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A Rare Tumor in a Rare Location: Ganglioneuroma

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Abstract

Ganglioneuroma is a rare benign tumor Neuronagliomas of the adrenal gland are a rare pathology. Therefore, this case will be of interest to urologists, surgeons, oncologists, and pathologists. Located in the adrenal gland (20%), along the sympathetic chain, and particularly in the posterior mediastinum (40%) and the retroperitoneum (30%). Ganglioneuroma poses a positive diagnostic and therapeutic problem. We report the case of a patient aged 48 admitted for pain in the right hypochondrium. He had no significant past medical or surgical history. On a physical examination, there were no noticeable findings except for mild tenderness and a mass on palpation. He underwent abdominal CT, which showed a relatively homogenous right adrenal tumor measuring. Endocrine work-up including urine catecholamine and cortisol levels was normal. Due to the tumor size and with consideration of the differential diagnosis of a malignant lesion, we elected to operate on the patient. Right adrenalectomy was performed, with no related complications. The final histopathologic report revealed adrenal ganglioneuroma. Although benign, the ganglioneuroma can present malignant aspects, in particular, CT scans and biological that can mislead the clinician, so histology remains the examination of choice for making the diagnosis.

Keywords

Tumor, Adrenal Glands, Gangioneuroma

1. Introduction

Ganglioneuromas are benign and well-differentiated tumors of the sympathetic nervous system. They arise from the great sympathetic chains extending from the base of the skull to the neck, mediastinum, retroperitoneum, and adrenal glands [1] [2]. Ganglioneuromas of the adrenal gland are extremely rare. These tumors

affect preferentially young people with the majority reported to occurring before the age of 20. These tumors are usually asymptomatic and, in the majority of cases, detected incidentally. When symptomatic, they present with non-specific symptoms related to their size or location with compression of neighboring structures [3]. GNs are usually hormonally inactive, some of them secrete catecholamines. Symptoms like virilisation, hypertension, and diarrhea may develop as a result of mixed hormone secretion. The imaging characteristics of adrenal GN are variable and some are very similar to other adrenal tumors such as adrenocortical carcinoma (ACC) and pheochromocytoma, a fact that is crucial in the clinic [4] [5]. Therefore, it is generally challenging to obtain a precise differential diagnosis of adrenal GN prior to surgery. Histopathological examination is currently the only tool to definitely diagnose this pathology.

The objective of this work is to focus on this rare pathology in light of recent data from the literature.

2. Case Report

A 48-year-old male was admitted to our hospital for pain in the right hypochondrium. He had no significant past medical or surgical history. On a physical examination, there were no noticeable findings except for mild tenderness and a mass on palpation. The results of laboratory tests were all found to be within the normal ranges. Because of his symptoms, he underwent abdominal CT, which showed a relatively homogenous right adrenal tumor measuring 20 × 18 × 11 cm (**Figure 1**). Endocrine work-up including urine catecholamine and cortisol levels was normal. Due to the tumor size and with consideration of the differential diagnosis of a malignant lesion we elected to operate on the patient. Exploratory laparotomy was performed to allow a definite diagnosis. Right adrenalectomy was performed, with no related complications. The surgical specimen was an elastic tumor, measuring 20 × 18 × 11 cm (**Figure 2**). On microscopy, the section

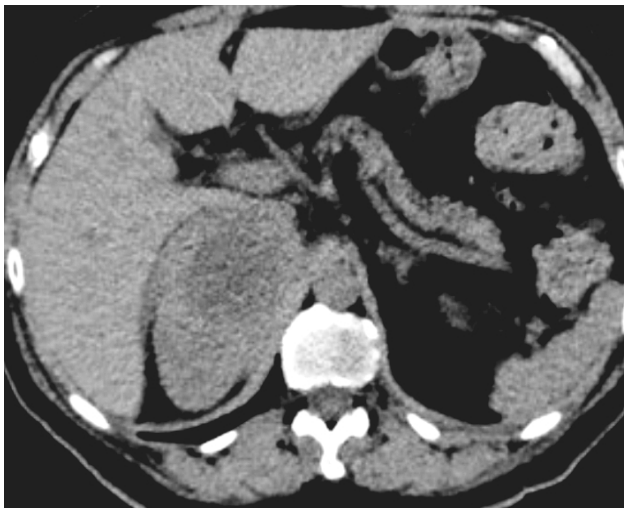

Figure 1. Abdominal CT showed a relatively homogenous right adrenal tumor measuring 20 × 18 × 11 cm.

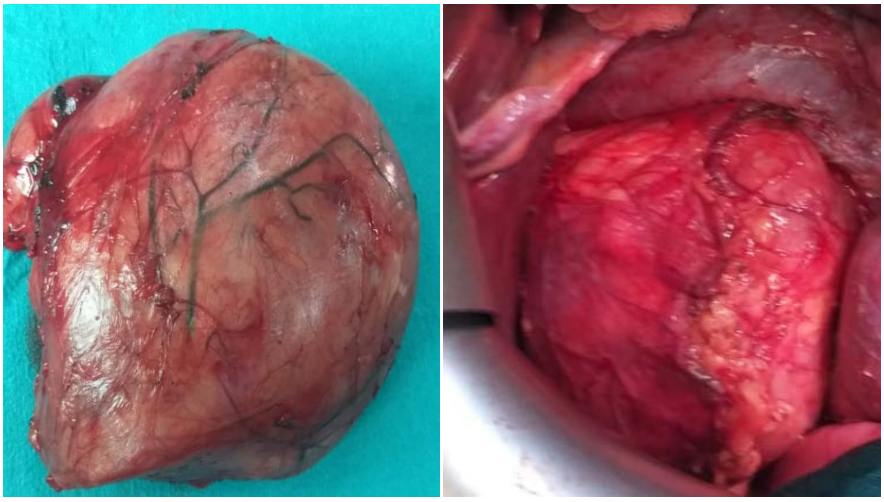

Figure 2. The surgical pictures of adrenal masse before and after resection.

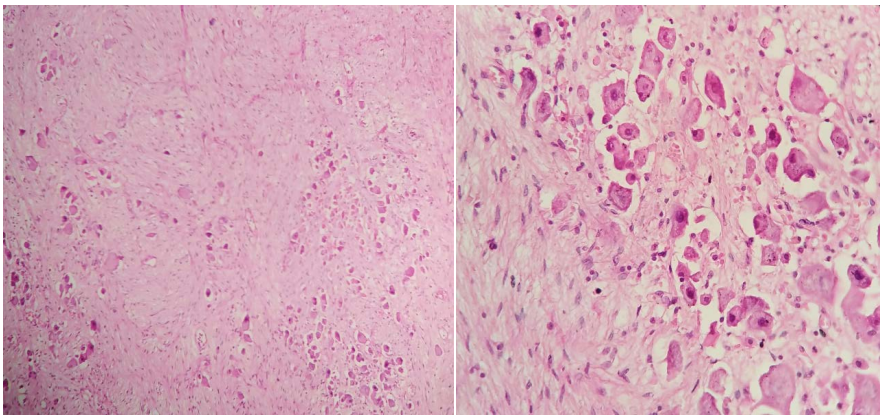

Figure 3. Tumor proliferation made on a schwanian background of mature ganglion cells.

showed irregular proliferation of spindle-shaped cells and scattered mature ganglionic cells with dystrophic changes and focal lymphocytic infiltration. No evidence was found for the malignancy. An immunohistochemical examination was requested for confirmation. The final histopathologic report revealed adrenal ganglioneuroma (**Figure 3**).

Postoperative recovery was uneventful. No recurrence was detected during the one-year follow-up visits.

3. Discussion

GN is a rare, differentiated and benign tumor arising from primordial neural crest cells that form the sympathetic nervous system [6]. On histology, it is composed of mature Schwann cells and ganglion cells with fibrous stroma [7] [8]. GN is a member of a group of neurogenic tumors group that includes ganglioblastoma and neuroblastoma. It differs from other neurogenic tumors in its benign potential [6].

Although GN is generally considered to occur more frequently in young people, some recent studies have shown that it may also be seen between the

ages of 40 and 50 [4] [9]. GN is most commonly found in the posterior mediastinum and retroperitoneum, and the involvement of the adrenal gland is relatively rare (21%) [10]. Adrenal GN is usually regarded as having silent hormonal functions and therefore can be asymptomatic. Occasionally, composite tumors with pheochromocytoma are seen and they, rarely, can secrete cortisol and androgen [11] [12].

Additionally, Geoerger *et al*. reported that up to 30% of patients with GN had elevated plasma and urinary catecholamine levels but that patients were rarely found to have symptoms of catecholamine excess [7]. By contrast, Koch *et al*. reported the case of a patient with a GN that was positive for vasoactive intestinal peptide, which is the product of ganglion cells [13]. Patients with vasoactive intestinal peptide-positive tumors such as GN and neuroblastomas may not have any symptoms or signs of vasoactive intestinal peptide secretion [14]. GN can also produce and secrete other hormones, such as testosterone, indicating the pluripotency of its precursor cell.

The differential diagnosis of an adrenal mass comprises a long list including adenoma, myelolipoma, cyst, lipoma, pheochromocytoma, adrenal cancer, metastatic cancer, hyperplasia and tuberculosis [7] [8].

The macroscopic characteristics of adrenal GN are an encapsulated mass with a firm consistency and a solid, homogeneous, grayish-white cut surface. Histopathological examination shows mature ganglion cells and Schwann cells among a fibrous stroma. Using microscopy they can be classified in two main groups: mature and maturing.

The mature type is composed of mature Schwann cells, ganglion cells and perineurial cells, whereas the maturing type consists of cells with different maturation levels, ranging from mature cells to neuroblasts with a similar stroma. According to immunohistochemical analysis, GN is characterized by reactivity for S-100, vimentin, synaptophysin and neuronal markers [12].

The imaging characteristic of adrenal GN on CT and MRI has been well described, but the precise diagnosis of adrenal GN using radiological evaluation prior to surgery is difficult. Qing *et al*. reported that the misdiagnosis rate of adrenal GN on CT and MRI before surgery is 64.7% [4]. Intratumoral calcification has been determined in 0% to 29% of the cases [4] [5].

PET scans can help complete the picture obtained by CT and MRI when making a differential diagnosis between adrenal GN and ACC or metastasis. One study reported that all cases of ACC had a SUV of 3.0 or higher, and that the sensitivity and specificity to distinguish ACC from adenoma were 100% and 98%, respectively [15] [16]. In another review of four patients with adrenal GN, the SUVs were between 1.5 and 2.9 [9].

Even though a lesion size greater than 4.5 cm seems to be a strong predictor of malignancy, this is not correctly confirmed by histologic evaluations. In our patient, although his tumor was larger than 18 cm, pathology confirmed a benign tumor. Recent studies recommended that nonsecretaryadrenal incidentalomas larger than 6 cm or with suspicious features of malignancy on imagining proce-

dures should be treated by adrenalectomy [6]-[17].

There is no medical treatment for such tumors. GN, although benign, can grow aggressively. Patients treated surgically for a benign neurogenic tumor have an excellent prognosis. Papavramidis *et al*. reported that adrenal GN should be resected by adrenalectomy, whereas retroperitoneal GN can be resected without adrenalectomy [18]. Laparoscopic adrenalectomy becomes the procedure of choice for the surgical removal of a vast majority of small-sized (<6 cm) adrenal lesions [6] [7] [17]. The prognosis for an adrenal GN following surgical resection is good.

4. Conclusion

Despite its rarity and benignity, the Adrenal ganglioneuroma is worth knowing. The main positive functional sign is pain without biological hypersecretion syndrome. His prognosis is extremely favorable after complete excision.

Consent

The authors have clear consent from the patient to patient to publish this article.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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