODUCTION

Adrenal ganglioneuroma is an extremely rare and benign tumor comprising Schwann cells and ganglion cells [1]. Adrenal ganglioneuroma is usually hormonally silent and tends to be discovered incidentally on imaging tests. There have only been 41 cases reported in the literature in English from 1961 to 2009, involving 22 women and 19 men, with a mean age of 44 years and with incidentaloma as the initial presentation [2]. As the incidence of the adrenal tumor increases with increasing size, laparoscopic adrenalectomy is gradually

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ABSTRACT

Introduction: Adrenal ganglioneuroma is an extremely rare and benign tumor comprising Schwann cells and ganglion cells. Adrenal ganglioneuroma is usually hormonally silent and tends to be discovered incidentally on imaging tests. Here, we present a case of a 39-year-old man with adrenal ganglioneuroma who underwent laparoscopic left adrenalectomy.

Case Presentation: A 39-year-old man was admitted to our department with left flank pain after he recovered from Covid-19. He had no symptoms of nausea, vomiting, headache, diarrhea, palpitation, or chest distress, and there was no lack of appetite or weight loss. He denied any medical conditions, such as hyperaldosteronism, hypercortisolism, and hypertension. On further examination with contrast-enhanced CT identified a hypo-isodense mass in the left adrenal gland, measuring 4.8 x 4.4 x 5.0 cm, and spotty dense calcification in the middle of the mass. All functional laboratory evolution showed negative including urinalysis and serum electrolyte. Because the non-functioning suprarenal tumor was presumed to be benign, a laparoscopic approach was planned for definitive extirpation of the left adrenal gland. CT triple-phase whole abdomen was done before the procedure which showed contrast-enhanced identified an isodense mass with 43 HU in the left adrenal gland, measuring 4.8 x 4.4 x 5.0 cm with absolute washout of 49.3% and relative washout of 30.7%. Laparoscopic left adrenalectomy was performed. The patient underwent successful laparoscopic extirpation of the left adrenal gland. The postoperative pathology revealed a 7.0 x 6.0 x 3.0 cm encapsulated tumor with a firm texture. Microscopically, the tumor showed a mixture of mature ganglion cells and fascicles of Schwann cells. Adrenocortical cells were found near the capsule.

Conclusions: Even though there have been some guidelines for the diagnosis of Adrenal ganglioneuroma, pre-operative misdiagnoses are still frequent. We recommend that complete operative resection should be considered once malignancy cannot be excluded by pre-operative evaluation. Laparoscopic adrenalectomy is a reasonable option at least for tumors ≤ 4.5 cm. Adrenal ganglioneuroma can be successfully excised laparoscopically with an appropriate and experienced surgeon.
getting popular in the management of adrenal lesions. We assess the feasibility of laparoscopic resection for adrenal neoplasms more than 4 cm in size through the present study. Here, we present a case of a 39-year-old man with adrenal ganglioneuroma who underwent laparoscopic left adrenalectomy.

CASE PRESENTATION

A 39-year-old man was admitted to our department with left flank pain after he recovered from Covid-19. He had no symptoms of headache, nausea, vomiting, diarrhea, blurred vision, palpitation, or chest distress, and there was no lack of appetite or weight loss. He denied any medical conditions, such as hyperaldosteronism, hypercortisolism, and hypertension. The patient used no medications and no known allergies and denied tobacco, alcohol, or drug use. On physical examination, the patient was afebrile with normal vital signs. His weight was 70 kg, and his height was 1.67 meters. He appeared well hydrated with moist mucous membranes. He had an unremarkable exam — no findings of abdominal striae, palpable mass, and any tenderness. Further examination with contrast-enhanced CT identified a hypo-isodense mass in the left adrenal gland, measuring 4.8 x 4.4 x 5.0 cm, and spotty dense calcification in the middle of the mass (Figure 1).

All functional laboratory evolution showed negative urinalysis and serum electrolytes (sodium, potassium, chloride, and calcium). Because the non-functioning suprarenal tumor was presumed to be benign, a laparoscopic approach was planned for definitive extirpation of the left adrenal gland. While preparing and discussing with the family, the patient underwent routine tests (ultrasound examination and serum electrolyte test) every 3 months with no size progression of the tumor and normal range serum electrolyte. 10 months later, CT triple-phase whole abdomen was done before the procedure which showed contrast-enhanced identified an isodense mass with 43 HU in the left adrenal gland, measuring 4.8 x 4.4 x 5.0 cm with an absolute washout of 49.3% and relative washout of 30.7%. Laparoscopic left adrenalectomy was performed (Figure 2) [3].

During the procedure, the patient was placed in a lateral decubitus position. The patient was placed on a bean bag that helped support the patient in the required position. A soft roll was placed under the contralateral axilla. The arms were secured with padding. The contralateral arm was generally positioned on an arm board, bolstered with pillows or soft padding, and secured with tape. The ipsilateral arm was similarly secured on top of the contralateral arm but could also be supported by a metal L-shaped support that was secured to the table. The patient was securely fastened to the table with a two-inch tape over the lower leg, thigh, pelvis, and chest. Also, the potential of open conversion should be considered during positioning.

Laparoscopic adrenalectomy was performed using a trans-peritoneal approach. We used three 5-mm working ports and one 12-mm camera port for left-sided procedures. The first 12-mm port was inserted in the umbilicus or at the lateral border of the rectus abdominis muscle just above the level of the umbilicus to accommodate the camera. Two subcostal 5-mm ports were placed, one in the midclavicular line and the other in the lateral border of the rectus abdominis muscle. The third 5-mm subcostal trocar was inserted in the anterior axillary line.

The patient underwent a successful laparoscopic extirpation of the left adrenal. No adhesions were found with surrounding organs, there was no complication intraoperatively, and there was 300 ml blood loss. The patient was recovering well after surgery and was discharged from the hospital on postoperative day 2.

Figure 1. The contrast-enhanced CT identified a hypo-isodense mass in the left adrenal. (A), (B), (C) Arterial phase, and (D), (E) venous phase.
The postoperative pathology examination revealed a 7.0 x 6.0 x 3.0 cm encapsulated tumor with a firm texture. Microscopically, the tumor showed an admixture of mature ganglion cells and fascicles of Schwann cells (Figure 3A-B). Adrenocortical cells were found near the capsule. Immunohistochemical (IHC) examination showed immunoreactivity for synaptophysin, S-100, neuron-specific enolase (NSE), and vimentin (Figure 3C-F). AE-1/3 was positive in the remnant of adrenocortical cells (Figure 3G). Ki67 showed a low proliferative index (Figure 3H).

The tumor consisted of mature ganglion cells and Schwann cells (Hematoxylin-Eosin 100x) C. Synaptophysin positivity in ganglion and Schwann cells (IHC 100X) D. S100 positivity in ganglion and Schwann cells (IHC 400X) E. NSE positivity in ganglion and Schwann cells (IHC 400X) F. Vimentin positivity in stromal cells (IHC 100x) G. AE-1/AE-3 positivity in the remnant of adrenocortical cells (IHC 100X) H. Ki67 showed low proliferation index (IHC 100x).

**DISCUSSION**

Adrenal carcinoma is rare, in patients who have no history of malignancy. The size seems to be a strong predictor of malignancy, and a lesion smaller than 4.5 cm is considered at low risk of malignancy. Ganglioneuroma is a benign neoplasm that arises from neural crest cells of the sympathetic ganglia or adrenal medulla. It represents the most well-differentiated tumor in the neuroblastomaganglioneuroblastomaganglioneuroma spectrum and may be primary or different from neuroblastoma or ganglioneuroblastoma [4]. The main

### Table 1. The main characteristic of adrenal ganglioneuroma, adopted from Nieman (2010) [5].

<table>
<thead>
<tr>
<th>Functioning (&gt;15%)</th>
<th>Nonfunctioning</th>
</tr>
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<tbody>
<tr>
<td>Adenoma (aldesterone or cortisol)</td>
<td>Adenoma</td>
</tr>
<tr>
<td>Carcinoma</td>
<td>Myelolipoma</td>
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<tr>
<td>Pheochromocytoma</td>
<td>Neuroblastoma/ganglioneuroma</td>
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<tr>
<td>Congenital adrenal hyperplasia*</td>
<td>Hemangioma</td>
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<tr>
<td>Macronodular or micronodular adrenal disease*</td>
<td>Metastases*</td>
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<tr>
<td></td>
<td>Cyst</td>
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<td>Bleeding*</td>
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<td>Granuloma*</td>
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<td>Amyloidosis*</td>
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<td>Infiltrate disease*</td>
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*Bilateral involvement
CONCLUSIONS

Even though there have been some guidelines for the diagnosis of Adrenal Ganglioneuroma, pre-operative misdiagnoses are still frequent. We recommend that complete operative resection should be considered once malignancy cannot be excluded by pre-operative evaluation. Laparoscopic adrenalectomy is a reasonable option at least for tumors < or = 4.5 cm. Adrenal Ganglioneuroma can be successfully excised laparoscopically with an appropriate and experienced surgeon.

DECLARATIONS

Competing interest

The authors declare no competing interest in this study.

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REFERENCES