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# A Rare Adrenal Incidentaloma: Case Report of a Right Adrenal Ganglioneuroma

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**Abstract** Adrenal ganglioneuroma (AGN) is an extremely rare and benign tumor that originates from the neural crest tissue of the sympathetic nervous system. The majority of cases are detected incidentally. It presents a challenging preoperative diagnostic differential with other solid adrenal tumors. However, the assessment and management of AGNs are similar to other adrenal tumors. We present a case of a 40-year-old female referred to our institution for a right solid adrenal mass that was detected incidentally on an abdominal CT scan (4×5×5cm). The patient underwent laparoscopic surgical excision of the tumor. The histopathology examination showed areas of spindle cells and scattered mature ganglionic cells compatible with AGN.

***Keywords:*** *adrenal ganglioneuroma, imaging features, laparoscopy*

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## 1. Introduction

Adrenal Ganglioneuroma (AGN) is a rare and benign tumor that originates from primitive neural crest cells. Histologically, the tumor is composed of Schwann mesenchymal cells and gangliocytes. Commonly, AGNs are hormonally silent and as a result can be asymptomatic even when the lesion is of substantial size. It is difficult to make a precise diagnosis preoperatively. Definitive diagnosis is done by the histopathological examination of the specimen. The assessment and management of these tumors are similar to other adrenal tumors.

## 2. Presentation of Case

A 40-year-old female, with no previous medical history, consulted for right lumbago evolving for 2 months without any other associated signs. The physical examination was without particularity. An abdominopelvic CT showed a right adrenal incidentaloma measuring 4 × 5 × 5 cm, well limited with lobulated contours, hypodense heterogeneous enhanced after injection and containing central calcifications (Figure 1). Complete preoperative hormonal evaluation was carried out to evaluate functionality and the lesion was considered a non-secreting tumor.

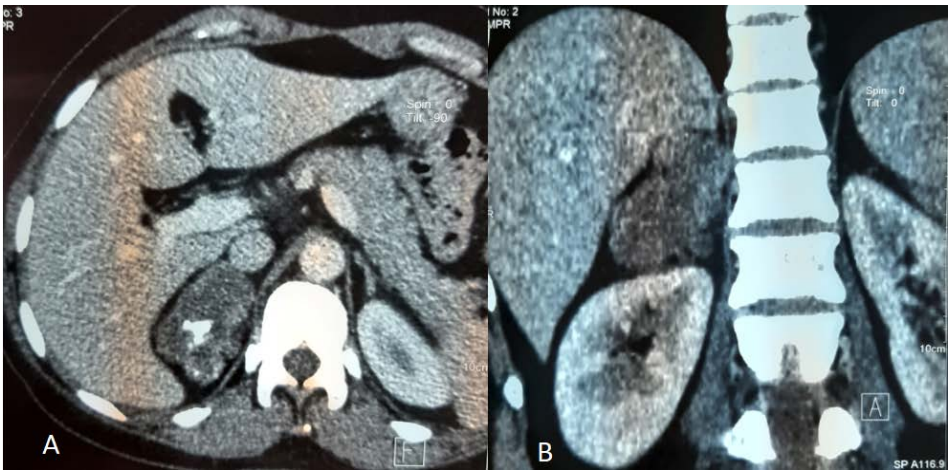


**Figure 1.** An abdominopelvic scan showed a right adrenal incidentaloma measuring 4 × 5 × 5 cm, well defined with lobulated contours, hypo-dense heterogeneous enhanced after injection with central calcifications: A) axial section; B) coronal section

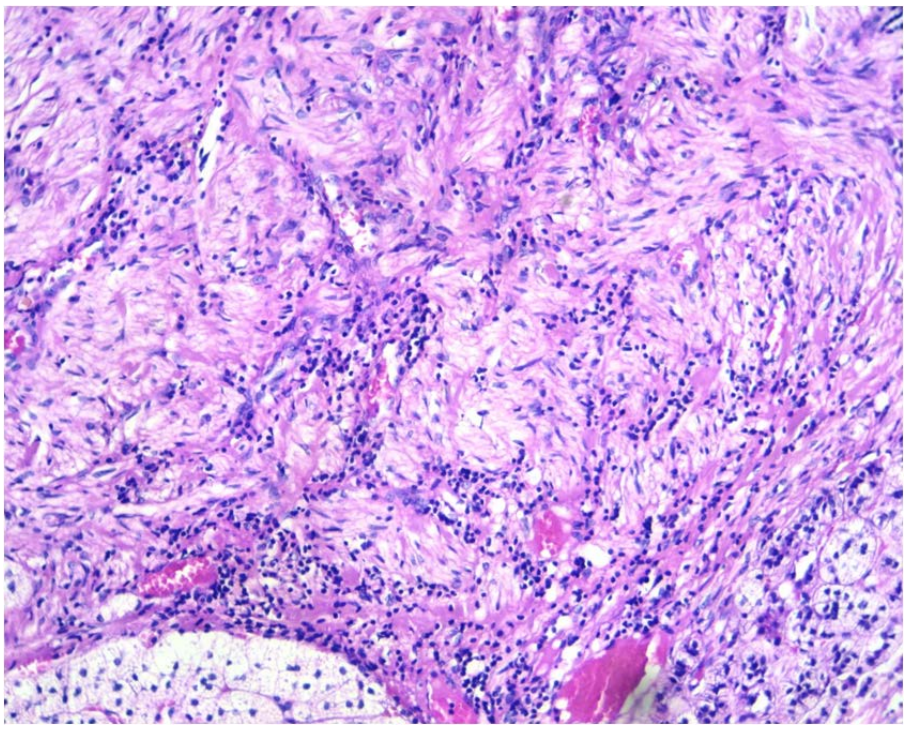

**Figure 2.** Tumor proliferation made of spindle cells without atypia; in contact with the adrenal fat. (HEx10)

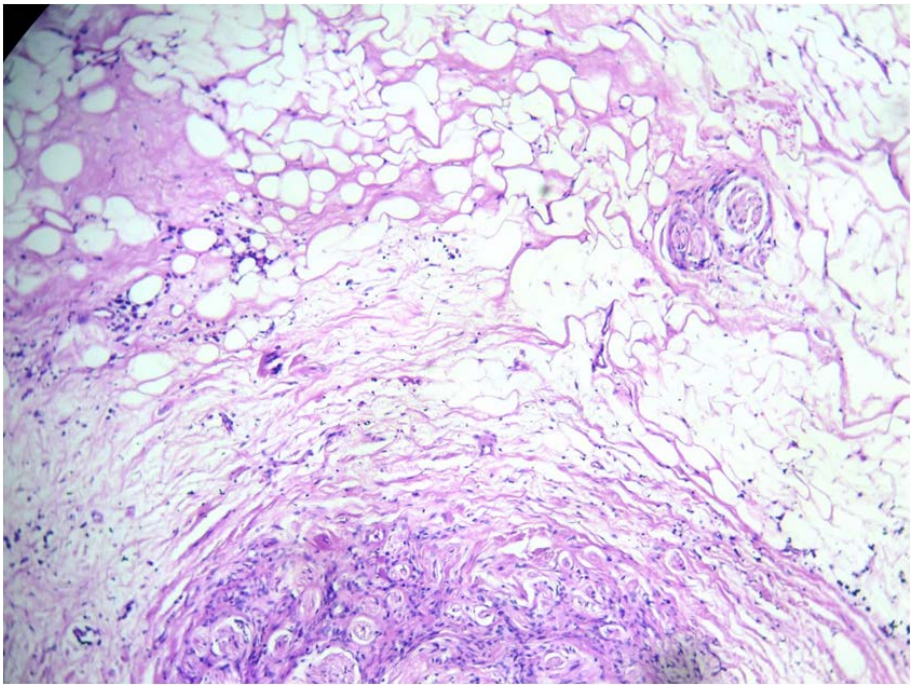

**Figure 3.** Spindle cells arranged in bundles containing layers of mature ganglion cells. (HEx40)

The patient underwent laparoscopic right adrenalectomy whose histopathological examinations confirm the diagnosis of AGN (Figure 2 and Figure 3). The postoperative period was uneventful.

## 3. Discussion

A GN is a rare differentiated benign tumor arising from primordial neural crest cells that form the sympathetic nervous system [1]. On histology studies, it is composed of mature Schwann cells and ganglion cells with a fibrous stroma [2,3]. GNs are a member of a group of neurogenic tumors which include ganglioblastomas and neuroblastomas. It differs from other neurogenic tumors in its benign nature [1]. GNs may arise in any region of the paravertebral sympathetic plexus [4,5], but are typically observed in the posterior mediastinum (39-43%) or the retroperitoneum (32-52%). In contrast, neuromas rarely occur in the adrenal glands [6,7]. Although GNs are generally considered to occur more frequently in young people, some recent studies have shown that they may also be seen in patients between the ages 40 and 50 [8,9].

Adrenal GNs are usually silent hormonally and as a result can be asymptomatic; even when the lesion is of substantial size [8,10]. On the other hand, it has been

reported that up to 30% of patients with GNs may have elevated plasma and urinary catecholamine levels, but without exhibiting any symptoms of catecholamine hypersecretion [2]. Additionally, it has been noted that ganglion cells can secrete vasoactive intestinal peptide (VIP), While pluripotent precursor cells sometimes produce steroid hormones, such as cortisol and testosterone [11,12].

Most GNs are discovered incidentally on imaging studies [7,13]. Ultrasonography shows a well-defined, homogeneous, hypoechogenic mass [14,15]. CT usually reveals a homogeneous or slightly heterogeneous mass that tends to surround major blood vessels without compression or occlusion. The tumor is well-defined, hypodense, and poorly enhanced by contrast medium [14,16]. Calcifications, typically fine and punctate, are seen in approximately 42-60% of GNs. Early enhancement of linear septae has been reported, and delayed heterogeneous contrast uptake has also been described in some cases. CT allows for an accurate description of the lesion and its relationship with vascular structures [17]. On magnetic resonance imaging, T1-weighted images tend to have homogeneously low or intermediate signal, whereas T2-weighted images have heterogeneously intermediate or high signal [18]. Arguably, the latter is caused by the presence of the myxoid matrix along with a relatively low number of ganglion cells [17]. Furthermore, gadolinium administration can result in delayed and progressive enhancement of the lesion [10,14]. Moreover, MIBG (131-metaiodobenzylguanidine) scintigraphy produces similar results in GNs, ganglioneuroblastomas and neuroblastomas [7,14]. Recently, PET scans have been proclaimed to facilitate the diagnostic process. Particularly, a Standardized Uptake Value (SUV) of 3.0 or higher has been suggested to distinguish malignant adrenal lesions from benign ones with a 100% sensitivity and 98% specificity [19]. However, Adas and al [20] did report an adrenal GN with a SUV of 4.1 that was determined to be histologically benign.

Preoperative diagnosis is difficult because of the lack of specific imaging findings [11,21]. Percutaneous biopsy may help, but the presence of undifferentiated components limits its utility [11,22]. The differential diagnosis of adrenal GN includes other adrenal solid masses such as pheochromocytoma, NB, GNB, adenoma, and carcinoma [2,11,22]. GNB and NB more frequently develop in children.

Ultimately, histopathological examination is required to confirm the diagnosis of GN. In the vast majority of cases, GNs are histologically benign lesions which can be classified into two main categories [2]. Firstly, “mature type” GNs comprise of mature Schwann cells, ganglion cells, and perineural cells within a fibrous stroma While completely lacking neuroblasts and mitotic figures [10]. Secondly, “maturing type” GNs consist of similar cellular populations with miscellaneous maturation degrees, ranging from fully mature cells to neuroblasts. Nevertheless, the detection of neuroblasts is typically indicative of neuroblastomas or ganglioneuroblastomas. These types of neurogenic tumors have the potential to evolve into GNs [14]. Characteristically, GNs exhibit immunohistochemical reactivity for specific markers such as S-100, vimentin, synaptophysin, and neuron-specific enolase [18].

Surgical management remains the gold standard for the treatment of primary adrenal GNs [2,23]. Surgical indications are similar to that of adrenal incidentalomas: if size exceeds 4-6 cm and/or suspicion of malignancy. AGNs are often non-secretory. Nonetheless, malignant GNs remain extremely rare occurrences [24]. Even though laparoscopic adrenalectomy is usually the procedure of choice, a number of variables (*e.g.*, hormonal activity, tumor location, and proximity to adjacent structures) also need to be taken into account when deciding on the best approach to manage these rare tumors [25]. Wide margin resections are usually unnecessary because adrenal GNs rarely metastasize or recur. Postoperatively, there is no need for adjuvant therapy in patients with AGNs and their prognosis is excellent [2,24].

## 4. Conclusion

AGNs are rare tumors and, despite some peculiarities used for radiologic diagnosis, preoperative diagnosis is difficult. Therefore, complete resection is recommended once malignancy cannot be excluded. The prognosis is excellent with surgical removal.

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