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Case Report

Adrenal Ganglioneuroma: Case Report and Literature Review

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Abstract

Introduction: Ganglioneuroma is a rare, benign neurogenic tumor that develops from sympathetic ganglionic cells. It is mainly located in the retroperitoneal region. ***Observation:*** This is a 39-year-old patient with no pathological history who has been seen for right back pain. Abdominal CT showed a right adrenal tumor with diameters of 40mm×22.5mm, a spontaneous density of >10 HU before injection, and > 31.5 HU after injection with a wash-out at late time < 40%. The hormonal balance reversed the secreting character of the mass, surgical treatment was indicated, histological analysis came back in favor of adrenal ganglioneuroma. ***Conclusion*:** Although benign, the ganglioneuroma may present malignant aspects, including scanographic that can mislead the clinician. Therefore, histology remains the examination of choice to make the diagnosis.

Keywords: Adrenalectomy, Ganglioneuroma, Laparoscopy.

INTRODUCTION

Ganglioneuromas are benign nerve tumours of children and young adults. They are rare [1], accounting for only 4–5% of adrenal tumors and less than 2% of adrenal incidentalomas [2-4].

They belong to the group of neurogenic tumors that develop at the expense of sympathetic ganglionic chains, a group that also includes ganglioneuroblastomas and neuroblastomas [5].

These tumors pose a problem of clinical and radiological diagnosis as well as a therapeutic problem because of their close anatomic connections with neighboring organs. Diagnostic confirmation is given by anatomopathological examination of the adrenalectomy piece [6].

We report the case of adrenal ganglioneuroma confirmed by histological examination.

OBSERVATION

We report the case of a 39-year-old woman with no particular pathological history, who was sent to us for the exploration of an adrenal mass discovered during an abdominal pelvic ultrasound requested for recurrent right back pain.

The clinical examination found a patient in preserved general condition, normotensive without clinical signs of hypercorticism or hyperandrogenicity.

Biological assays were found to be in favour of a non-secreting adrenal mass with correct kalemia, urinary methoxylates, 24-hour urinary cortisol, testosterone, and dihydroepiandrosterone sulfate (SDHEA) were normal.

Adrenal computed tomography (CT) showed an oval right adrenal mass, homogeneous hypodense, of regular contours, measuring 40mm× 22.5 mm of a spontaneous density >10 HU before injection, and > 31.5 HU after injection, discreetly enhanced at an early time, with late wash-out < 40% (Figure 1).

Given the symptomatic nature and the CT features, surgical treatment was indicated. The patient benefited from a right adrenalectomy performed by coelioscopic converted into laparotomy under costal due to the intimate adhesion with the liver, making the dissection hemorrhagic with simple operative follow-up.

The macroscopic analysis of the mass confirmed the well-limited character of the tumor of whitish color, fasciculate appearance, and firm

consistency; immunolabeling with the S100 protein showed diffuse positivity in fusiform cells and large lymph node cells (Figure 2). Facing these aspects, the diagnosis of adrenal ganglioneuroma was retained.

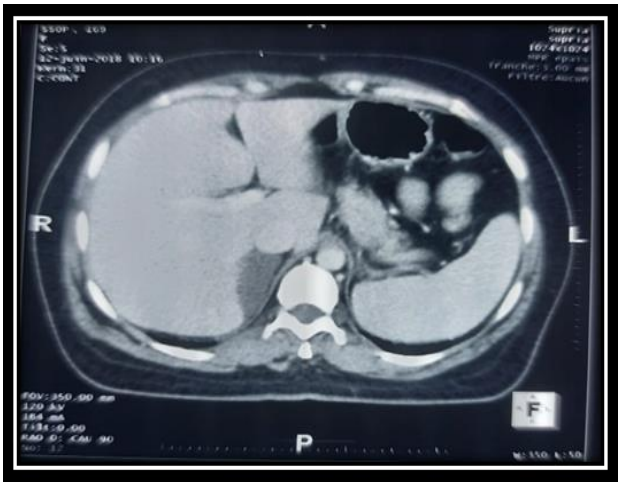

Figure 1: Axial Section: Abdominal CT with contrast injection showing solid right adrenal mass measuring 40mm×22.5 mm in diameters, oval, of regular contours, hypodense and homogeneous

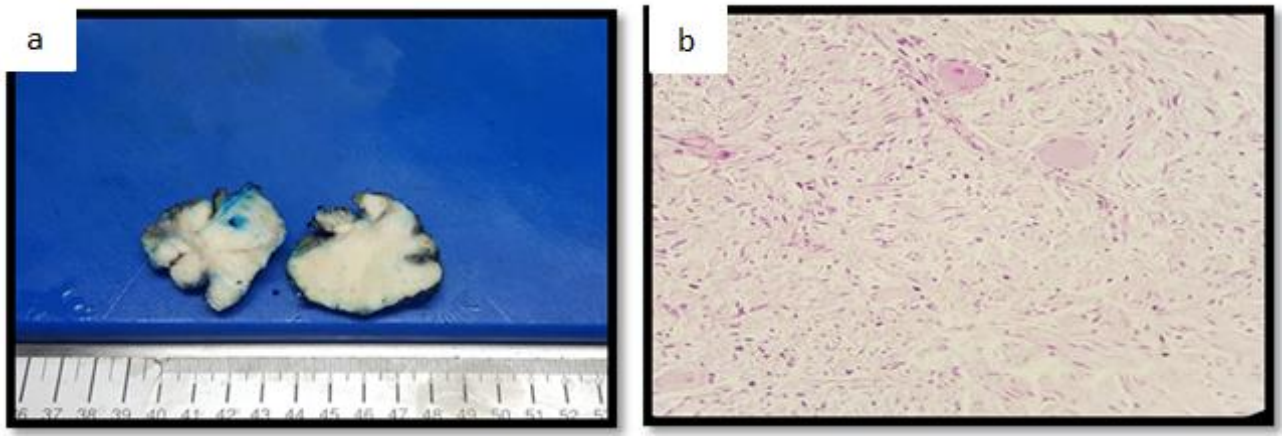

Figure 2: Macroscopic part (a): large, limited mass of whitish colour, fasciculate appearance and firm consistency; (b) Histological section: Immunolabeling by the S100 protein: diffuse positivity in both fusiform cells and large ganglionic cells

DISCUSSION

Ganglioneuroma is a rare, well-differentiated, benign neurogenic tumor consisting of mature sympathetic ganglionic cells and nerve fibers. It arises from neural crest cells like neuroblastomas and ganglioneuroblastomas [8].

This tumor is more common among children and young adults [9]. The median age at diagnosis is about 7 years, although it occurs at all ages [7, 10]. Female sex is most often affected with a sex ratio of 0.75 [11].

Retroperitoneal localization (37.5%) is the most common after mediastinal (41.5%), adrenal (21%) and cervical (8%) [7, 12]. Rarer locations include the spermatic cord, heart, and gastrointestinal tract [13].

In our case, the circumstances of discovery were abdominal pain, but the ganglioneuroma is often asymptomatic, discovered incidentally during a radiological assessment for another condition. It can sometimes manifest as abdominal mass or urinary, neurological, vascular, or digestive signs caused by compression of neighboring organs [13], which can even cause bowel obstruction [14].

Generally, adrenal ganglioneuromas are non-functional, with a normal hormonal assessment [15], as was the case with our patient. In rare cases, they may secrete cortisol and androgens [16], catecholamines, or VIP (intestinal polypeptide vasoactive), which cause diarrhea and high blood pressure [15, 17].

If biology is required to determine whether or not an incidentaloma secretes, morphological exploration is an effective tool for distinguishing benign from suspected malignant lesions and defining relationships with neighboring organs, particularly vessels [18]. A tumor size ≥4 cm, HU density ≥10 without injection, absolute washing <60%, relative washing < 40% 10 minutes after contrast injection,

invading surrounding tissue or metastases are signs of suspected malignancy. Calcifications are encountered in about half of the cases. These have a variable appearance, but are typically fine. Before injection, the tumor is homogeneous, hypodense, well limited and with regular contours. After injection, the contrast intake remains low to moderate. The mass becomes heterogeneous or remains homogeneous.

In some cases, magnetic resonance imaging (MRI) may be useful in assessing the invasion of surrounding vascular structures. It shows a tumor in homogeneous hypo-signal in T1 and hyper or iso-signal in T2 depending on the amount of stroma contained in the lesion. Contrast enhancement after gadolinium injection is not specific [15].

The ganglioneuroma still has a differential diagnosis with other retroperitoneal tumors, primarily ganglioneuroblastoma and neuroblastoma, but these tumors are suspected in the presence of radiological signs of locoregional invasion and the presence of an infiltrating character during surgery.

In front of a secreting adrenal incidentaloma, presenting radiological criteria suspect of malignancy or when it becomes functional or suspect during the follow-up period, the surgical indication arises [23]. In our case, given the impossibility of affirming or reversing the malignant character, a right adrenalectomy was performed, with simple operative follow-up. The diagnosis of adrenal ganglioneuroma was retained after a histological and immunohistochemical study with diffuse positivity in fusiform cells and large ganglionic cells by the S100 protein.

However, a complete analysis of the surgical part allows us to eliminate not only a neuroblastomatous contingent but also a pheochromocytoma within the ganglioneuroma [5].

Macroscopically, the tumor is well limited, lobulated, and sometimes encapsulated. The size is variable and often voluminous. When cut, it is smooth, firm, grey-white, and fasciculated. The presence of necrotic or hemorrhagic remodeling must make one suspect an immature contingent.

Histologically, the tumor consists of mature ganglionic cells with abundant eosinophilic cytoplasm and eccentric nuclei with a prominent nucleola. The background is of variable abundance, made of Schwann cells, sometimes arranged in bundles.

The evolution of ganglioneuromas is slowly progressive. Their prognosis is good in the case of complete excision. The complications are mainly mechanical [11].

In our patient, no local or remote recurrence was observed. Malignant transformation into ganglioneuroblastoma has been reported, but it is rare, which is why extended surveillance is important.

CONCLUSION

Adrenal ganglioneuroma is a rare benign tumor, non-secreting, developed from the sympathetic nervous system, which deserves to be known and evoked in front of an adrenal mass. Diagnosis is often late. Imaging, in particular CT and MRI, confirms the tumor's retroperitoneal seat, its relationship to neighboring organs, and predicts its resectability. However, ganglioneuroma may have malignant aspects that can mislead the clinician. Only histological study allows us to make the diagnosis. The prognosis is extremely favorable after surgery.

Conflict of interest: The authors declare no conflict of interest.

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