Adrenal Ganglioneuroma: Case Report

Adrenal Ganglionöroma: Olgu Sunumu

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Abstract

Ganglioneuroma (GN) is a rare benign tumor arising mainly from neural crest cells and consisting of Schwann and ganglion cells. GN rarely occurs from the adrenal medulla and is observed most commonly in children and young adults. It is not associated with hormonal activity and is clinically asymptomatic. We present a 35-year-old woman whose abdominal ultrasonography for abdominal pain revealed in the right adrenal gland, a biochemically normal mass that was minimally hypointense on T1-weighted and hyperintense on T2-weighted magnetic resonance images (MRI) and which was pathologically diagnosed as GN after adrenalectomy. *Turk Jem 2011; 15: 74-6*

Key words: Adrenal gland, ganglioneuroma, magnetic resonance imaging

Özet

Ganglionöroma (GN) nöral krest hücrelerinden kaynaklanan schwann ve ganglion hücrelerinden oluşan nadir görülen benign tümördür. GN adrenal medullada nadir olarak oluşur ve genellikle çocuklarda ve genç erişkinlerde görünür. Hormonal aktivite göstermez ve klinik olarak asemptomatiktir. Karın ağrısı şikayetiyle yapılan abdominal ultrasonografisinde sağ adrenalde kitle tesbit edilen, biyokimyasal olarak normal olan ve manyetik rezonans (MR) aksiyel T1 ağırlıklı serilerde minimal hipointens, T2 ağırlıklı serilerde hiperintens görünümde olan, adrenalektomi sonrası patolojik olarak ganglionöroma tanısı alan 35 yaşında bir kadın olguyu sunuyoruz. *Türk Jem 2011; 15: 74-6*

Anahtar kelimeler: Adrenal bez, ganglionöroma, manyetik rezonans

Introduction

Ganglioneuroma (GN) is a rare benign tumor arising mainly from neural crest cells. Histologically, it is composed of Schwann and ganglion cells. The tumor is mostly located along the length of the sympathetic chain. Retroperitoneum and posterior mediastinum are the most common locations for GN. GN rarely occurs from the adrenal medulla. It is generally difficult to diagnose and is observed most commonly in children and women. The tumor is not associated with hormonal activity and is often clinically silent (1-3). We present the case of a 35-year-old woman who had mass in the right adrenal gland which was pathologically diagnosed as GN after adrenalectomy.

Case

Abdominal ultrasonography performed for abdominal pain in a 35-year-old woman revealed a mass of 18x55 mm in the right adrenal gland. Computed Tomography (CT) showed a minimally hypodense, smoothly marginated mass without calcification (Figure 1). Right adrenal magnetic resonance imaging (MRI) revealed a smoothly marginated mass of 35x33x21 mm which was minimally hypointense on axial T1-weighted images (Figure 2) and hyperintense on T2-weighted images, relative to the paravertebral muscles (Figure 3). The patient was normotensive and hormonal parameters were normal (Table 1).

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Figure 1. Computed Tomography, minimally hypodense, smoothly marginated mass lesion

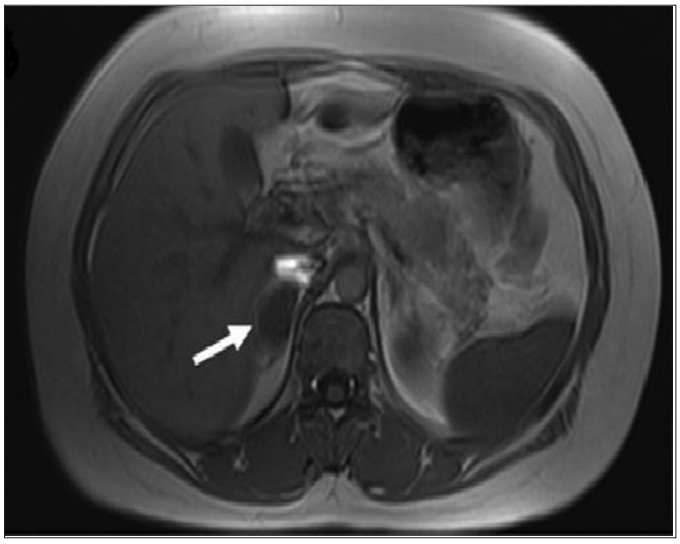

Figure 2. MRI imaging, mass lesion is minimal hypotense in T1-weighted axial images

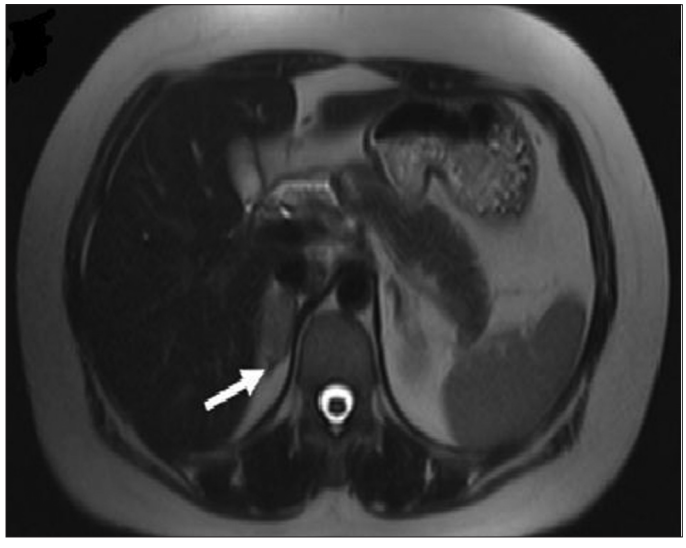

Figure 3. MRI imaging, mass lesion is hyperintense on T2-weighted axial images

Table 1. Preoperative and postoperative hormonal parameters

	Preoperative	Postoperative	Normal
Cortisol (ug/dl)	4.16	11.95	6.2-19.4
11-deoxycortisol (ng/ml)	1.99	2.16	< 8
17-OH progesterone (ng/ml)	3.37	0.79	0.95-5
DHEA-SO4(ug/dl)	109	110.1	33.9-340
Androstenedione(ng/ml)	1.10	2.21	0.5-4.7
Urine Normetanephrine(ug/24h)	65	-	< 390
Potassium (mmol/l)	4.76	4.10	3.6-5.1

Cortisol was found as 1.08ug/dl in 1-mg dexamethasone suppression test. Right adrenalectomy was performed upon patient's request. Pathological evaluation revealed a smoothly marginated, elastic, homogeneous, grey-white 4x4-cm nodular lesion. Microscopic evaluation revealed a benign tumor, composed of spindle cells with Schwann differentiation and ganglion cells with diffuse distribution, arising from the adrenal medulla and separated from the cortex with smooth margins without any capsule. The patient was diagnosed as GN after observing positivity for chromogranin and synaptophysin in ganglion cells and for S-100 in Schwann cells in immunohistochemical evaluation. During the short postoperative ACTH stimulation test, cortisol values at 0, 30, 60, 90 and 120 minutes were 8.5, 17.4, 20.1, 22.1, 23.8 ug/dl, respectively.

Discussion

Although malignant transformation has been reported, adrenal GNs are usually rare and benign. They are observed most commonly in children and young adults (2,4). In a series of patients, average age was found as 40 years (2). These tumors have an equal frequency in male and female patients (5). Our case was a 35-year-old woman.

Most GNs are asymptomatic and they are incidentally discovered during imaging. However, some big GNs may cause pressure symptoms. Most GNs do not secrete catecholamines or steroid hormones (1,6). In a series of 17 adrenal GN patients, 11 were diagnosed during controls, 2 were diagnosed after imaging studies for low back pain and the remaining four were diagnosed after imaging studies for abdominal pain, billiary colic, abdominal trauma and fatigue, respectively. None of these patients had hormonal secretion (2). In our patient, a mass was detected in the right adrenal gland during abdominal ultrasonography performed for abdominal pain and her hormonal parameters were normal.

When GNs are evaluated radiologically, the specific features observed in CT are as follows: presence of punctuate calcifications, absence of vessel involvement and a non-enhanced attenuation of less than 40 Hounsfield Units. In MR, the features observed are low non-enhanced T1-weighted signals and slightly high and heterogeneous T2-weighted signals with a late and gradual enhancement on dynamic MRI. Even though many aggressive tumors such as adrenal

carcinoma share some of these findings, diagnosis of GN should be considered in the presence of all or most of these findings (3). In our case, CT revealed a smoothly marginated mass without calcification and minimally hypodense relative to the paravertebral muscles. Non-enhanced MRI demonstrated a smoothly marginated mass lesion that was minimally hypointense on T1-weighted axial images and hyperintense on T2-weighted images, relative the paravertebral muscles.

Fine-needle aspiration (FNA) biopsy can be useful to diagnose adrenal lesions. However, in as much as 20% of cases, the material is insufficient for diagnosis. FNA biopsy should be avoided when pheochromocytoma or echinococcal parasitic cysts are suspected. Using FNA biopsy in routine evaluation of adrenal incidentalomas is controversial due to concerns with risk of metastatic spillage and lack of consensus on diagnostic accuracy (2,7).

According to the 2002 National Institutes of Health (NIH) consensus statement, adrenalectomy should be considered both in patients with clinically unapparent functional adrenal tumors and in those with nonfunctional adrenal incidentalomas >6 cm. Adrenalectomy should be strongly recommended if imaging findings suggest that the lesion is not an adenoma with rapid growth rate and decreased lipid content (7,8). Some authors recommend adrenalectomy in case of presence of one or more of the following: tumor size ≥6 cm, suspicious appearance on MRI and CT, and patient's fear of malignancy (2,7). Our case was operated due to severe malignancy fear and anxiety too.

In pathological investigation, adrenal GNs are, in most cases, sublobular or nodular in shape, with grey or greyish sections. Morphologically, they are composed of ganglion cells and spindle-shaped cells. Typically, GNs lack the external lamina. They stain positively for vimentin, S-100, synaptophysin and chromogranin A (2,9). In the pathological investigation of our case, there was a smoothly marginated, elastic, homogeneous grey-white nodular lesion, while microscopy revealed spindle cells with Schwann differentiation and ganglion cells with diffuse distribution. In immunohistochemical evaluation, a positive staining for chromogranin and synaptophysin in ganglion cells and for S-100 in Schwann cells were observed and, thus, the patient was diagnosed as GN.

Surgical resection is the first treatment of choice in the treatment of adrenal GNs (1). Our case was treated surgically. Clinical and laboratory features of the postoperative patient were normal and, thus, the patient was checked and discharged. We present this case report to emphasize the rareness of GNs and its place in excluding malignancy of adrenal masses.

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