DOI: 10.4274/tjem.2804

Adrenal Schwannoma: A Very Rare Cause of Adrenal Incidentaloma

Adrenal Schwannoma: Nadir Bir İnsidenteloma Nedeni

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Abstract

Adrenal schwannomas are very rare tumors. They originate from the myelin sheath of the nerves in the adrenal medulla. While adrenal schwannomas can rarely cause some clinical symptoms, such as abdominal pain and back pain, most of them present as a nonsecretory adrenal mass in an asymptomatic patient. We here in report a 41-year-old male who was referred to our endocrinology outpatient clinic due to an adrenal mass detected on abdominal ultrasound during evaluation of abdominal pain. Triphasic computed tomography of the abdomen demonstrated a 29x26 mm right adrenal mass with a density of 24 HU which did not show washout. Hormonal work-up revealed that adrenal tumor was nonsecretory. Since the radiological characteristics of the lesion were suspicious for a malignant tumor, the patient underwent right adrenalectomy, and the pathological examination demonstrated adrenal schwannoma. *Turk Jem 2015; 19: 109-111*
Key words: Adrenal incidentaloma, schwannoma, rare tumor

Özet

Adrenal Schwannomalar oldukça nadir tümörlerdir. Adrenal medullada sinir kılıf hücrelerinden kaynak alırlar. Nadiren müphem karın ağrısı ve sırt ağrısına yol açsalar da genellikle asemptomatik hastalarda fonksiyon göstermeyen adrenal insidentelomalar olarak tesadüfen tespit edilirler. Biz bu çalışmamızda 41 yaşında karın ağrısı nedeniyle yapılan abdominal US de sağ adrenalde 30x30 mm lezyon saptanan hastaya Trifazik Dinamik batın BT yapılmıştır. BT'sinde sağ sürrenal gövdesinde 29x26 mm çapında 24 HU dansitesinde washout göstermeyen kitlesi saptanmış olup yapılan 1 mg dexm. Süpr testi ve idrar katekolaminleri normal bulunmuştur. Hipertansiyon ve hipokalemisi olmayan hasta malinite öntanısı ile üroloji kliniğinde sağ sürrenalektomi yapılmış ve schwannoma olarak tespit edilmiştir. *Turk Jem 2015; 19: 109-111*
Anahtar kelimeler: Adrenal insidentaloma, schwannoma, nadir tümör

Introduction

An adrenal incidentaloma is an adrenal mass, usually ≥1cm in diameter, and discovered during a radiologic examination performed for indications other than an evaluation for adrenal disease (1,2). The prevalence of adrenal incidentalomas was found to be 4% on abdominal ultrasound imaging performed for an unrelated reason. In post-mortem studies, the prevalence of adrenal masses smaller than 1 cm was reported to be 65% (3,4). Adrenal incidentalomas can be either benign or malignant. Among the benign lesions, adenoma, pheochromocytoma, myelolipoma, and adrenal hyperplasia are the most common ones while the adrenal schwannomas are seen extremely rarely. In a study from Canada, 381.200 adrenal incidentaloma specimens were analyzed and adrenal schwannoma was detected in only one case (5). Schwannomas are generally well circumscribed, solitary, and encapsulated tumors. Although they are observed between the ages of 22 and 55 years in most cases, they can be seen at any age. Both sexes are affected equally. Most typically, they grow slowly. In addition, clinical findings of retroperitoneal schwannomas occur late because of the anatomical localization of the tumor (6,7). Here, were present a case of schwannoma as a very rare cause of adrenal incidentalomas.

Case Report

A 41-year-old male had been admitted to an outside hospital with a two-year history of abdominal pain and dyspeptic complaints, and had been found to have a 30x30 mm well-circumscribed, hypoechoic, solid nodule on the abdominal ultrasound examination. He was subsequently referred to our endocrinology outpatient clinic for further evaluation of the adrenal mass. Triphasic computed tomography (CT) of the abdomen demonstrated a 29x26 mm right adrenal mass with a density of 24 HU which did not show washout (Figure 1). He was not a smoker and had a normal weight (body mass index (BMI)=24 kg/m^2) without any cushingoid features on physical examination. His systolic and diastolic blood pressure values were normal (120/80

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mmHg). Routine laboratory results were unremarkable, including a normal serum level of potassium. Hormonal evaluation of the adrenal mass, including midnight plasma and salivary cortisol, urinary free cortisol, plasma cortisol after overnight 1 mg dexamethasone test, and urinary metabolites of catecholamines were all within normal range. Since CT features were suspicious for a malignant mass, metastatic lesion was questioned and a thoracic CT scan was obtained for the primary focus. Thoracic CT findings were normal except for the splenectomy which was performed 6 years previously due to hereditary spherocytosis.

Although the lesion was considered to be a nonfunctioning adrenal mass, the patient underwent right adrenalectomy because of suspicious findings on triphasic CT of the abdomen (Figure 1). Pathological examination demonstrated schwannoma (Figure 2, 3). Immunohistochemical analysis revealed cells that were uniformly S-100 positive, and were negative for CD34, CD177, desmin, and SMA. Ki-67 proliferation index was about 3-4%.

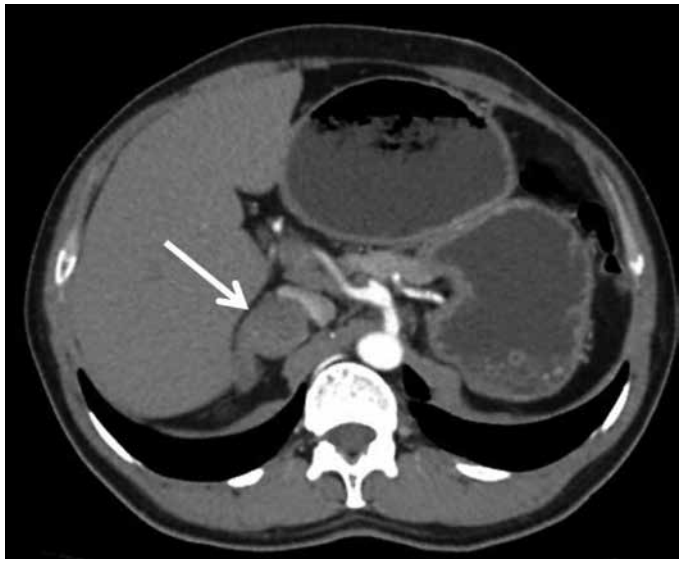

Figure 1. Computed tomography showing an 30 mm, well-circumscribed, enhanced round tumor with solid component arising from the right adrenal gland

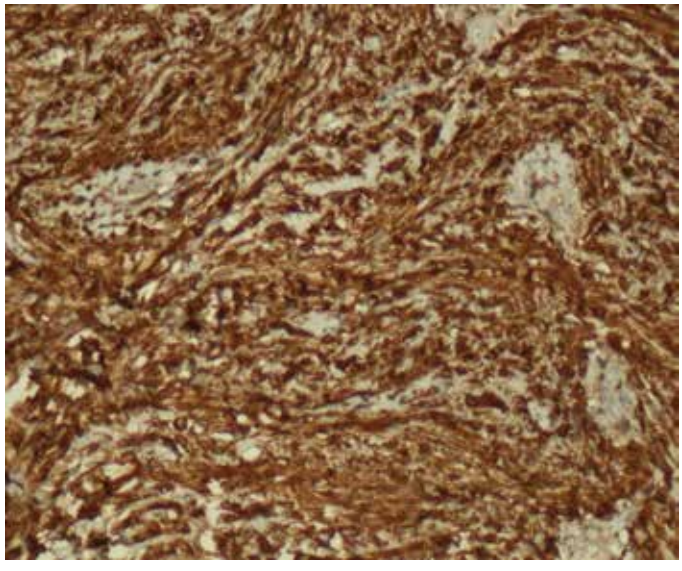

Figure 2. Fusiform cells in parallel bundles (Hex200)

Discussion

The approach to adrenal masses varies depending on whether the tumor is hormonally active or not and whether the tumor is benign or malignant (1). Most adrenal incidentalomas are benign lesions. The frequency of pheochromocytomas and adrenocortical carcinomas has been found to be 1.5-23% and 1.2-12% respectively (2). In the adrenal glands, adrenal metastasis from some cancers, mostly lung cancer, breast cancer, kidney cancer, melanoma, and lymphoma, can be seen. The rate of fat content within the mass determines the tomographic density (Hounsfield units, HU). As fat content in the mass increases, HU decreases. Lesions that have a density of below than 10 HU on a non-contrast CT scan are benign with a sensitivity of 98% whereas lesions with a non-contrast CT density of >20 HU should be considered a malignant lesion.

Adrenal schwannomas are benign nerve sheath tumors commonly originating from the nerve sheath of peripheral, motor, sensory, sympathetic, or cranial nerves within the head, neck, and upper and lower extremities (8). They have been first described by Verocay in 1908 (9). Adrenal schwannomas are exceedingly rare and, most of them are typically found incidentally as non-functioning adrenal masses.

Although clinical findings of schwannomas are usually faded, sometimes patients experience some minor clinical symptoms, such as vague, poorly localized abdominal pain, as seen in our patient. No specific biochemical tests are available for the diagnosis of schwannomas. These tumors are mostly diagnosed incidentally due to widespread use of imaging modalities. The operative exploration is quite often needed for establishing a definitive diagnosis. Small schwannomas are encapsulated, homogeneous tumors, where as large schwannomas show cystic degeneration, hemorrhage, and central necrosis (7,10). Pheochromocytoma, neuroblastoma, and ganglioneuroma are the majority of the tumors arising from the adrenal medulla

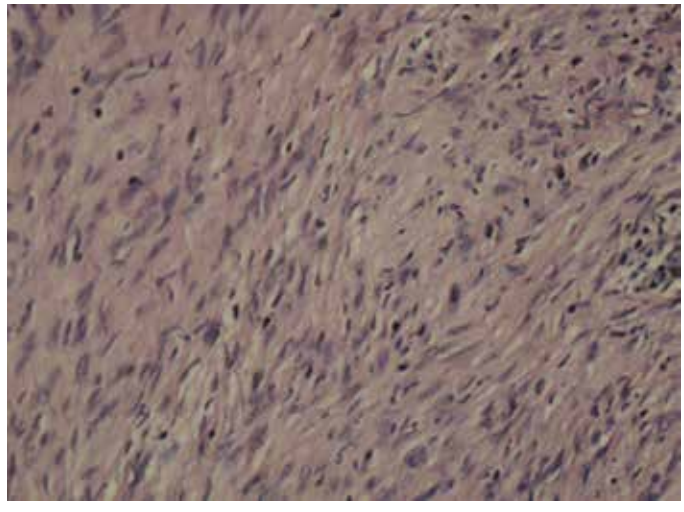

Figure 3. Diffuse S100 positivity in the tumor (immunoperoxidase, x200)

while schwannoma of the adrenal gland is very rare. Among all reported cases of schwannoma originating from the adrenal medulla, no case of schwannoma arising from the adrenal cortex has been described till now. This is likely the consequence of the nerve in the adrenal cortex developing quite poorly compared with that in the medulla and only a few thin nerves running along the vasculature (11).

Adrenal schwannoma arises from either of 2 myelinated nerve systems innervate to the adrenal medulla. One of them is the sympathetic nerve from the upper lumbar plexus, and the other is the phrenic nerve or vagus nerve. Schwannomas have been reported to vary in size from a few millimeters to 15 millimeters. Definitive diagnosis is possible only after histological examination of the operative specimen which typically demonstrates neoplastic cells that simulate the appearance of differentiated Schwann cells that are well circumscribed and composed of spindle cells organized as cellular areas with nuclear palisading and paucicellular areas (12). These tumors are positive for S-100 protein and vimentin (13). Although schwannomas are usually asymptomatic, they can cause some minor clinical symptoms, such as abdominal pain, back pain, or hematuria. Although these neoplasms are usually nonsecretory, rare cases of retroperitoneal schwannomas secreting noradrenalin have been reported (9,14). No cases of recurrence or metastasis has been reported during post operative follow-up (15).

In conclusion, adrenal schwannomas are rare tumors whose radiological appearances look like malignant lesions. No laboratory or imaging methods are available to distinguish these tumors from other adrenal neoplasms.

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Financial Disclosure: The authors declared that this study has received no financial support.

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