

A Giant Cystic Pheochromocytoma of the Adrenal Gland: MDCT Findings: Case Report

Adrenal Bezin Dev Kistik Feokromositomu: MDBT Bulguları

Berat ACU, MD, Assis. Prof.,^a
Taylan KARA, MD, Msc,^b
Tansu PINARBAŞILI, MD,^a
Doğan KÖSEOĞLU, MD, Assoc. Prof.^c

Departments of
^aRadiology,
^bPathology,
Gaziosmanpaşa University
Faculty of Medicine, Tokat
^cClinic of Radiology,
Beyhekim Hospital, Konya

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Yazışma Adresi/Correspondence:
Taylan KARA, MD, Msc
Beyhekim Hospital,
Clinic of Radiology, Konya,
TÜRKİYE/TURKEY
taylankara@gmail.com

ABSTRACT Pheochromocytoma is a rare tumor that originated from chromaffin cells of adrenal medulla and secretes catecholamines. Cystic pheochromocytoma is an uncommon variant of adrenal pheochromocytoma. We report a giant cystic pheochromocytoma detected at the adrenal gland. A 38 year old female who has been treated with the diagnosis of iron deficiency anemia since 5 years, presented with abdominal pain. Because of a giant cystic mass detected on ultrasonography, contrast enhanced multidetector-row computed tomography (MDCT) was performed. On MDCT, a 9x9x8cm measured well-defined cystic mass with enhanced mural nodule originated from left adrenal gland was detected. After the surgery the histopathological diagnosis was pheochromocytoma. Pheochromocytomas are usually solid masses with high vascularity. It can rarely be seen as a cystic mass formation. To diagnose the cystic pheochromocytoma is important because of the surgical risk during the preoperative management of the patient.

Key Words: Pheochromocytoma; adrenal gland neoplasms

ÖZET Feokromositom adrenal medullanın kromaffin hücrelerinden köken alan ve katekolamin salgılayan nadir bir tümördür. Kistik feokromositom, adrenal feokromositomun az rastlanan bir varyantıdır. Bu makalede adrenal bezde saptanan dev kistik feokromositom olgusunu multidetektörlü bilgisayarlı tomografi (MDBT) görüntüleriyle sunuyoruz. Beş yıldan beri demir eksikliği anemisi nedeniyle tedavi gören 38 yaşındaki kadın hasta karın ağrısı nedeniyle başvurdu. Karın ultrasonunda sol adrenal bez lojunda dev kistik kitle saptanması üzerine hastaya kontrastlı MDBT yapıldı. MDBT’de sol adrenal bezden kaynaklanan 9 x 9 x 8 cm boyutlu iyi sınırlı, duvarı ve içerisinde kontrast tutan mural nodülü olan dev kistik kitle izlendi. Cerrahi sonrası kitlenin histopatolojik tanısı feokromositom ile uyumluydu. Feokromositomlar genellikle yüksek damarlanma gösteren solid olarak izlenen kitlelerdir. Nadiren kistik kitle olarak da karşımıza çıkabilirler. Kistik feokromositomun tanısı, preoperatif dönemde hastanın cerrahi riskini değerlendirmek açısından önemlidir.

Anahtar Kelimeler: Feokromositoma; adrenal bez tümörleri

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Pheochromocytoma is a rare tumor that originated from chromaffin cells of adrenal medulla and secretes catecholamines.¹ Cystic pheochromocytoma is an uncommon variant of adrenal pheochromocytoma. We report a giant cystic pheochromocytoma with multidetector-row computed tomography (MDCT) findings.

CASE REPORT

A 38 year old female who has been treated with the diagnosis of iron deficiency anemia since 5 years, presented with abdominal pain. Haemoglobin value was 8.4 g/dL. Abdominal ultrasonography (US) was performed. Because of a giant cystic mass detected on US, MDCT was performed. On MDCT, a 9 x 9 x 8 cm measured well-defined cystic mass originated from left adrenal gland was detected (Figure 1-4). The mass had wall and contrast enhanced mural nodule, displaced left kidney to the inferior. According to this findings the diagnosis was cystic neoplasm or hydatid cyst of adrenal gland. A 24 hour urine sample for metanephrine-normetanephrine and vanillylmandelic acid as well as serum catecholamines were extremely high.

The patient underwent to surgery with prediagnosis of pheochromocytoma. During the pathological examination, subcapsular tumoral lesion consisting of nested pattern of atypical cells with polygonal cytoplasm and eccentric nucleus was seen (Figure 5). So the histopathological diagnosis was pheochromocytoma.

DISCUSSION

Pheochromocytoma is a rare tumor that originated from chromaffin cells of adrenal medulla which are embryologically derived from the neuroectoderm and secretes catecholamines.¹ The prevalence of pheochromocytomas confirmed at autopsy is

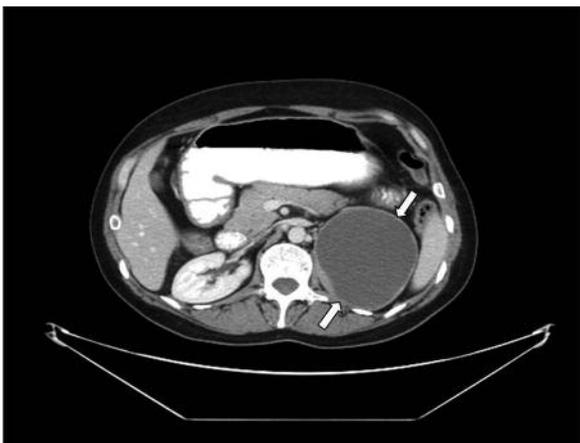


FIGURE 1: Axial postcontrast abdominal CT shows well defined cystic mass with wall at left adrenal gland (arrows).



FIGURE 2: Axial postcontrast abdominal CT shows well defined cystic mass and mural nodule within the wall (arrow).



FIGURE 3: Coronal reconstruction of postcontrast abdominal CT shows cystic mass at the left adrenal gland that compresses left kidney and displaces to inferior (arrows).

0.13%.² Because of releasing of catecholamines, approximately 90% of patients with pheochromocytomas present with hypertension.³ It is reported that more than 90% of patients with a pheochromocytoma will have one or more symptoms of palpitation, diaphoresis and headache.⁴

Approximately 10% of pheochromocytomas are bilateral, malignant, extraadrenal, extraabdominal, familial, pediatric and without increasing blood pressure; this is called as “rule of 10’s” for pheochromocytoma.⁵

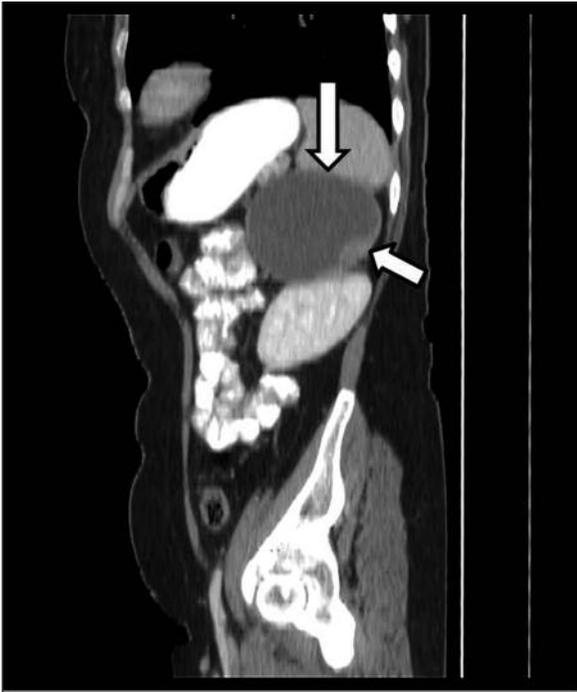


FIGURE 4: Sagittal reconstruction of postcontrast abdominal CT shows cystic mass with mural nodule in the wall compressing left kidney (arrows).

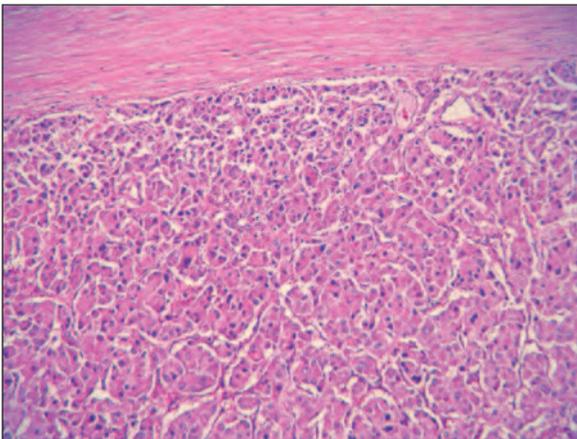


FIGURE 5: Cystic pheochromocytoma (hematoxylin-eosin, X20). A subcapsular tumoral lesion consisting of nested pattern of atypical cells with polygonal cytoplasm and eccentric nucleus (X20, HE).

Close to 90% of pheochromocytomas originate in the adrenal medulla. Extraadrenal pheochromocytomas are also known as catecholamine-secreting paragangliomas.⁶ The most common localization of extraadrenal pheochromocytomas are at the aortic bifurcation in the organ of Zuckerkandl.

Although the patients with pheochromocytoma are usually asymptomatic, chronic lumbar

pain and upper abdominal pain can be observed because of the increasing mass diameters.¹ Elevation of catecholamines metabolites such as metanephrine-normetanephrine and vanillylmandelic acid in 24 hour urine sample and serum catecholamines levels are also diagnostic.^{7,8}

Unless treating effectively, pheochromocytoma may have serious complications, such as myocardial infarction, arrhythmias, hypertensive encephalopathy, cerebrovascular accidents, sudden death, dissecting aortic aneurysm and heart failure.^{9,10}

Pheochromocytomas are usually unilaterally solid masses with high vascularity. The typical appearance of pheochromocytoma is seen as a round, well defined, homogenous mass with soft tissue density measuring over 3 cm on CT.¹¹ Although the attenuation of the mass is similar to that of the liver on unenhanced images, after the contrast injection the mass will enhance because of the vascularity.¹²

Because pheochromocytomas are lipid poor tumors, no signal intensity decreasing during out-of-phase sequences is seen on MRI.⁶

Pheochromocytomas can rarely be seen as a cystic mass formation. In a study including 31 patients with pheochromocytoma, the incidence of cystic pheochromocytoma has found 19%.¹ The characteristic appearance of cystic pheochromocytoma is a relatively thick walled mass, with or without septa, that has persistent wall enhancement after contrast injection. Cystic components of pheochromocytoma reflects necrosis and liquefaction within the mass and these parts have a low attenuation on CT and hyperintense signal on T2 weighted images on MRI.¹

Differential diagnosis of the cystic pheochromocytoma is as the followings: hydatid cyst of adrenal gland, adrenal adenoma and adrenal cyst. To diagnose the cystic pheochromocytoma is important because of the surgical risk during the preoperative management of the patient.

CONCLUSION

Although cystic pheochromocytoma is a rare presentation of pheochromocytoma, it should be kept in mind as a differential diagnosis of adrenal masses.

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