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Case Report

Cystic adrenal mass revealing a pheochromocytoma in the setting of multiple endocrine neoplasia: A case report ☆

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ABSTRACT

Pheochromocytoma, a neuroendocrine tumor, represents a rare medical condition characterized by the excessive secretion of catecholamines. These tumors often exhibit distinctive features on imaging studies, notably appearing hypervascular. Furthermore, they may present as cystic masses with thin walls, a characteristic that becomes more evident following the administration of contrast medium. The cystic form of adrenal pheochromocytoma, as exemplified in our case, is particularly uncommon, thus underscoring the importance of recognizing its atypical presentation. Accurate diagnosis hinges on a thorough understanding of both the clinical manifestations and radiological findings suggestive of pheochromocytoma. However, definitive confirmation typically necessitates histological examination of the surgical specimen post-adrenalectomy. By shedding light on this rare variant, our case emphasizes the critical role of comprehensive diagnostic approaches in managing such complex medical conditions. Additionally, it underscores the significance of multidisciplinary collaboration among clinicians, radiologists, and pathologists to ensure timely and accurate diagnosis, ultimately guiding appropriate treatment strategies and optimizing patient outcomes.

Introduction

Pheochromocytoma, a neuroendocrine tumor derived from chromaffin cells, is known for its secretion of catecholamines, contributing to its distinct clinical manifestations. While typically presenting as a hypervascular mass, occasionally with necrotic areas, the presence of cystic formations adds complexity to its diagnosis and management [1]. This case highlights the rarity of cystic variants of pheochromocytoma, par-

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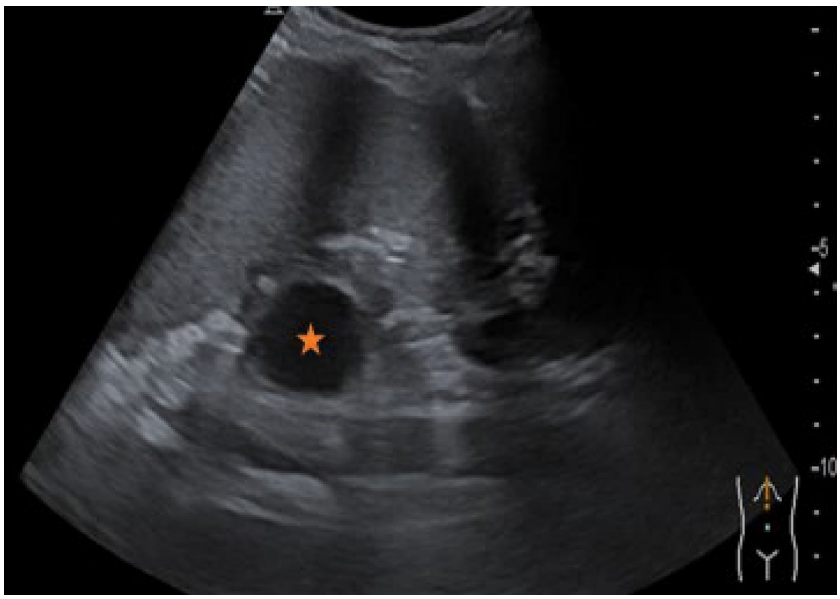

Fig. 1 – Abdominal ultrasound: left adrenal cystic mass (orange star).

ticularly in the context of a young woman with multiple endocrine neoplasia type 2 (MEN2), emphasizing the importance of thorough evaluation and multidisciplinary care within a university hospital setting.

Case report

A 24-year-old woman, currently a housewife, urgently sought medical attention at the emergency department. Despite medical treatment, her hypertension has persisted for 2 months, causing severe symptoms such as pulsating headaches and excessive sweating. It is important to note that she has a medical history of medullary thyroid carcinoma, which necessitated a total thyroidectomy, and hyperparathyroidism that remains untreated, resulting in high parathyroid hormone levels of 115 pg/mL and calcium levels of 12 mg/dL.

The physical examination revealed a patient in good general health, with a body mass index of 20 kg/m^2, a blood pressure of 220/100 mmHg, and a pulse rate of 70 beats per minute.

Additional examinations were carried out in search of a pheochromocytoma within the framework of MEN2. The biological workup found a high level of methoxylated derivatives: The metanephrine/24 h level registered at an alarming 4.5 mol/24 h, far exceeding the reference value of 1.40 mol/24 h. Similarly, the normetanephrine/24 h level was recorded at an astonishing 8.40 mol/24 h, surpassing the reference value of 3.60 mol/24 h. These results are highly concerning and require immediate attention.

Abdominal ultrasonography showed a cystic mass in the left adrenal cavity, pure anechoic, thin-walled, avascular on color Doppler, measuring 32 × 31 mm (Fig. 1). Abdominal CT scan showed a cystic mass of the left adrenal gland, clear boundaries, pure liquid density, and enhanced wall after contrast agent injection, measuring 33 × 31 mm. No signs of the locoregional extension were found on the CT scan, particularly no retroperitoneal adenomegaly (Fig. 2). The diagnosis of adrenal pheochromocytoma was made.

The patient was admitted to a general surgery ward for median laparotomy surgery. Ten days before the surgery, a medical preparation was administered to stabilize blood pressure levels and replenish blood volume. This involved a combination of an alpha and beta blocker, namely phenoxybenzamine at a dosage of 20 mg taken orally three times a day and propranolol at 20 mg taken orally 3 times daily.

Intraoperative exploration, using drugs to prevent blood pressure variations, revealed a cystic mass developing on the left adrenal gland (Fig. 3). A left adrenalectomy by median laparotomy with supra- and sub-umbilical incision was performed carefully, respecting the vascular pedicles. The opening of the posterior parietal peritoneum and the liberation of the left colonic angle allowed the reclination of the left renal vein and the approach of the left adrenal gland. There was no hemodynamic instability during the mobilization and resection of the cystic mass and the postoperative course was simple.

No radiotherapy or chemotherapy was performed. The histological study of the surgical specimen confirmed the diagnosis of cystic pheochromocytoma, showing a tumor composed of nests (Zellballen) of epithelioid cells with abundant eosinophilic cytoplasm and a vascular stroma (Fig. 4).

MEN 2 was retained in our patient because of the involvement of the 3 glands: thyroid, parathyroid, and adrenal, and was confirmed by the RET gene mutation.

The patient's hospital stay after surgery was 10 days. No surgical complications or acute or late postoperative adrenal insufficiency were observed. Clinical symptoms disappeared with the normalization of blood pressure and methoxylated derivatives dosage.

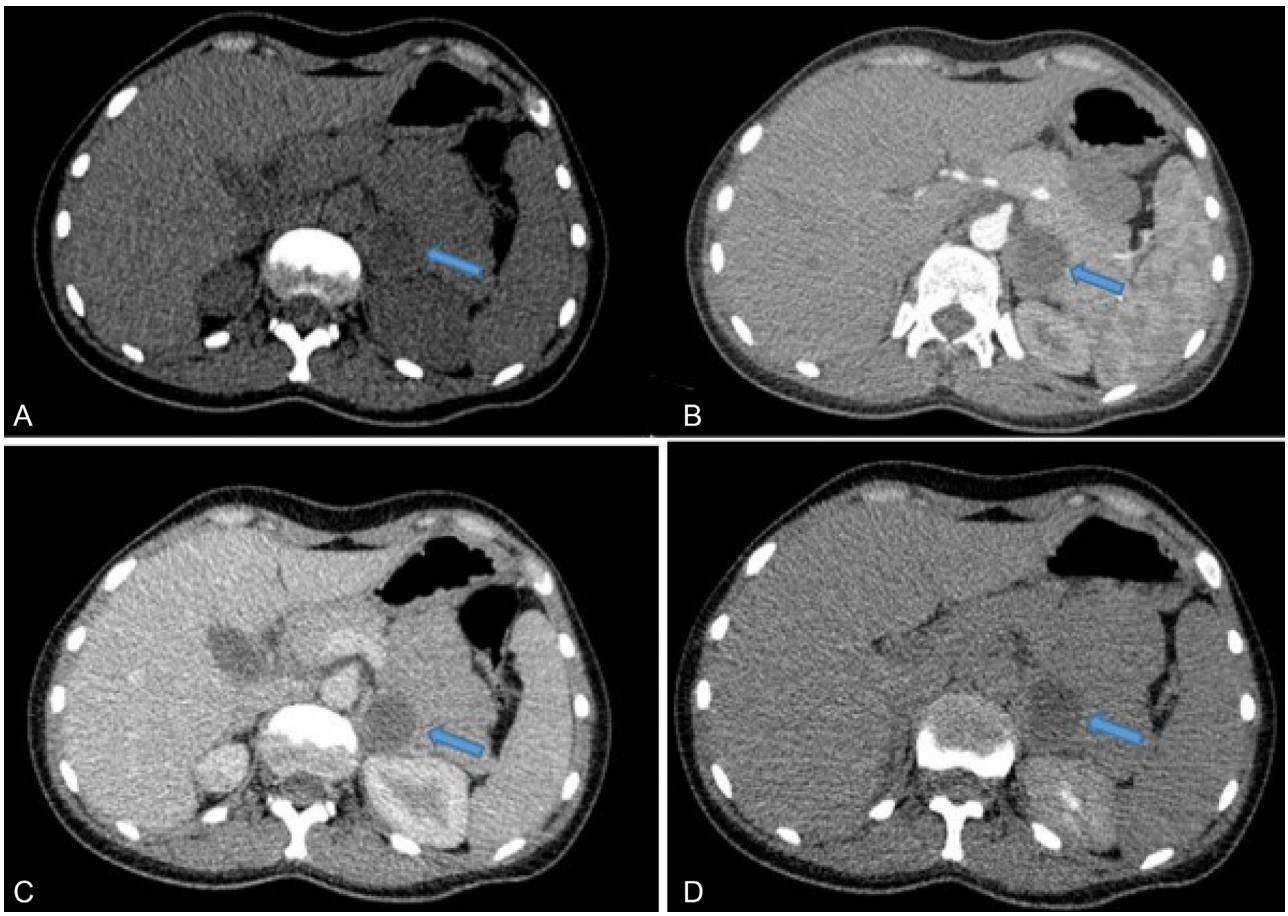

Fig. 2 – Abdominal CT: axial slices without injection (A) and with contrast agent injection at arterial (B), portal (C), and late (D) phase: hypodense left adrenal mass with non-enhanced fluid density after PDC injection (blue arrow).

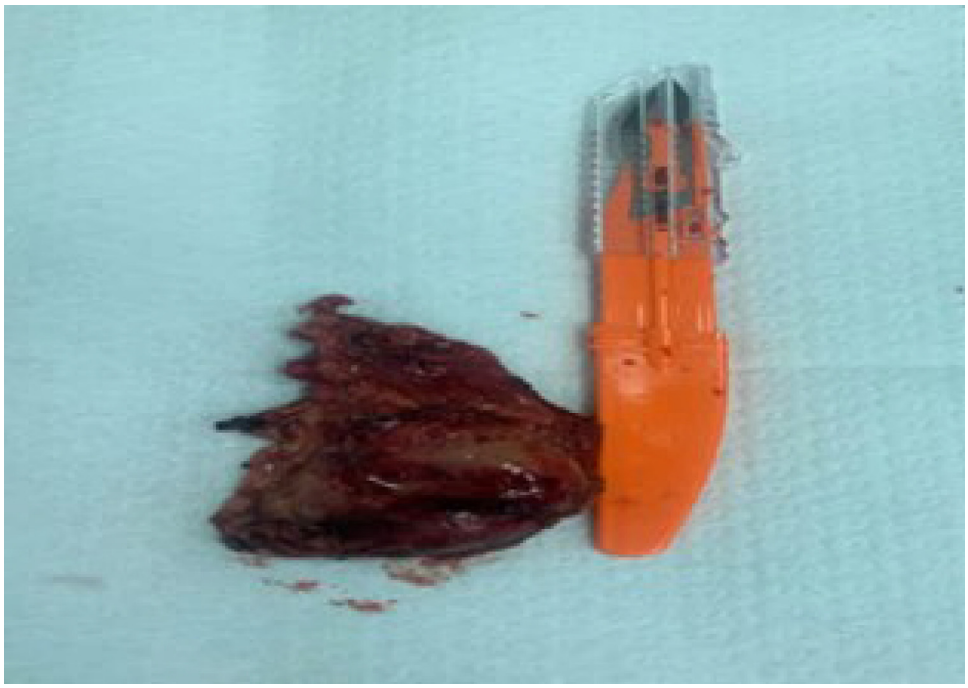

Fig. 3 – Surgical specimen: left adrenal cystic mass.

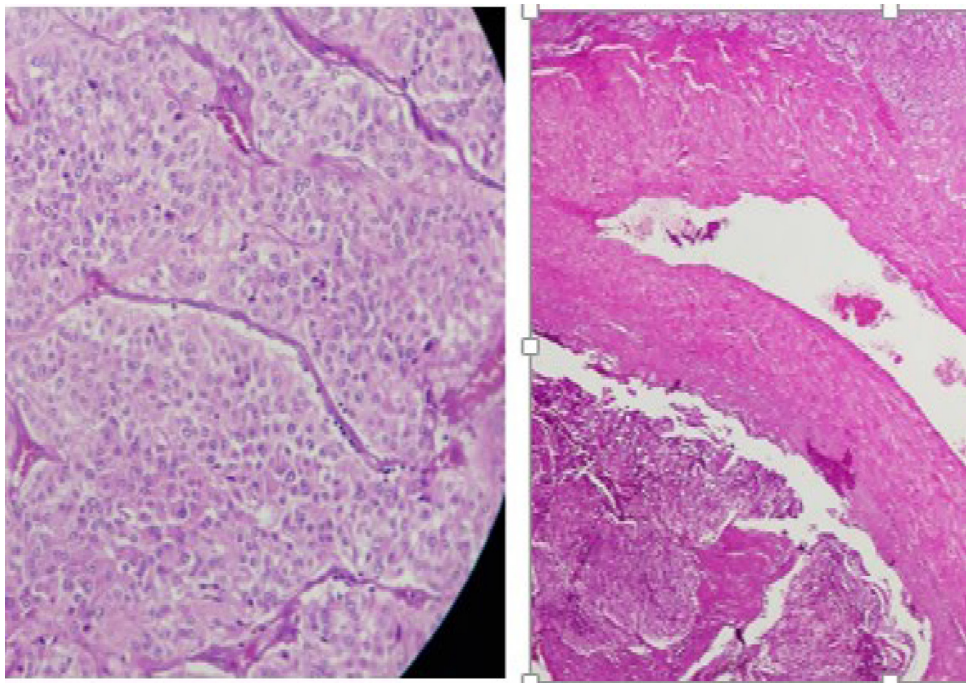

Fig. 4 – Histological aspect of a cystic pheochromocytoma (Magnification x 40) : tumor composed of nests (Zellballen) of epithelioid cells with abundant eosinophilic cytoplasm and a vascular stroma.

Discussion

Pheochromocytoma is a chromaffin cell tumor originating from the sympathetic system, often secreting catecholamines. The vast majority of pheochromocytomas are single and benign, intra-adrenal in 70% of cases [1]. An uncommon variant of adrenal pheochromocytoma is the cystic form.

Although small nonfunctional adrenal masses are asymptomatic, usually discovered incidentally on abdominal imaging, most functional adrenal masses are suspected clinically and confirmed by a biologic workup before a radiologic study. The clinical manifestations of pheochromocytoma are due to excess catecholamine production and include hypertension associated with Menard's triad of "headache/palpitations/sweating," which is found in nearly 90% of pheochromocytoma cases [2,3]. The first stage in diagnosis includes a bioassay of metanephrines and normetanephrines. The sensibility of the urinary VanillylMandelic Acid(AVM) for diagnosing a pheochromocytoma is 98%. The urinary metanephrines have a sensitivity of at least 90% and a positive predictive value of 83% [4]. Pheochromocytomas can be part of a syndrome associated with multiple tumors, including MEN2, as in our patient's case. In this case, genetic counseling is necessary to search for a germline mutation [1].

Abdominal CT is the recommended imaging for the characterization of adrenal lesions, with acquisition performed in spontaneous contrast and after injection of contrast medium at the portal (70 sec) and late time (10 min). The scannographic appearance of pheochromocytoma varies from a small, solid, homogeneous lesion to a large, hypervascular, heterogenous mass that may contain areas of central necrosis.

The cystic form of adrenal pheochromocytoma is extremely rare (0.064%-0.18% in autopsy series), and less than 500 cases have been reported in the Western literature [5]. Terrier and Lecene proposed the first classification of adrenal cysts in 1906, later simplified by Abeshouse. Foster proposed categorizing this tumor into 4 types based on histological type: parasitic cyst, endothelial cyst, epithelial cyst, and pseudocyst. Endothelial cysts are the most common, accounting for 45% of all cases in the world's medical literature [5].

The finding of a cystic adrenal mass on abdominal CT may be difficult to distinguish from another suprarenal cystic lesion. The diagnosis is made after a confrontation with clinical and biological data. Typically, adrenal cysts are round or oval in shape with a thin wall. CT attenuation is identical to that of water with a density of about 5 and 15 Hounsfield units, and only the wall enhances after injection of iodinated contrast medium [6]. The unusual formation of adrenal cysts in the pheochromocytoma range is thought to be due to hemorrhage and extravasation, followed by liquefaction with encapsulation forming a fibrous wall [7].

Adrenalectomy is the treatment of choice. The laparoscopic retroperitoneal or transperitoneal approach remains the gold standard for the surgery of pheochromocytoma smaller than 6mm and weighing less than 100 g, whatever its component [8–10]. Currently, the reference approach for pheochromocytoma surgery is laparoscopy; laparotomy is reserved for adrenal masses larger than 8 cm or in the presence of perirenal fibrosis or in case of hemostasis disorders [10]. For our patient, the surgery was performed by laparotomy for technical reasons, namely the non-availability of the laparoscopic column and the lack of experience of the surgeons in the laparoscopic approach.

As in our patient, initiation of adequate preoperative drug therapy is necessary to control blood pressure and reduce the risk of perioperative complications. Dissection of cystic pheochromocytomas may be accompanied by severe hemodynamic variations such as arterial hypertension and arrhythmia that can be controlled by intraoperative administration of drugs such as alpha-blockers, beta-blockers, and calcium channel blockers. The histological study of the surgical specimen confirms the diagnosis.

Conclusion

Pheochromocytoma is a rare and serious pathology especially in its familial form. Adenomas and metastases are the most frequent causes of adrenal incidentalomas. However, the possibility of other much rarer forms of adrenal masses, particularly the cystic form, should not be ignored. Abdominal imaging (ultrasound, CT, and MRI) is very useful for differential diagnosis.

Patient consent

Informed consent for publication was obtained from patient.

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