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Case Report

Large Ganglioneuroma Case Mimicking as An Adrenal Adenoma

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Abstract

Ganglioneuroma (GN) is a rarely seen benign tumor originating from neural crest cells and consisting of ganglion and Schwann cells. Adrenal GNs occur most frequently in the fourth and fifth decades of life. They have an equal frequency in male and female patients and are usually found incidentally during imaging. It is not related to hormonal activity and is clinically asymptomatic. We aimed to present a 49-year-old female patient whose magnetic resonance image performed for abdominal pain was found a biochemically normal mass in the right adrenal gland and then was pathologically diagnosed as GN after right adrenalectomy.

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Introduction

Ganglioneuroma (GN) is a rare tumor with benign behavior mainly originating from neural crest cells.[1] Histologically, it consists of Schwann and ganglion cells.[2] The lesion is mostly located along the length of the sympathetic chain.[3] While they are usually located in the posterior mediastinum or retroperitoneal space, they rarely occur from adrenal medulla.[3] Retroperitoneum and posterior mediastinum GNs are usually diagnosed in children and young adults, while adrenal GNs occur most frequently in the fourth and fifth decades of life. In the case series, it has been determined that GNs are seen equally in males and females.[4,5] Adrenal GNs can be seen as single or together with other neuroendocrine tumors.[4] Since these lesions do not tend to be hormonally active, they are discovered incidentally during imaging techniques. The imaging features of adrenal GN are variable, and some may resemble adrenal tumors such as adrenocortical carcinoma and malignant pheochromocytoma.[4,5] For this reason, it is generally difficult to diagnose adrenal GN preoperatively precisely. In the current case, we aimed to present a 49-year-

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Table 1. Laboratory parameters of the patient at the admission

Variables	Results	Reference range
Glucose (mg/dL)	91	74-106
Creatine (mg/dL)	0.63	0.66-1.09
ALT (U/L)	9	3-35
Sodium (mEq/L)	139	136-146
Potassium (mEq/L)	4,11	3.5-5.1
Calcium (mg/dL)	10,51	8.8-10.6
LDH (U/L)	185	25-247
ACTH (pg/mL)	29.8	0-46
Cortisol (μg/dL)	15.99	6.7-22.6
Aldosteron (ng/dL)	13.73	7-30
Renin (ng/mL/saat)	4.06	0,98-4,18
DHEA-S (ug/dL)	180.3	56.2-282.9
Urine normetanephrine (ug/24 saat)	254.17	100-500
Urine netanephrine (ug/24 saat)	93.35	50-250
Urine ndrenaline (ug/d)	6.26	0-20
Urine noradrenaline (ug/d)	37.42	15-80
Urine nopamine (ug/d)	195.58	65-400
ESR (mm/h)	14	0-20

ALT: alanine aminotransferase, LDH: lactate dehydrogenase, ACTH: adrenocorticotropic hormone, DHEA-S: dehydroepiandrosterone sulfate, ESR: erythrocyte sedimentation rate.

old female patient pathologically diagnosed as GN after adrenalectomy, which was found incidentally mass in the right adrenal gland.

Case Report

A 49-year-old female patient was admitted to our center when a mass of 81x48x92 mm in the right adrenal gland was revealed in abdominal magnetic resonance imaging (MRI) performed for abdominal pain (Figure 1). She had hypertension as comorbidity, which was under control with 10 mg amlodipine. A physical examination was normal. The results of laboratory and hormonal analysis were all found to be within the normal ranges (Table 1). Endocrine investigation for excessive hormone secretion, including urine catecholamine and hormonal parameters, were normal. Cortisol was found as 1.1 ug/dL in the 1 mg dexamethasone suppression test. MRI revealed a mass of 81x48x92 mm, which was hypointense on axial T1-weighted images and heterogeneously hyperintense on T2-weighted images, relative to the paravertebral muscles (Figure 1). Right adrenalectomy was performed on the patient due to the size of the lesion after excluding pheochromocytoma. The surgical specimen was evaluated, and immunohistochemical examination showed positive staining of S-100, neuron-specific enolase (NSE), and synaptophysin. The tumor was diagnosed GN after right adrenalectomy.

Discussion

Although malignant cases have been reported, ganglioneuroma is a rare, generally benign tumor arising from primordial neural crest cells.[1,6] Although most retroperitoneum and posterior mediastinum GN cases are under the age of 20, adrenal GNs are diagnosed in the fourth and fifth decades.[4,5,7] GN have an equal frequency in male and female patients.[4] They are usually asymptomatic and found incidentally during imaging. Symptoms may develop due to the pressure from mass in large lesions. In a case series comprising 17 adrenal GN patients, eleven of them were diagnosed during controls incidentally, four of them were diagnosed after imaging studies for abdominal pain, and two of them were diagnosed after imaging studies for low back pain. None of these patients had hormonal secretion.[5] Similarly to previous reports, in the present case, a mass was

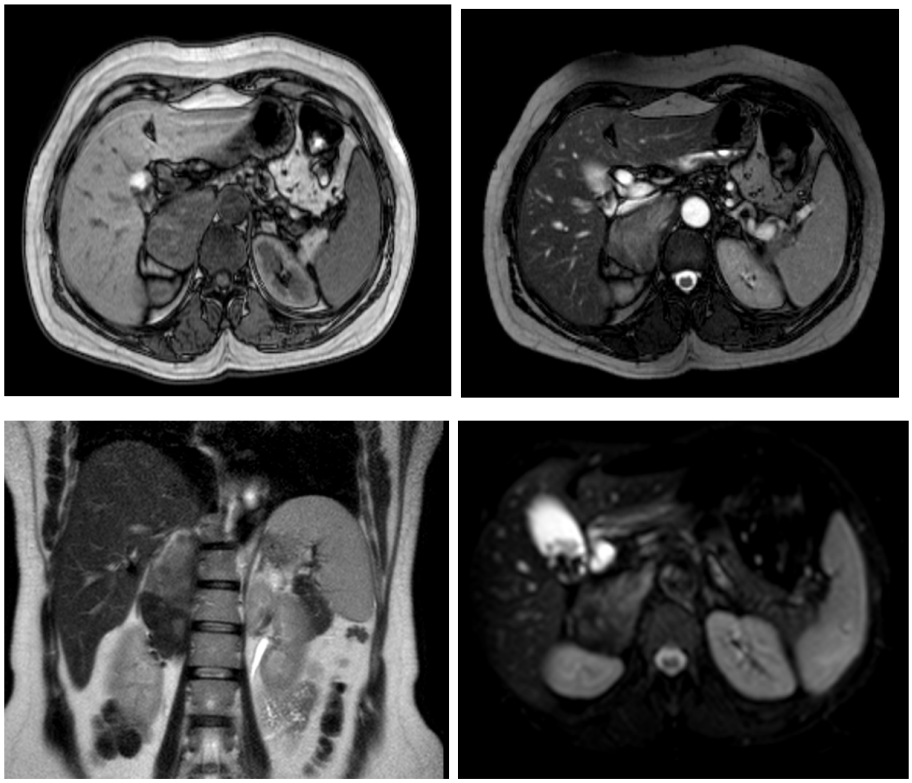

Figure 1. Mass lesion is in the right adrenal region on MR imaging

detected in the right adrenal gland on abdominal MRI performed for abdominal pain at 49-year-old female patient.

Most GN do not secrete catecholamines or steroid hormones, but it has been reported that up to 30% of patients may have elevated urinary catecholamine levels. Rarely ganglion cells can secrete vasoactive intestinal peptide (VIP) or produce steroid hormones. Diarrhea, hypertension, or hypokalemia may be observed due to VIP secretion.[3,6,8] Computed tomography (CT) findings are usually compatible with a well-defined, encapsulated, solid lesion. These tumors can been appeared punctuate calcifications, and nonenhanced attenuation of less than 40 Hounsfield Units on CT imaging. Punctate calcifications are observed at a frequency rate from 20% to 69% and are considered highly indicative of GNs. On MRI, T1-weighted images tend to have homogeneously low or intermediate signal, whereas T2-weighted images have heterogeneously intermediate or high signal.[4,5] Unfortunately, radiology findings are not pathognomonic of adrenal GNs, and preoperative misdiagnosis frequency of adrenal GNs based on MRI and CT findings has been observed to be 64.7%.[5] In the current case, a large mass in the right adrenal gland was located on MRI, which has not clinically significant hormone secretion, and the imaging findings could not eliminate the suspicion of malignancy.

The treatment of adrenal incidentalomas varies according to the functional status of the lesion and whether it is malignant. Regardless of size, functional adrenal lesions with clinically significant hormone excess should be surgically removed. Non-functional lesions <4 cm can be followed up, and surgery should be planned in case of progression (surgical resection if the lesion enlarges by more than 20% or at least a 5 mm increase in maximum diameter).[9] A clear approach has not been determined for asymptomatic adrenal lesions between 4-6 cm. However, surgery should be recommended for adrenal lesions larger than 6 cm. In a study conducted by Qing et al.[5], the mean pathologic size of the adrenal GNs was found at 6.3±3.1 cm. In the present case, performed right adrenalectomy due to the size of the mass and suspicion of malignancy. In pathological evaluation, adrenal GNs are composed of spindle-shaped cells and ganglion cells. They stain positively for S-100, vimentin, chromogranin A and synaptophysin.[4,10] In the

present case, immunohistochemical evaluation, positive staining for NSE and synaptophysin in ganglion cells and for S-100 in Schwann cells were observed. So, the patient was diagnosed as GN.

In conclusion, the detection of adrenal lesions has become easier nowadays due to the ease of access to imaging examinations. GNs are difficult to diagnose preoperatively because they resemble other tumors radiologically. Adrenal GN should be kept in mind in the differential diagnosis, especially in large asymptomatic masses that are not hormonally active.

Conflict of Interests

Authors declare that there are none.

Acknowledgment

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