

Cystic Pheochromocytoma-An Unusual Presentation of the Adrenal Mass

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A cystic pheochromocytoma or central hemorrhagic pheochromocytoma is sometimes difficult to differentiate from a benign adrenal cyst by CT or MRI studies. Radionuclide study with ^{131}I -MIBG may serve as a useful tool for preoperative diagnostic purpose. In the present study, we report a case of cystic pheochromocytoma mimic benign adrenal cyst and discuss the procedure of approach.

Key words: cystic pheochromocytoma, ^{131}I -MIBG adrenal scan

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Pheochromocytoma is a catecholamine-producing tumor, which is mainly originated from adrenomedullary chromaffin cells. The most common clinical manifestation is hypertension, but it can also mimic a variety of diseases such as hyperthyroidism, essential hypertension, and emotional disorders. More than 90% of pheochromocytomas appear to be benign in nature. However, the possible associated complications of hypertensive crisis may be fatal [1]. On the contrary, if correctly diagnosed and properly treated, pheochromocytoma is usually curable.

Clinical manifestations of suspicious pheochromocytoma include typically intermittent hypertension, palpitation,

sweating, headache, nausea and abdominal pain. Modern imaging modalities, such as ultrasound, computer tomography (CT) or magnetic resonance imaging (MRI), in conjunction with laboratory parameters, such as elevated plasma levels of noradrenaline or urinary vanillylmandelic acid (VMA), pheochromocytoma can be accurately diagnosed. But for atypical cases, such as cystic, central hemorrhagic or ectopic pheochromocytoma, it is sometimes difficult to diagnose.

By using the radioiodinated metaiodobenzylguanidine (MIBG) adrenal scan, we successfully demonstrated a case of cystic pheochromocytoma whose clinical manifestations and laboratory data were obscure for the diagnosis of pheochromocytoma. The tumor was interpreted as an adrenal cyst by CT and MRI studies initially.

Case Report

A 34-year-old man presented with paroxysmal palpitation, dizziness, dyspnea and epigastralgia. Abdominal sonography revealed a cystic mass measuring about 7 cm in diameter over the right adrenal gland. CT and MRI also suggested an adrenal gland cyst. However, serial 24-hour urinary vanillylmandelic acid (VMA) levels were slightly higher than normal on three consecutive days. Findings of ultrasound-guided fine needle aspiration (FNA) cytology were non-diagnostic. Sudden onset of elevated systolic blood pressure up to 220 mmHg was noted during the aspiration procedure, alerting the clinician the need for further investigation.

Nuclear medicine studies using ^{131}I -MIBG clearly demonstrated a suprarenal mass, which was diagnosed as a pheochromocytoma. Adrenalectomy with the removal of the mass was performed. Grossly, it was a $7 \times 6 \times 3\text{-cm}^3$ cystic mass containing about 90 ml of coffee-brown-like fluid.

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Figure 1. (A) CT and (B)MRI studies both show a well-defined cystic mass with an irregular inner margin in the right suprarenal region. The T1-weighted image of MRI shows different signal level. A renal cyst with central hemorrhage was highly suspected

Pathologic examination proved the mass to be a pheochromocytoma, and a positive chromograinin-A immunohistochemical stain further confirmed the diagnosis.

Discussion

Many diagnostic tools have been used for the evaluation of an adrenal mass. Sonography [2,3], excretory urography, angiography [4], CT and MRI are currently the most commonly used modalities. Of these, sonography was thought to be the method of first choice for the diagnostic procedure in most medical centers. Although it is non-invasive, non-radioactive, rapid, safe and relatively cheaper than other procedures, its resolution is often a major problem.

A previous report by Bush et al. [5] pointed out the fact that the cystic pheochromocytoma is sometimes difficult to

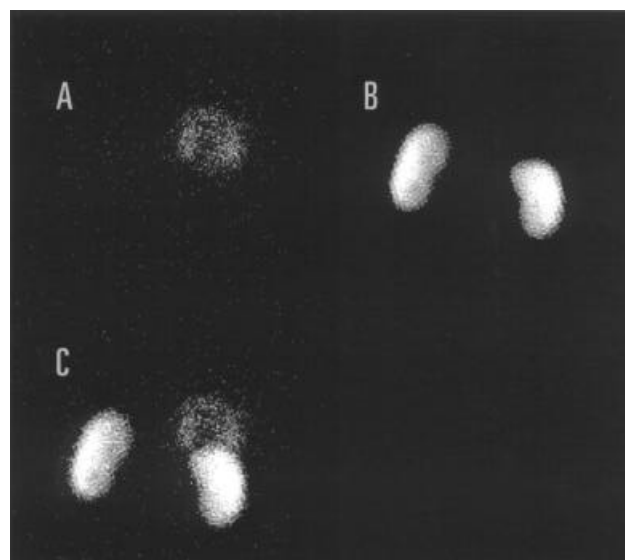


Figure 2. (A) ^{131}I -MIBG imaging combined with (B) $^{99\text{m}}\text{Tc}$ -DMSA renal scanning demonstrated a local "hot" area over the right suprarenal region (C), representing a pheochromocytoma

be recognized by routine diagnostic modalities. It is especially true when differentiating an atypical cystic pheochromocytoma from a benign adrenal cyst because of the vague clinical symptoms and signs, and insufficient laboratory information. Ultrasound-guided FNA is a useful diagnostic procedure in the evaluation of a suprarenal mass, especially in the patient with equivocal roentgenographic findings over the renal area [6]. However, it may also lead to a misinterpretation in the case of cystic pheochromocytoma. In addition, the procedure may induce hypertensive crisis, which is a fetal complication if not treated cautiously.

In this case, no remarkable clinical manifestation was demonstrated. The only biochemical abnormality was a mild increase of urinary VMA value. Available radiological studies favored cystic mass over the right adrenal gland. The only clue implicating a pheochromocytoma was the abrupt increase of systolic blood pressure following the needle aspiration. Under such condition, radionuclide study might play a critical role in identifying the existence of pheochromocytoma. A positive ^{131}I -MIBG study usually indicates a strong evidence for the existence of pheochromocytoma. In contrast, no definite uptake over suprarenal region substantially

excludes the possibility. The study can therefore assist urologists to do better surgical intervention and resolve the problem.

The use of radioiodinated MIBG as the radiopharmaceutical for the detection of functional neuroendocrine tumors was first reported in 1981 by Sisson et al. [7] and was proved to be an effective method in the clinical evaluation of pheochromocytoma. The chemical structure of MIBG is similar to norepinephrine and guanethidine, but without the adrenergic pharmacological effect. When the radioiodinated (^{123}I or ^{131}I) MIBG was injected intravenously into the body, it was taken up from the plasma into chromaffin cells and would localize pheochromocytoma. It has been used widely in initial diagnosis, follow-up, detection of metastases, and localization of ectopic adrenal tissue. The sensitivity of the detection of pheochromocytoma is about 87.4% with specificity up to 98.9%. The most common possibility for a false negative outcome is resulted from medical interference. Drugs such as phenothiazines, tricyclic antidepressants, labetalol, haldol, guanethidine, and reserpine are competitive agents for the binding of MIBG to chromaffin cell [8]. It is, therefore, suggested to discontinue the above-mentioned medicines two to six weeks prior to MIBG studies. Changes of normal anatomic landmark, such as dilated renal pelvis or other nonpheochromocytoma neoplasm, such as carcinoid tumors [9,10] or paragangliomas might also lead to a false positive MIBG study.

A typical solid pheochromocytoma can be easily recognized by a clinician using classic diagnostic modality. However, the cystic pheochromocytoma, which structurally resembles a benign adrenal cyst, is sometimes difficult to have a rational diagnostic approach. Sonography and roentgenographic studies including CT and MRI may be inconclusive in this situation. Radionuclide imaging using MIBG, an analog of guanethidine, may provide a unique means to detect the presence of sympatho-adrenal tissues. We, therefore, highly recommend MIBG scanning for

patients with clinical manifestations of pheochromocytoma. This non-invasive technique may provide reliable, and functional information in the evaluation of typical and atypical pheochromocytoma.

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囊狀嗜鉻細胞瘤

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典型的嗜鉻細胞瘤多伴隨有明顯的臨床症狀，如高血壓等，一般以超音波、電腦斷層攝影或磁振造影等檢查，配合尿液中VMA濃度變化，多可正確診斷。然而，對於非典型的病患，其臨床症狀不明顯、囊狀嗜鉻細胞瘤或中央部分壞死等情形，經常造成誤判。輕微者疏忽而致症狀持續，嚴重者可能因不當處理引發高血壓危機而致死；在此類非典型的病患的診斷中，核子醫學的碘-131-MIBG腎上腺髓質造影檢查，扮演著非常重要的地位。

病患為一34歲男性，因偶發性的心悸和呼吸緊促而求診。由於臨床症狀不明顯，尿中VMA濃度僅稍微偏高，而CT、MRI和超音波均顯示為一腎上腺囊腫；在患者進行細針穿刺時，發生高血壓危機。經碘-131-MIBG腎上腺髓質造影檢查證實為一少見的囊狀嗜鉻細胞瘤。患者後經手術摘除腫瘤，目前情形良好，特於此提出討論。

關鍵詞：嗜鉻細胞瘤，碘-131-MIBG腎上腺髓質造影

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