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Case Report / Olgu Sunusu

Nadir Görülen Bir Adrenal İnsidentaloma Vakası

An unusual case of Adrenal Incidentaloma

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ÖZET

Adrenal insidentalomalar, başka amaçlarla yapılan radyolojik görüntülemeler sırasında tesadüfen tespit edilen kitlelerdir. Adrenal insidentalomaların büyük kısmını nonfonksiyonel adenomlar oluşturmaktadır. Adrenal insidentalomalarda sessiz bir klinikle seyreden cushingsendromu veya feokromasitoma gözlenebilmektedir. Ancak adrenal insidentalomalı bir vakada cushingsendromu ve feokromasitomanın birlikte bulunması son derece nadirdir. Burada, laboratuartetkikleri subklinikcushingsendromu ile uyumlu, ürinerkatekolaminmetabolitleri normal olan ancak histopatolojik olarak feokromasitoma tanısı konulan bir atipik adrenal insidentaloma vakası sunulmaktadır.

Anahtar Kelimeler: Feokromasitoma, Adrenal insidentaloma, Subklinikcushingsendromu

ABSTRACT

Adrenal incidentalomas are masses accidentally discovered while conducting radiological examinations for other purposes. A major part of adrenal incidentalomas are non-functional adenomas. Silently developing Cushing's syndrome or pheochromocytoma can be observed in adrenal incidentalomas. However, coexistence of Cushing's syndrome and pheochromocytoma at the same time in the same case is quite rare. In the present study, an atypical adrenal incidentaloma case is presented, whose laboratory examinations were compatible with Subclinical Cushing's syndrome, urinary catecholamine metabolites were normal, but who histopathologically had pheochromocytoma diagnosis.

Keywords: Pheochromocytoma, Adrenal incidentaloma, Subclinicalcushing's syndrome

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INTRODUCTION

Adrenal incidentalomas are masses of larger than 1 cm sizes accidentally observed in radiological examinations conducted for other purposes. Incidence of adrenal incidentalomas is 2.3% in autopsy studies and 0.5-2.0% in abdominal CT screenings (1). When detected, adrenal incidentalomas need to be evaluated for malignity and hormonal hyperfunction (2). Although most of the adrenal incidentalomas are non-functional adenomas, aldosteron secreting adenomas, Cushing's syndrome cases and pheochromocytoma can also be observed. The present study deals with an atypical adrenal incidentaloma case whose laboratory evaluations suggested subclinical Cushing's syndrome, who had normal levels of urinary catecholamine metabolites and eventually had pheochromocytoma diagnosis based on histopathological evaluation.

CASE

Thirty-eight years old male patient presented with dyspepsia to another health institution. During abdominal USG examination, a mass was detected in right adrenal area, and then the patient was referred to our clinic for a detailed examination. Blood pressure of the patient, who had been using low dose calcium channel blocker for the last two years, was 120/80 mmHg and pulse rate was 78 beats/min. There was no feature in physical examination. System examinations and routine laboratory tests were normal. A mass of 33x36 mm detected in right adrenal gland by upper abdominal MR examination was found to be compatible with pheochromocytoma (Figure 1). Basal cortisol level of the patient was 13.5 µg/dl (6.2-19.4). Overnight 1 mg, 2-day low-dose and 8 mg dexamethasone suppression tests showed no suppression (9.2, 7.6 and 8.3 µg/dl). Due to some technical limitations, adrenocorticotropic hormone (ACTH) level was not measured. Urine catecholamine level was within normal limits. Following laboratory measurements were recorded: Metanephrine 27.11 µmol/24h (normal, 5-374); Normetanephrine 342.04 µmol/24 h (5-778); vanillylmandelic acid 2.02 mg/24h (1.6-7.3); Dopamine 386.43 µg/24 h (190-450). Hypophysis was normal in MR.

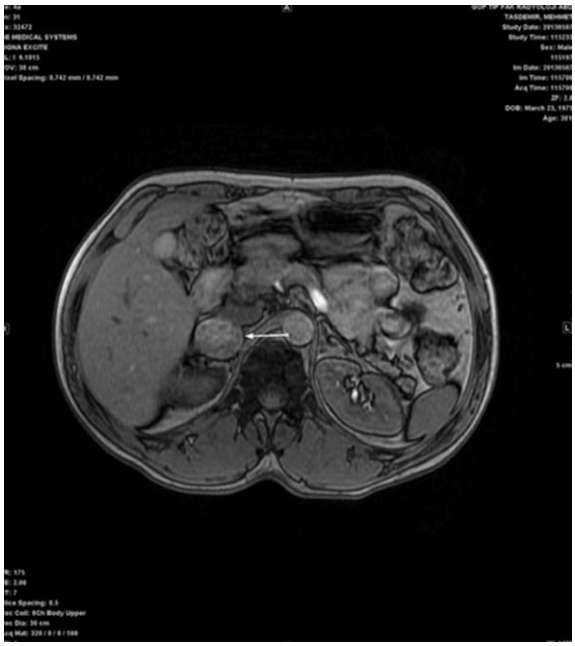

Figure **1A:** Smooth-edged, more hyperintense than spleen parenchyma lesion of 33x36 mm size in right adrenal gland in T1A out-of phase series.

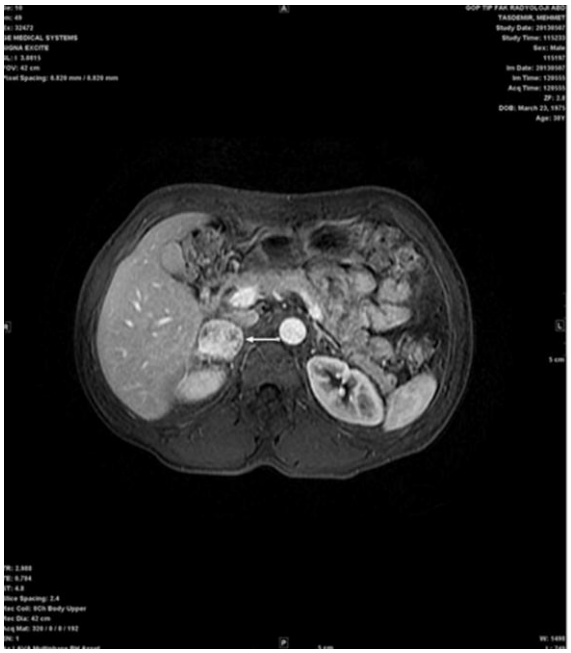

Figure **1B:** Heterogeneously contrasted, smooth-edged, clearly hyperintense mass lesion in right adrenal gland in contrasted T1A series

Gastrointestinal system endoscopic and additional screenings performed to detect malignity was considered normal. The patient was treated for hypertension using calcium channel broker and referred to operation with a diagnosis of subclinical Cushing's syndrome. The mass excised after an uneventful operation was studied histopathologically and pheochromocytoma diagnosis was made (Figure 2, 3).

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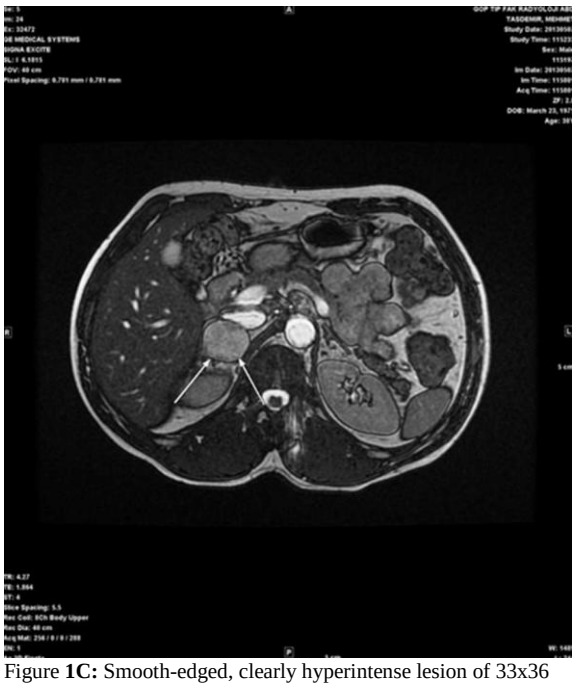

Figure **1C:** Smooth-edged, clearly hyperintense lesion of 33x36 mm size in right adrenal gland in T2A FLAIR series**.**

The patient did not need antihypertensive medication in post-operative period and discharged from the hospital without any problem. In post-operational dexamethasone suppression test, suppression was evident and laboratory findings indicating Cushing's syndrome disappeared.

DISCUSSION

Adrenal incidentalomas are becoming increasingly prevalent thanks to developments in common use of imaging methods. Non-functional adenomas constitute a major part of adrenal incidentalomas. In a study dealing with 380 adrenal incidentaloma patients, 63% were non-functional adenomas, 15% were subclinical Cushing's syndrome, 7% were pheochromocytoma, 6% were myelolipoma, 4% were carcinoma, 3% were metastasis and 2% were aldosteronoma (3). Due to their higher lipid contents, benign lesions are distinguished from the malign ones through the evaluations of imagings such as CT and MR (3). In addition to malignity evaluation, functionality of the mass in terms of hormone production is also important. Functional adenomas generally produce a hormone, but sometimes two different adenomas can be observed in the same gland.

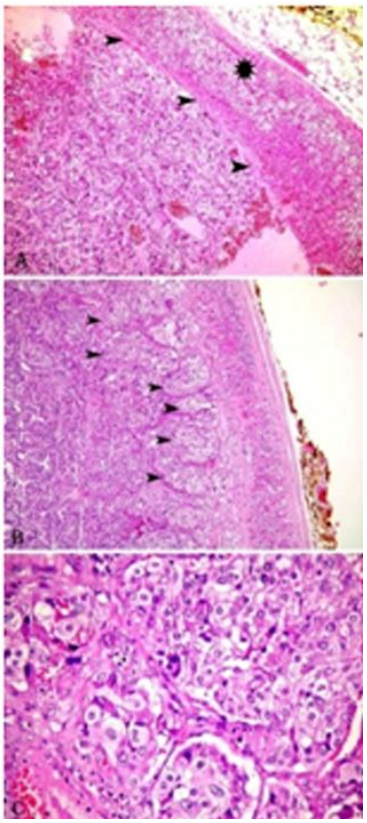

Figure 2:
2A. Vessel-rich tumor lesion separated from normal adrenal gland (asterisk) through fibrous capsule (arrowheads) (HE, X5).
2B. Organization of cells forming the tumor in "Zellballen" pattern (HE, X10).
2C. Tumor cells of large granular cytoplasm and Pleomorphic nucleus (HE, X30).

Cases have been reported in which both pheochromocytoma and adrenocortical adenoma coexisted. Hwang et al. (4) observed in an incidentaloma case both functional pheochromocytoma and adrenocortical adenoma in the same gland. Cotesta et al. (5) detected non-functional cortical adenoma accompanied by pheochromocytoma in post-operation period in two patients who had pheochromocytoma diagnoses.In another adrenal incidentaloma case reported by Sato et al. (6), the observed mass was non-functional, but after the operation, pheochromocytoma and adrenocortical adenoma were confirmed.

In another adrenal incidentaloma case reported by Sato et al. (6), the observed mass was non-functional, but after the operation, pheochromocytoma and adrenocortical adenoma were confirmed. It has been proposed that both catecholamine and peptides secreted from pheochromocytoma such as adrenomedullin, somatostatin and neuropeptide Y affect adrenocortical steroid production (7).

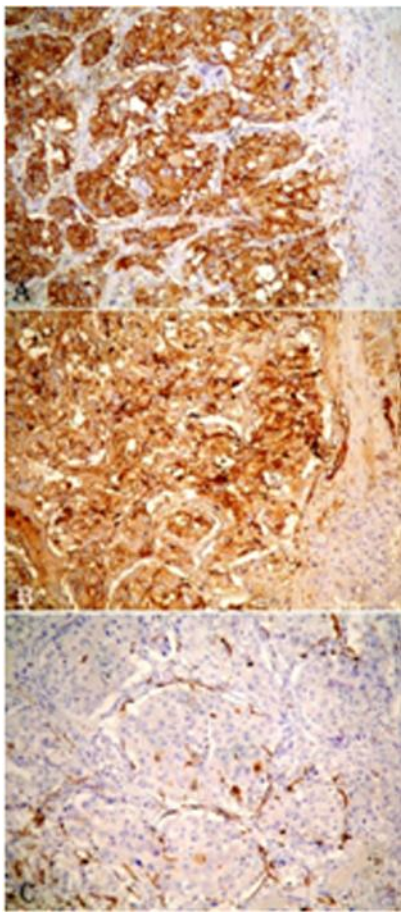

Figure 3:
S-100 positive sustentacular cells with common sinaptofizin (**A**) and chromogranin (**B**) expression surrounding the Zellballen organ (**C**) (DAB, X25).

Prolonged periods of ACTH, catecholamine and several other peptide secretions might lead to adrenal cortical hyperplasia or adenoma formation (4).

There are studies on literature reporting Cushing's syndrome cases depending upon ectopic ACTH production (8-10). In a review paper, Nijhoff et al. (10) mentioned 24 case reports of ectopic Cushing's syndrome due to ACTH production from pheochromocytoma. Most of these cases had clear Cushing's syndrome or pheochromocytoma clinics, and only two of them did not show clinical manifestations of excess catecholamine (11, 12). Our case was of an adrenal incidentaloma and did not have any indication of Cushing's syndrome or pheochromocytoma. While laboratory findings showed normal catecholamine and metabolite levels, dexamethasone tests were compatible with Cushing's syndrome. The case was evaluated as subclinical Cushing's syndrome, but histopathological examinations of the mass excised by a smooth operation confirmed pheochromocytoma. Adrenocortical hyperplasia or adenoma was not observed. ACTH was not evaluated in our case due to technical limitations, but laboratory evaluations indicating Cushing's syndrome, normal hypophysis MR examinations, normal tension after the operation, and evident suppression in post-operative dexamethasone suppression tests supported the diagnosis of our case as Cushing's syndrome of pheochromocytoma origin.

In summary, although rare, pheochromocytoma and subclinical Cushing's syndrome can be seen in the same case. Care should be observed especially for cases, such as ours, which manifest themselves as adrenal incidentaloma and present a silent clinical picture.

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