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RARE ADRENAL INCIDENTALOMA: GANGLIONEUROMA

H.S. Rekhi, Malkiat Singh, Sudesh Partap, Baljeet Kaur, Rajat Talesra*, Samrat Singh, Shubham Chhabra
Department of General and Minimum invasive Surgery, GMC & RH, Patiala

Corresponding Author : *Dr Rajat Talesra
Department of General and Minimum invasive Surgery, GMC & RH, Patiala
E-mail : talesrarajat@gmail.com

ABSTRACT

Adrenal ganglioneuromas (AGNs) are rare, benign, well-differentiated tumors of neural crest origin. They are usually hormonally non-functional and asymptomatic, frequently discovered incidentally during imaging for unrelated concerns. We present the case of a 19-year-old female who exhibited vague gastrointestinal and autonomic symptoms. Imaging revealed a right adrenal mass suggestive of malignancy. However, hormonal evaluation was unremarkable. Surgical excision was performed, and histopathology confirmed the diagnosis of adrenal ganglioneuroma. This case underscores the diagnostic challenge of such rare adrenal incidentalomas and highlights the importance of multimodal imaging and histopathology in guiding management.

INTRODUCTION

Adrenal incidentalomas are adrenal masses incidentally discovered during imaging for non-adrenal indications. With the widespread use of ultrasonography and cross-sectional imaging, their detection rate has significantly increased(1). Most incidentalomas are benign adrenocortical adenomas, but rare tumors such as ganglioneuromas must also be considered.

Adrenal ganglioneuromas (AGNs) are rare, benign neurogenic tumors derived from neural crest cells. They represent the most mature form in the spectrum of neuroblastic tumors, which includes neuroblastomas and ganglioneuroblastomas(2). AGNs account for about 20% of all ganglioneuromas and often occur in adolescents and young adults(3). These tumors are usually hormonally inactive and asymptomatic, often discovered incidentally or when large enough to cause mass effect.

Due to their rarity and non-specific presentation, AGNs pose a diagnostic challenge. Imaging can mimic malignant adrenal tumors, and definitive diagnosis is often made postoperatively via histopathology(4). This case report illustrates such a scenario.

CASE REPORT

A 19-year-old female presented to the Surgery OPD with complaints of episodic diarrhea and vomiting for the past three months. She also reported dull aching pain in the right epigastrium and right flank for four weeks. Additionally, she experienced episodes of tachycardia, palpitations, chronic sweating, perioral pigmentation, and intermittent headaches.

On general examination, perioral pigmentation was noted. Abdominal and systemic examinations were within normal limits.

Ultrasonography of the abdomen revealed a hypoechoic mass measuring 45 × 38 mm with calcifications in the right adrenal area, abutting the upper pole of the right kidney. Based on this, a functional workup was initiated, which revealed:

- Serum ACTH: 24.3 pg/mL (normal <46 pg/mL)
- Serum cortisol: 13.28 µg/dL (normal 6.72–22.6 µg/dL)
- 24-hour urinary VMA: 8.41 mg/24 hrs (normal 0.4–15.44 mg/24 hrs)

All values were within normal limits, suggesting a non-functional adrenal lesion.

A contrast-enhanced CT (CECT) of the abdomen showed a well-defined, heterogeneously enhancing soft tissue

lesion measuring 4.9 cm (AP) × 3.4 cm (T) × 4.2 cm (CC) in the right adrenal region, with an average attenuation of 35–40 HU. The lesion showed calcific foci, abutted the liver (segment VI) with loss of fat planes, indented the upper pole of the right kidney, and contacted the IVC and right crus of the diaphragm. These radiologic features raised suspicion of pheochromocytoma or adrenocortical carcinoma.

Given the inconsistency between the non-functional biochemical profile and radiological suspicion of malignancy, contrast-enhanced MRI (CEMRI) was done. It suggested a differential of adrenal ganglioneuroma or atypical adenoma.

Due to the tumor's size, imaging features, and associated symptoms, an open right adrenalectomy was performed via a posterolateral approach. The mass was excised completely and intact, with no evidence of local invasion.

Postoperatively, the patient had an uneventful recovery. Clear fluids were started the evening after surgery, followed by semisolids on the first postoperative day. She was discharged on the fourth day in satisfactory condition, and sutures were removed on postoperative day twelve.

Histopathological examination (H&E stain) revealed a well-encapsulated tumor composed of Schwannian spindle cells arranged in fascicles and parallel bundles, with scattered large polygonal ganglion cells possessing eosinophilic cytoplasm and prominent nucleoli. The stroma showed mixed inflammatory infiltrates and dystrophic calcification. Peripherally compressed normal adrenal tissue was identified. These findings confirmed the diagnosis of adrenal ganglioneuroma.

As the tumor was benign, no further imaging surveillance was pursued.

DISCUSSION

Adrenal ganglioneuromas are rare, benign tumors originating from sympathetic ganglia and are considered the most differentiated form of neuroblastic tumors(2). They are more commonly found in the posterior mediastinum and retroperitoneum, with adrenal involvement being uncommon(5).

AGNs usually occur in patients over the age of 10 and are more common in females(3). They are typically non-functional, although rare cases of hormonal secretion (catecholamines, VIP, or cortisol) have been reported(6). In our case, although the patient presented with symptoms resembling catecholamine excess, including palpitations, headaches, and sweating, the functional workup was normal.

Imaging plays a crucial role in the detection and preliminary evaluation of AGNs. On CT, they often appear as well-circumscribed, hypodense lesions with mild to moderate enhancement and may contain punctate or coarse calcifications(7). MRI provides better tissue characterization; AGNs generally show low signal intensity on T1 and high signal intensity on T2-weighted images due to their myxoid content(8).

However, imaging findings are often non-specific, and AGNs may mimic malignant lesions such as adrenocortical carcinoma or pheochromocytoma, as was suspected in our case. Hence, histopathological examination remains the gold standard for diagnosis. The presence of mature ganglion cells, spindle-shaped Schwann cells, and absence of mitotic figures or necrosis confirms the diagnosis(6).

Surgical excision is both diagnostic and therapeutic. Open adrenalectomy is preferred for large tumors or when malignancy cannot be ruled out, while laparoscopic adrenalectomy may be reserved for smaller, well-demarcated lesions(9). Complete resection typically results in an excellent prognosis, with rare recurrence or malignant transformation.

CONCLUSION

Adrenal ganglioneuroma is a rare, benign tumor that should be considered in the differential diagnosis of adrenal incidentalomas, especially in young patients with atypical symptoms. Although often asymptomatic, these tumors may present with non-specific gastrointestinal or autonomic features. Radiologic evaluation can raise suspicion but may not definitively distinguish AGNs from malignant tumors. Surgical excision remains the cornerstone of diagnosis and management, offering excellent outcomes when complete resection is achieved. Awareness and early recognition of this rare entity are essential to avoid unnecessary anxiety and overtreatment.

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