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Adrenal Ganglioneuroma

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Abstract **Case Report**

Introduction: Ganglioneuromas are benign, well differentiated tumors arising from the sympathetic nervous system. However, they arise rarely in the adrenal glands. Ganglioneuroma (GN) is a very rare (0–6% of incidentalomas) tumor that arises from sympathetic ganglion/chain and is made of mature ganglion cells, Schwann cells, neurites and nerve fibers. Most cases of adrenal ganglioneuromas are incidentally diagnosed since they are mostly asymptomatic and produce symptoms rarely due to compression of neighboring structures. ***Case Report:*** We hereby present a rare case of an adult patient followed by a benign colic sigmoid polypeptide with epigastralgia for which a pelvic abdomino CT scan was requested, demonstrating a left 56 mm adrenal mass. The patient was referred to the incidental alert service, with a non-secretory adrenal mass. The patient was operated with left adrenalectomy, with anatomo-pathological study a mature benign ganglioneuroma, of the very rare tumors reported in literature. ***Conclusion:*** The diagnostic approach to an adrenal incidentaloma is well codified, determining a precise etiologic orientation, since thorough endocrine exploration and radiological characteristics confirm the accurate diagnosis of certain tumors. Although in certain cases confirmation of diagnosis can only be made by histological examination, as is the case with this patient. Adrenal ganglioneuromas occur rarely and difficult in diagnosing preoperatively since symptoms vary and are mostly nonspecific. Ganglioneuroma should not be missed as a differential diagnosis of an adrenal mass. Although in certain cases confirmation of diagnosis can only be made by histological examination, as is the case with this patient.

Keywords: Ganglioneuroma, Adrenal, Adult, Laparoscopic excision, Histopathologic examination.

INTRODUCTION

Ganglioneuroma are benign tumors from neural crest cells, most often located in the posterior mediastinum and retroperitoneum, and are rarely localized in the adrenal gland. We report the case of a patient. Ho was referred to the incidental alert service.

The most common sites are the posterior mediastinum and the retroperitoneal space [1]. Most cases of adrenal ganglioneuromas are incidentally diagnosed since they are mostly asymptomatic and produce symptoms rarely due to compression of neighboring structures. Retroperitoneal ganglioneuromas are usually non-secreting and asymptomatic but when they reach large sizes they cause pressure symptoms locally [2]. We hereby present a rare case of an adult female patient presenting with pain in abdomen diagnosed with a non-secretory adrenal mass who underwent laparoscopic excision of the mass. She was finally diagnosed on pathological examination to be an adrenal ganglioneuroma, one of the very rare tumors reported in the medical world literature [3, 4].

CASE PRESENTATION

We report the case of a 45-year-old patient followed by a benign colic sigmoid polypeptide with epigastralgia for which a pelvic abdomino CT scan was requested, demonstrating a left 56 mm adrenal mass. The patient was referred to the incidental alert service. At the anamnesis, it does not report a paroxysmal acute attack, or weight gain, or signs of hypercatabolism. A physical examination did not reveal arterial hypertension or signs of hypercorticism, with no milk-spot stain. Routine laboratory investigations including the hormonal balance had shown normal results, including urinary methoxylates and urinary free cortisol, thyroid function tests were within normal range. The patient performed adrenal CT, specifying adrenal mass characteristics, which was hypodense in spontaneous contrast (26UH) moderately elevated after injection of the contrast medium (45UH) with a Wash out at 89% (figure 1 et 2). The patient was operated with left adrenalectomy, with anatomo-pathological study a mature benign ganglioneuroma.

Postoperatively, the patient presented no signs of adrenal insufficiency, with an annual follow in search of neurofibroma.

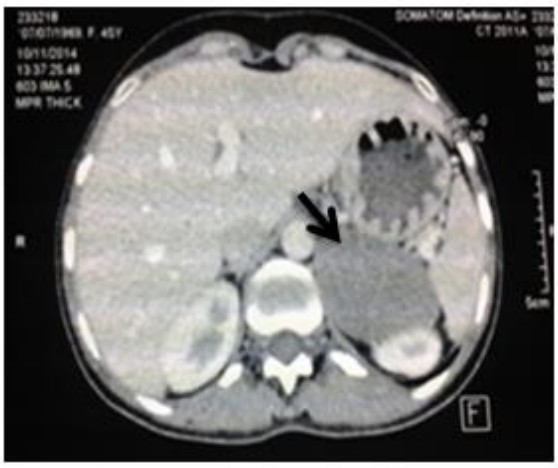

Fig-1: Adrenal scanner without injection

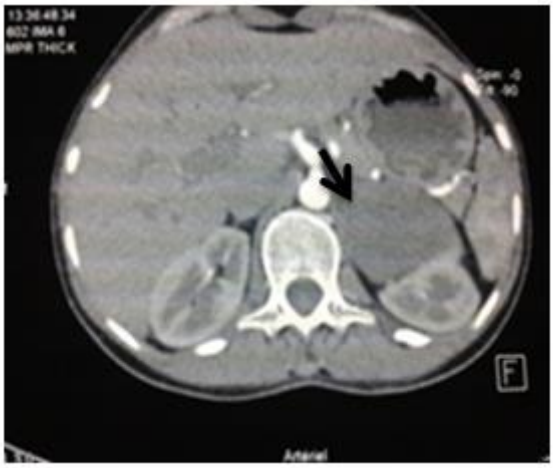

Fig-2: Adrenal scanner without injection

DISCUSSION

The incidence of adrenal ganglioneuroma is 1/ million population being most commonly sporadic in occurrence but could be associated with other syndromes like multiple endocrine neoplasia type 2/neurofibromatosis type 2. Clinically, the majority of these tumors is asymptomatic and hormonally non-secretory (incidentalomas). The patient described in this case presented the most common manifestation of adrenal ganglioneuroma, an incidentaloma, and the mass was diagnosed after the imaging study.

Adrenal incidentalomas are uncommon in patients younger than 30 years of age, but increase in frequency with age. It affects men and women equally. In more than 85% of cases, lesions are benign nonfunctioning adenomas.

There have only been 41 cases reported in the literature in English from 1961 to 2009, 22 involving women and 19 involving men, with a mean age of 44 years and with incidentaloma as the initial presentation.

Functioning tumors and carcinomas are not generally incidentalomas, since their diagnosis is based on specific signs and symptoms. However, in some cases, secretion of catecholamines, vasoactive polypeptides and androgens have been described (in association with pheochromocytomas) [5]. Compressive symptoms may occur. The prognosis is very good with surgical removal [6].

Adrenal carcinoma is rare, especially in patients who have no history of malignancy.The size seems to be a strong predictor of malignancy, and a lesion smaller than 4.5 cm is considered at low risk for malignancy. These patients can be followed up with annual abdominal CT and hormonal tests. However, there is no consensus on how to follow up non-surgical patients. For masses that seem to be benign in the imaging study (< 10 Hounsfield Units – HU; contrast washout >50%), small (< 3 cm) and nonfunctioning,

biochemical and imaging revaluation can be done every 1 to 2 years, and later assessment if clinical changes.

The fish flesh appearance of the tumor raised the hypothesis of an injury of neural origin, which was confirmed by histopathology. Ganglioneuroma (GN) is a benign neoplasm, arises from the neural crest cells - the sympathetic ganglia and the adrenals. It is composed of Schwann cells, ganglion cells and fibrous tissues. The differential diagnosis of a tumor with high resemblance to ganglioneuroma is a neuroblastoma. In neuroblastoma, levels of urinary homovanillic acid (HVA) and vanillylmandelic acid (VMA) are usually raised, while the levels of urinary HVA and VMA in ganglioneuroma are within normal ranges [7]. Neuroblasts are not a part of mature GN. Thus the Meta iodo benzyl guanidine uptake of a GN is lower than that of a neuroblastoma.

The GN should be diagnosed when the following findings are noted in a case of an adrenal mass: (1) non secreting, (2) absent vessel involvement, (3) calcifications in the gland, and (4) magnetic resonance imaging (MRI) findings of a non-enhanced T1-weighted signal with late and gradual enhancement. The final treatment for adrenal ganglioneuromas is resection by either open or laparoscopic method [8-10].

Table-1: The main features of adrenal ganglioneuroma are summarized in (Table 1) [11]

Main characteristics of adrenal ganglioneuromas
Clinical: Incidentaloma /compressive symptoms may occur
Radiology: Homogeneous, encapsulated mass, with well-defined edges and without invasion of nearby structures/calcification (40%-60%)/intense signal on T2-bright
Laboratory: Nonfunctioning/Rare: secretion of catecholamines, vasoactive polypeptides and androgens
Microscopy : Mature ganglion cells and Schwann cells among a fibrous stroma

Although the appearance, including the presence of calcification, may overlap with that of primary adrenal carcinoma, the lack of both clinical symptoms and local invasion may help suggest the diagnosis of adrenal ganglioneuroma.

CONCLUSION

Ganglioneuroma is a benign tumor of the sympathetic nervous system that occurs in all age groups but is more common before the age of 60 years. Ganglioneuromas may arise anywhere along the paravertebral sympathetic plexus and occasionally are found within the adrenal medulla [2]. Rarely, neuroblastoma may mature into ganglioneuroma. Also, ganglioneuroma may occur in association with other tumors such as pheochromocytoma.

Adrenal ganglioneuromas do not typically secrete exogenous hormones; thus, systemic manifestation of disease would not be cxpected. The tumor is commonly identified serendipitously in patients undergoing radiologic study for other reasons. Thus,ganglioneuroma should not be missed as a differential diagnosis of an adrenal mass. Histopathologic examination plays a crucial role in diagnosis.

Conflits d'intérêts

Les auteurs ne déclarent aucun conflit d'intérêts

Contributions des auteurs

Tous les auteurs ont contribué à la conduite de ce travail. Tous les auteurs déclarent également avoir lu et approuvé la version finale du manuscrit.

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