Left Adrenal Ganglioneuroma Treated by Laparoscopic Adrenalectomy in a 41-Year-Old Woman: A Case Report

Patient: Female, 41-year-old
Final Diagnosis: Adrenal ganglioneuroma
Symptoms: Asymptomatic • incidental finding
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Rare disease
Background: Ganglioneuromas are differentiated tumors originating from the neural crest. Although their occurrence is rare, they usually involve the posterior mediastinum and retroperitoneum. However, they rarely occur in the adrenal gland. Adrenal ganglioneuromas (AGNs) are hormonally inactive tumors that are mostly discovered incidentally during abdominal imaging performed for unrelated reasons. As preoperative diagnosis is challenging owing to their heterogeneous nature, adrenalectomy is the most effective method to ascertain an AGN diagnosis. We report a case of left adrenal ganglioneuroma treated by laparoscopic adrenalectomy. In addition, we have presented a relevant literature review to provide further information about this rare tumor.

Case Report: A 41-year-old woman presented to the Emergency Department with left flank pain associated with dysuria. She was diagnosed with renal colic, which was confirmed by computed tomography of the kidneys, ureter, and bladder. Additionally, an incidental solid lesion in the left adrenal gland was discovered. She was treated conservatively for her acute condition at the Emergency Department and discharged in a good condition. Further work-up including magnetic resonance imaging revealed a left large triangular suprarenal mixed soft tissue mass. She underwent laparoscopic left adrenalectomy. The final histopathology showed an AGN.

Conclusions: We present a case of a large AGN in a patient with systemic lupus erythematosus. Because it is a rare tumor with a heterogeneous presentation, its preoperative diagnosis is challenging. Thus, adrenalectomy is required to confirm the diagnosis. The prognosis is excellent and recurrence is extremely rare after tumor resection.

Keywords: Adrenalectomy • Ganglioneuroma • Laparoscopy

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Background

Ganglioneuromas (GNs) are rare, differentiated tumors originating from the neural crest. The most common site of GN occurrence is the posterior mediastinum (41.5%), followed by the retroperitoneum (37.5%). Rarely, they occur in the adrenal gland (20%) and neck (8%). GN occurrence in the intestine, spermatic cord, bone, and heart has also been reported in rare cases [1,2].

Adrenal GNs (AGNs) are rare benign tumors located in the adrenal medulla. They account for 0.3-2% of all adrenal masses and have an excellent prognosis. AGNs comprise mature Schwann cells, ganglion cells, and nerve fibers [2]. Considering the rarity of AGNs, their pathogenesis is not fully understood [3]. Moreover, their heterogeneous presentation makes the preoperative differential diagnosis of AGNs challenging [1-4].

Most patients with AGNs are asymptomatic, and these tumors are mostly discovered incidentally during abdominal imaging performed for other unrelated reasons [3,4]. Importantly, no specific imaging features are considered as pathognomonic for AGNs [3,4].

Given the difficulty of making a preoperative diagnosis of AGN and the possibility of malignancy, particularly if the tumor size is larger than 6 cm, all reports recommend adrenalectomy as the appropriate treatment for AGNs [1-4]. Furthermore, most studies do not recommend wide excision because AGNs rarely recur or metastasize [1-3].

This report presents the case of a 41-year-old woman with left adrenal ganglioneuroma who was treated by laparoscopic adrenalectomy.

Case Report

A 41-year-old woman with systemic lupus erythematosus on hydroxychloroquine visited our Emergency Department with concerns of pain on the left flank that persisted for 6 h; this was associated with dysuria. No other symptoms were noted. The patient had no family history of the same illness. On examination, the patient was alert, oriented, and experienced pain. Her vital signs were normal. The abdomen was soft and lax, with tenderness on the left flank, but organomegaly or lymphadenopathy was not observed. Laboratory findings showed a normal complete blood count, except for a low platelet count (87 000/µL; normal range: 150 000-400 000/µL). Laboratory tests of serum electrolytes and renal function showed the following findings: Na, 139 (136-145 mmol/L); K, 3.8 (3.5-5.1 mmol/L); urea, 1.86 (3.2-8.2 mmol/L); and creatinine, 48 (49-90 mmol/L). Computed tomography (CT) of the kidneys, ureter, and bladder showed left nephrolithiasis, and a solid lesion (7.0×4.0×4.8 cm) was incidentally detected in the left adrenal gland (Figure 1). Chest CT findings were normal.

The patient was diagnosed with renal colic and was treated conservatively with pain killer in the Emergency Department and was discharged in good condition. She was also referred to an endocrine surgery clinic. Further work-up for adrenal incidentaloma was obtained. Laboratory test results showed the following findings: 24-h urine vanillylmandelic acid, 12.6 (0-34.3 µmol/24 h); urine metanephrine, 319 (<1622 nmol/24 h); urine normetanephrine, 909 (<2129 nmol/24 h); total urine metanephrine, 1228 (<3751 nmol/24 h); serum aldosterone, 131 (<640 pmol/L); and urinary free cortisol, 65 (<486 nmol/24 h).

Figure 1. Axial abdominal computed tomography scan showing a large triangular (7.0×4.0×4.8 cm) complex mass of solid and relatively cystic components in the left suprarenal region.

Figure 2. Magnetic resonance imaging in axial T2-weighted sequence showing a large triangular mass of mixed low and high T2 signal intensity (solid and cystic components) replacing the left adrenal gland.
Additionally, magnetic resonance imaging revealed the presence of a large triangular left suprarenal mixed soft tissue mass (47×86×41 mm) (Figure 2). Furthermore, tiny peripheral nodular enhancement of the lateral wall of the mass was noted in the contrast series. Based on these findings, surgical intervention was planned. The patient was placed in the right lateral decubitus position during the surgery. Four ports (one 11-mm and three 5-mm ports) were used, and laparoscopic left adrenalectomy was successfully performed. The patient’s postoperative course was uneventful, and she was observed on a follow-up in the clinic 6 months after surgery, with no active concerns.

The resected mass measured approximately 10×10 cm (Figure 3). Gross appearance exhibited a 2.5×1×0.5-cm adrenal gland attached to a firm, well-circumscribed encapsulated, gray-white lesion (8.5×6×4 cm). Microscopic examination also revealed adrenal gland tissue with attached well-circumscribed benign proliferating neural crest tissue neoplasm, mostly composed of an admixture of mature-looking ganglion cells and Schwann cells (Figure 4). There was no evidence of malignancy. These findings are consistent with those of AGN. Written and verbal informed consent was obtained from the patient for the publication of this case report and accompanying images.

Discussion

AGNs are extremely rare tumors that affect the adrenal medulla. This report describes a case of a large AGN that was discovered incidentally and required adrenalectomy. This approach is in line with those in previously published reports [1-4].

An AGN typically affects the right adrenal gland, which is in contrast to the present case [5]. Although the etiology of AGN is unknown, the occurrence of AGN may be familial. Genetic factors, including mutations in the tyrosine kinase receptor ERBB3, play a role in GN pathology [2]. It is most prevalent in the fourth and fifth decades, and both sexes are equally affected [2]. In the present case, there was no family history of AGN, and the patient’s age fell within the range mentioned in previous reports.

AGNs are hormonally silent and are usually discovered incidentally, as seen in our case. Abdominal and/or back pain is the most frequently reported symptom secondary to a mass...
effect [1-4]. In case of excess secretion of catecholamines or steroid hormones (indicative of a hormonally active disease), the patient may develop specific symptoms such as diarrhea, hypertension, weakness, and virilization [1,3,6], but these symptoms were absent in our patient.

Abdominal CT is a valuable imaging tool for detecting adrenal masses [1-3]. CT findings that are indicative of AGNs include the observation of a well-defined, encapsulated mass with a lobular and solid appearance and an iso- to hypo-attenuating lesion in contrast to muscle signals. Furthermore, AGN cases do not have involvement of major blood vessels. Punctuate or discrete calcifications are observed in 20-69% of cases [1-3,7]. On the contrary, magnetic resonance imaging features include a hypointense T1-weighted signal and a hyperintense heterogeneous T2-weighted signal [7]. However, none of these features can differentiate AGNs from malignant adrenal tumors [2,4], which is in line with our case findings.

Adrenal cortical adenoma, composite pheochromocytoma, adenocortical cancer, neuroblastoma, and ganglioneuroblastoma are among the preoperative differential diagnoses [5]. Neuroblastomas and ganglioneuroblastomas originate from ganglion cells and are more prevalent in younger patients and children [5]. Furthermore, they metastasize to the bones, lungs, and liver in 50% of patients and behave more aggressively. Pheochromocytomas are usually symptomatic and have a familial tendency. Additionally, a 24-h urine catecholamine level is typically high in patients with this tumor [5]. Overall, AGNs have an excellent prognosis and do not require adjuvant treatment [1-3].

Adrenalectomy may be performed as an open technique or laparoscopically. Laparoscopic techniques have many advantages, including fewer complications, shorter hospital stays, and less postoperative pain; therefore, they have become the criterion standard for treating patients with adrenal tumors [5,8]. In our case, a laparoscopic approach was performed and was uneventful. Our case is unique; in addition to the presence of an AGN, the patient had systemic lupus erythematosus. To the best of our knowledge, this is the first case report of such an incidental combination.

Conclusions

AGN is a rare benign neurogenic tumor that is difficult to diagnose preoperatively, especially in asymptomatic patients. Adrenalectomy is therefore required to confirm the diagnosis. The prognosis is favorable, and recurrence is extremely rare after resection. We believe that this case report presents both clinical and radiological features of this condition that enrich the literature on such a rare tumor. Further larger case series are needed to gain a better understanding of the pathogenesis and to resolve the debate regarding the preoperative diagnosis and management of AGNs.

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Declaration of Figures’ Authenticity

All figures submitted have been created by the authors who confirm that the images are original with no duplication and have not been previously published in whole or in part.

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