Adrenal ganglioneuroma incidentally discovered in a patient with dysuria: a case report

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Abstract. – A case of adrenal ganglioneuroma incidentally discovered during an abdominal ultrasound examination in a 26 year-old woman patient with recurrent episodes of dysuria. After a diagnostic work-up with laboratory and abdomen CT scan, the patient underwent a laparotomic removal of the adrenal lesion. Histopathological examination of the adrenal mass confirmed the diagnosis.

Key Words: Adrenal incidentaloma, Dysuria, Adrenal ganglioneuroma, Urinary infections.

Introduction

Due the increasing use and improved technology of imaging studies, the prevalence of adrenal incidentalomas is increasing and many clinical questions arise1,2. Adrenal incidentalomas are characterized by size, growth, imaging characteristics and functional status. These criteria are used as surrogates to determine whether an adrenal tumor is likely to be malignant or if it should be resected based on its functional status3,4. Ganglioneuroma originates from cells of the ganglia and the adrenal medulla5. Adrenal ganglioneuroma are rare, often asymptomatic tumors that are usually hormonally inactive and incidentally discovered6,7.

Here we report a case of adrenal ganglioneuroma incidentally diagnosed in a 26 year-old woman patient with recurrent episodes of dysuria. Histopathological examination of the adrenal mass confirmed the diagnosis.

Case

A 26 year-old woman was admitted in our Department for evaluation of an incidentally discovered right adrenal mass. She had been in her usual state of health until the actual symptoms started 3 months ago, when she developed episodes of dysuria with fever (38.5°C). A urine culture showed growth of Escherichia coli, and ciprofloxacin was prescribed. Recurrent episodes of dysuria persistent and during investigation of this symptom, a heterogeneous, hypogenic right adrenal solid mass, measuring 60 × 40 mm, was incidentally detected on abdominal ultrasonography. Our patient was unmarried, and she is not sexually active. Her family history was remarkable for arterial hypertension. Physical examinations revealed a body temperature of 36.8°C, the patient’s blood pressure was 120/80 mmHg, and her pulse 72 beats per minute and regular; her waist circumference was 70 cm and body mass index was 23 kg/m². None signs of hypercortisolism (facial plethora, fullness, dorso-cervical and supraclavear fat pads, striae rubrae), and hyperandrogenism (hirsutism) were present.

Laboratory tests showed a normal complete blood count, normal levels of serum potassium (4.1 mEq/l; standard range 3.5-5 mEq/l), serum sodium (141 mEq/l; standard range 136-145 mEq/l) and creatinine (0.8 mg/dl; standard range 0.6-1.5 mg/dl). In the supine position, morning plasma aldosterone (10 ng/dl; standard range 0.75-15 ng/dl), plasma renin activity (0.4 mg/ml/h; standard range 0.2-2.7 ng/ml/h), plasma ACTH (26.8 pg/ml; standard range 10-60 pg/ml), Δ4 androstenedione (1.56 ng/ml; standard range 0.4-3.39) and DHEAS (134 mcg/dl; standard range 120-360) were normal.

Early morning (08:00 h) plasma cortisol was 1.5 µg/dl (standard value <5 µg/dl) after oral ad-
administration of 1 mg of dexametasone at 23:00 the previous night. Her 24-hours urinary excretion of aldosterone (17.6 µ/24h; standard range 2.8-25 µg/24h), cortisol (31 µg/24h; standard range 10-100 µg/24h), total metanephrines (120 µg/24h; standard range 20-345 µg/24h) were all normal.

Electrocardiogram (EKG) showed a sinus rhythm and chest roentgenogram was unremarkable.

Ambulatory blood pressure monitoring (ABPM) shows normal values of systolic and diastolic blood pressure (daily average 115/70 mmHg) with conserved the normal circadian rhythm.

Abdominal CT showed a well demarcated homogenous, hypodense, right solid adrenal mass (80 × 70 × 50 mm) without contrast enhancement (Figure 1). From these findings, the patients underwent a surgical exploration of the right adrenal mass because of a suspicious of malignancy, according to NIH statement of adrenal mass3. In particular, NIH recommended that non-functioning adrenal incidentalomas larger than 60 mm or with suspicious features of malignancy on imaging studies should be treated using adrenalectomy because of increased prevalence of malignancy.

Right adrenalectomy was performed at the Surgery Department by the transabdominal route under general anesthesia.

The macroscopic specimen showed that the right adrenal gland was occupied by a large tumor (100 × 80 × 60 mm), which compressed the covering adrenal cortex, and it was well encapsulated (Figure 2).

The cut surface of tumor was solid with whitish grey color without evidence of hemorrhage or necrosis. Microscopically the section showed that tumor was composed of a matrix of fibroblast with nests of Schwann cells and mature ganglia cells, the surrounding areas were composed by normal adrenal tissue. No invasion of the capsula neither of the vessels was present. This histological examination consists with diagnosis of adrenal ganglioneuroma (Figure 3).

After a period of 6 and 12 months of follow-up patient was doing well, laboratory data were normal and abdominal magnetic resonance
Figure 3. The picture shows the interface between adrenal tissue and ganglioneuroma, with fibrillar stroma and ganglion cells at low [A], and medium [B] power (A: 100×; B: 250×; H&E stain).

In particular, low alteration (<10 HU) on unenhanced CT scans or rapid attenuation washout on delayed contrast-enhanced CT images predict the presence of a lipid-rich benign adenoma with high accuracy\textsuperscript{12}. When the lesion has an alteration of major than 10 HU, further work-up seems to be necessary, since it is probably a lipid-poor lesion.

In more than 1000 cases evaluated by a multicenter study organized by the adrenal incidentalomas study group of Italian Endocrinology Society (AI-SIE) most of adrenal incidentalsomas were non-hypersecretory adenomas (74%), the prevalence of cortisol-secreting adenomas was 9.2%, pheochromocytoma was the second most prevalent form of hyperfunctioning tumor, occurring in 4.2% and adrenocortical carcinoma was about 4% of the overall series. Other tumors, such as adrenal ganglioneuromas were rare (1.5%)\textsuperscript{2}.

Ganglioneuroma is a benign tumor of the sympathetic nervous system, originating from the neural crest, from undifferentiated cells. It is more often found along the paravertebral sympathetic plexus, and occasionally from the adrenal medulla\textsuperscript{6,7}.

Approximately 100 cases of adrenal ganglioneuromas and 112 cases of extra-adrenal ganglioneuromas have been reported in the Japanese literature\textsuperscript{13}. Moreover, a review of pure adrenal ganglioneuroma studies in the English literature allowed us to identify 48 cases between 1961 and 2010\textsuperscript{14,35}, and as our report, the main presentation was adrenal incidentaloma. Because of this lack of hormonal production, adrenal ganglioneuromas are usually clinically silent lesions.
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detected in patients undergoing abdominal imaging studies for unrelated reasons. In the current case, the tumor was found incidentally following ultrasonography of the abdomen while the patient was being investigated for recurrent urinary infections.

Adrenal ganglioneuromas appear homogenous on enhanced CT imaging but have a peculiar contrast enhancement pattern that consists of delayed heterogeneous uptake, as was seen in our patient. The delayed gradual contrast enhancement in thought to be due to the presence of myxoid matrices in these tumors, resulting in delayed progressive accumulation of contrast material in the extracellular space\(^{15,16}\). However, only histopathological examination can confirm the diagnosis of ganglioneuroma and to distinguish it forms its malignant relatives\(^{16}\).

Laparoscopic adrenalectomy should be considered for incidentally discovered adrenal masses that are shown to be hormonally active or suspected to represent a malignant primary lesion\(^1,8\). Several Authors limit the laparoscopic adrenalectomy to lesions than 6 cm in size\(^2\), whereas others have performed laparoscopic adrenalectomy on tumors up to 13 cm in diameter without any significant morbidity\(^37\). In the present case, open right adrenalectomy was preferred, because laparoscopic surgery increases the risk of tumor recurrence. However, an analysis conducted by Hayes et al\(^38\) revealed that these adrenal ganglioneuromas the tumors are consistent with a prolonged disease-free survival even when excision in not possible. In rare cases, tumor recurrence may arise. Clinical and imaging follow-up for long periods are important measures.

In conclusion, in our case presented with recurrent urinary infections and incidentally adrenal mass the final diagnosis was a pure adrenal ganglioneuroma. Surgery consisted in a laparotomic adrenalectomy with complete excision of the tumor. The clinical and radiological follow-up upon 12 months revealed no recurrence of the mass.

References


