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Giant Suprarenal Tumour Case Report and Review

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1. Introduction

Suprarenal tumors evoke considerable interest and represent various diagnostic challenges to physicians and surgeons. Adrenal tumors consists the most portion of suprarenal tumors, which can be stratified into adrenal medullary and adrenocortical tumors. Approximately 60% of adrenocortical tumors are hormonally active and show specific signs and symptoms. Patients with a nonfunctioning adrenal tumor usually present with abdominal discomfort due to the mass effect of the tumor. An imaging feature that differentiates benign from malignant adrenal neoplasms is the tumor size.

Here we report a case of giant left adrenal tumour (3.8 kg in weight) with cushings clinical manifestation. Open surgery was done, full recovery was there, follow up was uneventful

2. Background

There are an estimated 1–2 cases per million per year of adrenocortical carcinoma (ACC) in the USA [1]. ACC represents a rare and aggressive malignancy [1]; it is the second most aggressive endocrine malignant disease after anaplastic thyroid carcinoma. ACC is more common in the female population; it has a bimodal age distribution of 5 to 20 years and 40 to 50 years [2]. Out of all the adrenal masses, 60% are hyper-functioning (hormone-secreting) and the rest, 40%, are nonfunctioning (non-hormone secreting) [3].

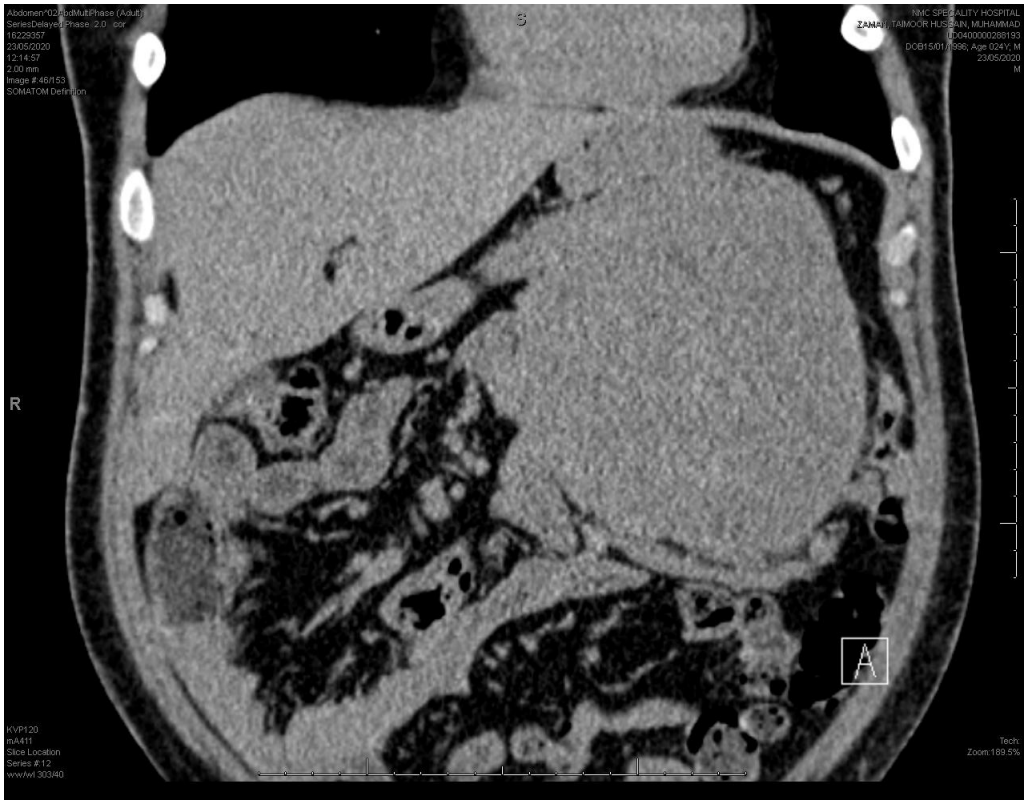
Functional ACC can clinically manifest early with virilization, feminization, or Cushing's syndrome, while non-secretory adrenal masses are diagnosed late and incidentally (adrenal incidentalomas) due to a mass effect or metastatic disease [4]. Nonfunctioning ACC is correlated with a poor prognosis due to the late diagnosis, local invasion, or recurrence and distant metastases [1, 3, 4].

On the other hand, adrenal cysts are usually benign lesions with a discovery at autopsy range of 0.064–0.18%, in which adrenal pseudocysts account for an extremely small amount [4].

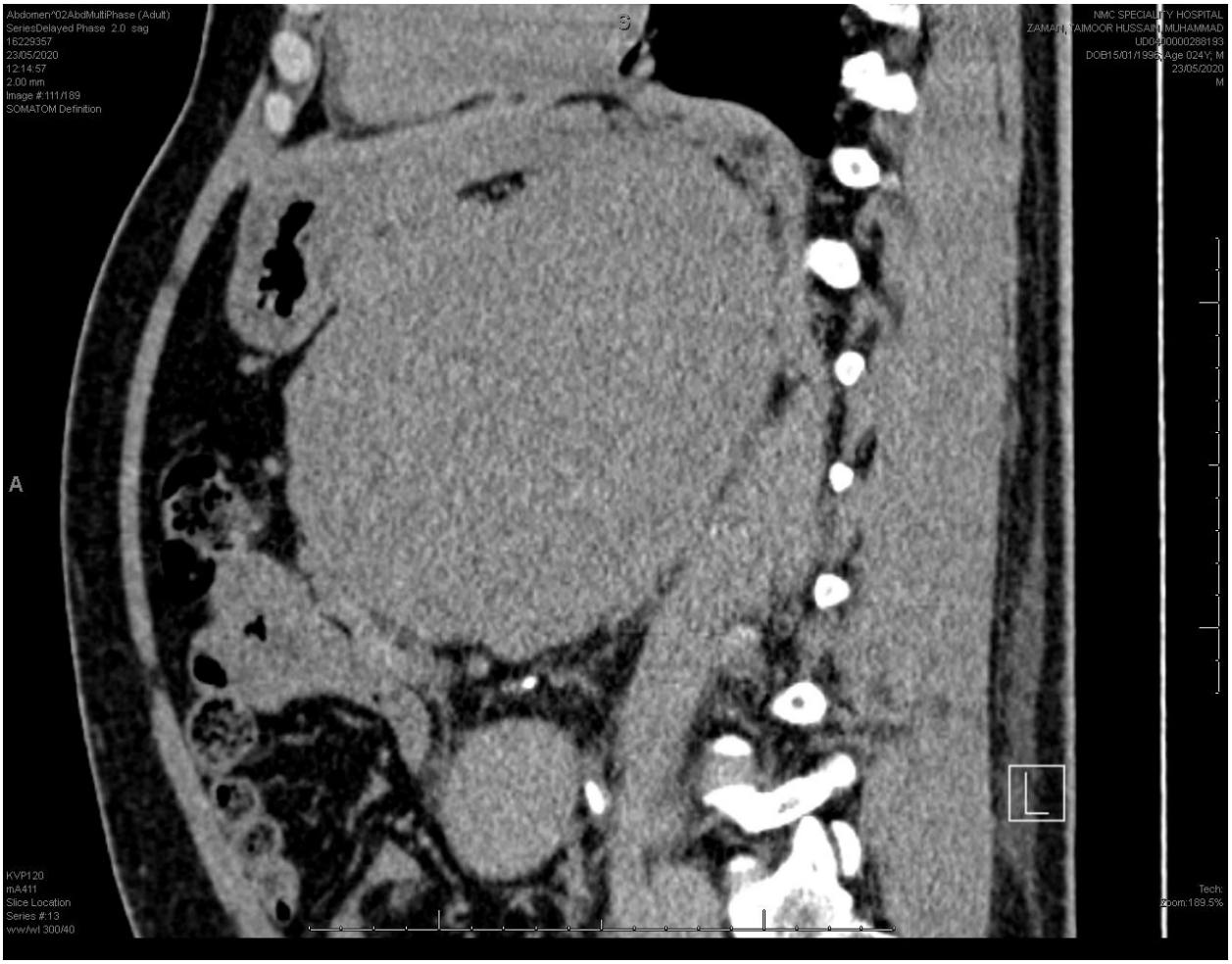
We present one case, who were referred to our Department of surgery and were diagnosed as having a large nonfunctioning ACC. Patient underwent open adrenalectomy.

3. Case Report

A 28-year-old man was referred from the endocraniologist due to the presence of a palpable mass in his left loin region. (fig 1) he was presented many times for the last few months in the emergency unit because of hypokalemia and hypogylcemia attacks associated with other symptoms such as abdominal pain, faceflushing, headaches, weight gain.

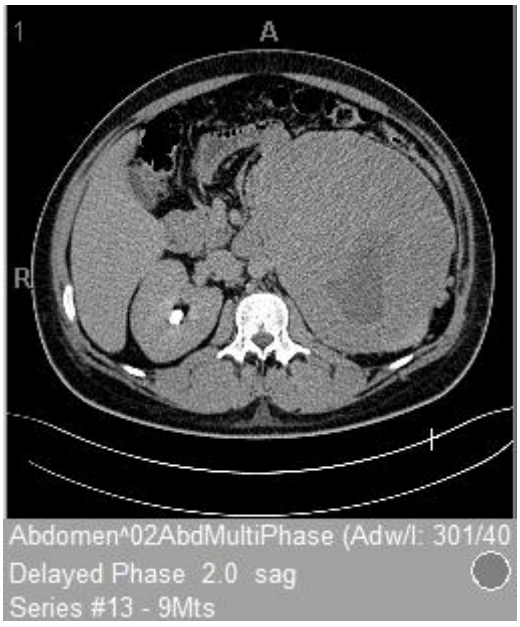


A physical examination revealed the presence of a palpable mass over the upper left quadrant of the abdomen that extended up to the flank region; however, the mass lesion was not found to be tender. Results of abdominal computed tomography (CT) scan revealed a huge left suprarenal mass. There were evidences of calcification and central necrosis within the tumor, which had also downwardly displaced the left kidney (fig no.). All laboratory tests were normal, including urinalysis, complete blood count, and serum chemical studies. (5) An adrenal endocrine survey, including cortisol, testosterone, aldosterone, progesterone, and catecholamine, was also within normal limits. Metastatic evaluation, including a whole-body bone scan and positron emission tomography (PET), revealed nothing. (fig 2)



Differential diagnosis Abdominal CT scan after contrast medium enhancement revealed an inhomogeneous mass with irregular enhancement of the solid components. Calcifications and central necrosis were also visible. (6) No surrounding tissues or organs were involved. The possible differential diagnoses were adrenal medullary-type tumors (pheochromocytomas and rare ganglioneuromas), adrenocortical tumors [adenomas, myelolipomas, oncocytic neoplasms,and adrenocortical carcinomas (ACCs)], neurogenic tumors .schwannomas), and retroperitoneal sarcomas (malignantfibrohis-tiocytomas and leiomyosarcomas).

As is the case with any mass involving the adrenal glands, it is important to differentiate the lesion from an adrenal adenoma. An imaging feature that differentiates benign from malignant adrenal neoplasms is the tumorsize (7).1A diameter of<5 cm usually suggests a benign origin. Furthermore, measurement of the Hounsfield units on an unenhanced CT scan is of great value when differentiating malignant from benign lesions. (fig3)



An attenuation value of>10 HU on an unenhanced CT scan or an enhancement washout of<50% and delayed attenuation of>35 HU (on approximately 10e15-minute delayed enhanced CT scan) suggests a malignancy.2This young male patient had a giant nonfunctional suprarenal tumor that gave attenuation values of approximately 26e35 HU on the unenhanced CT scans and values of approximately 56e78 HU on enhanced CT scans, and therefore, an ACC was suspected. However, it was not possible to rule out the rare possibility of an oncocytic neoplasm, because a pathological feature that is useful when identifying an adrenal oncocytic tumor is a fibrous capsule.(8).

Management Considering the huge size of the mass and the potential possibility of malignancy, the patient underwent open surgery via a trans abdominal approach using a midline incision; this facilitated maximal exposure for the complete surgical excision of the tumor and also minimized tumor spillage. An ovoid, well-circumscribed tumor (25x30 cm) distinct from the left kidney was found. The tumor had compressed the spleen and pancreas and stretched the descending colon, but had not invaded these organs. The tumor was completely removed (9). The patient was discharged on the 7thday after the operation. No adjuvant chemotherapeutic drug such as mitotane was administered. The patient has had advised for regular follow-ups including chest X-rays and CT scans every 4 months and a yearly PET scan. There has been no evidence of recurrence over the last 2 years. (19).

Pathology

The final pathological diagnosis was an ACC. Upon sectioning, we found that the encapsulated tumor was a heterogeneous mass with a brownish-pink to yellow color on the cut surface and showed multiple areas of myxoid change and necrosis.

The surgical margin was free of tumor. A microscopic examination revealed a mass composed of round to polygonal neoplastic cells with abundant eosinophilic cytoplasm. These nuclei were hyperchromatic and pleomorphic, and had obvious nucleoli and mitotic activity [2 per 50 high power fields (HPFs)]. Tumor necrosis was evident. Neither capsular nor vascular invasion was identified. The pathological stage was T2N0M0 (European Network for the Study of Adrenal Tumor, the ENSAT staging system) (20). Immunohistochemical analysis revealed that the tumor cells were strongly positive for cytokeratin and desmin, but were weakly positive for vimentin and S-100 protein, and negative for Ki67.

4. Discussion

The overall 5-year survival rate of ACC ranges from 16 to 44% [4]. However, a more recent study recorded a 5-year overall survival rate that reached 60% and concerned patients with a 34% range of synchronous metastatic disease. Positive prognostic factors that could improve overall survival are early-stage disease in the absence of lymph node and distant metastases, an age of < 40 years, and negative resection margins (Ro), as in our case [1, 4].

On the other hand, adrenal pseudocysts are cystic lesions surrounded by a fibrous tissue wall with an absence of a recognizable epithelial or endothelial lining layer that characterizes true cysts. Adrenal pseudocysts are associated with malignant ACC in 7% of cases [5].

Approximately 60% of patients with ACC present with symptoms and signs of hormonal secretion. Our case ACC were not functional. Hormone secretion is not a discriminating feature between benign and malignant adrenocortical masses. The silent clinical nature of nonfunctioning ACC results in poor outcomes, while the majority of patients present with locally advanced and/or metastatic disease. The work-up for adrenal masses must include determination of whether the mass is functioning or nonfunctioning and whether it is benign or malignant [1, 5].

Radiographic studies in the form of CT or magnetic resonance imaging (MRI) can help define the size of the mass and rule out metastases. Emerging evidence suggests that fluorodeoxyglucose positron emission tomography (FDG-PET) with CT is superior to CT alone [6]. However, FDG-PET/CT is still considered a complementary study and is not recommended for ACC work-up [6].

Large tumors of > 4 cm raise high clinical suspicion of malignancy, as in our cases, and favor radical adrenalectomy [7, 8].

Despite the poor prognosis in this patient group, chemotherapy has a limited role in the treatment of ACC, and surgical resection has been shown to have the best outcomes [9, 10]. However, recent studies have shown a role for adjuvant chemotherapy in prolonging recurrence-free survival and overall survival [11]. There is no established duration of adjuvant chemotherapy.

Allolio et al. considered that complete surgical resection (Ro excision) offers the best chance for long-term survival in patients with stage I–III ACC, although these patients should be candidates for chemo-irradiation in order to increase disease-free survival [12]. In cases of recurrence, surgery should be considered a first-line option. However, mitotane has a valuable role in stage IV cases or in the presence of recurrent disseminated disease [12, 13].

Studies demonstrated mitotane's associated toxicity and its ability in: (1) inhibition of adrenocortical steroid biosynthesis by inhibiting cholesterol side chain cleavage and 11 β-hydroxylation, and (2) induction of hepatic clearance affecting extra-adrenal disposition of cortisol [14]. Concerning the routine use of mitotane, there is a lot of controversy as partial response occurs for 5–30% of patients with ACC treated with this anticancer agent. High-risk patients are currently being treated for 5 years with mitotane.

Alternatively, in patients with proven lung metastasis, the published evidence suggests that en bloc excision of involved organs and pulmonary metastasectomy could improve overall survival.(17)

Postoperative surveillance for recurrence should be performed every 3 months for the first 2 years and then every 6 months for 5 years.

Adrenal pseudocysts most commonly arise from hemorrhage within the adrenal gland, secondary to extreme stress, birth, trauma, surgery, or malignancy. The differential diagnosis of adrenal pseudocysts is from endothelial epithelial (18) and parasitic cysts and rarely from mesothelial cysts, lymphangiomas, dermoid cysts, or cystic adrenal carcinomas. (19,20)

Most pseudocysts on CT reveal well-demarcated, round or oval masses with fluid density with content of septa, blood, and soft tissue components. The wall of a pseudocyst seems to have occasional calcification on CT. MRI is pathognomonic for visualizing the complicated intracystic components [15].

Regarding surgical treatment, the open approach is preferable if the patient is symptomatic, the mass is > 6 cm, or there is a possibility of malignancy. Laparoscopic surgery is indicated for small tumors or pseudocysts provided there is no peri-adrenal infiltration and subsequent capsular disruption [15, 16].

5. Conclusions

Approximately 60% of patients with adrenocortical carcinoma present with symptoms and signs of hormonal secretion. Our cases' adrenocortical carcinomas were not functional. Hormone secretion is not a discriminating feature between benign and malignant adrenocortical masses. The silent clinical nature of nonfunctioning adrenocortical carcinoma results in late diagnosis, while the majority of patients present with locally advanced and/or metastatic disease.

Adrenocortical carcinoma is a rare endocrine tumor with a poor prognosis that can be diagnostically challenging and demands high clinical suspicion. The work-up for adrenal masses must include determination of whether the mass is functioning or nonfunctioning and whether it is benign or malignant.

References

[1] Sivasankar A, Jeswanth S, Johnson MA, Ravichandran P, Rajendran S, Kannan DG, et al. Acute hemorrhage into adrenal pseudocyst presenting with shock: diagnostic dilemmas–report of three cases and review of literature. Scientific World Journal. 2006;6:2381–2387. [PMC free article] [PubMed] [Google Scholar]
[2] Karayiannakis AJ, Polychronidis A, Simopoulos C. Giant adrenal pseudocyst presenting with gastric outlet obstruction and hypertension. Urology. 2002;59(6):946. [PubMed] [Google Scholar]
[3] Koksoy FN, Yucel O, Celik A, Igdem AA. Laparoscopic management of a giant adrenal cyst: case report. Surg Laparosc Endosc Percutan Tech. 2001;11(6):379–381. [PubMed] [Google Scholar]
[4] Tait DL, Williams J, Sandstad J, Lucci JA., III Benign adrenal cyst presenting in a pregnant patient. Am J Perinatol. 1997; 14(8):461–464. [PubMed] [Google Scholar]
[5] Trauffer PM, Malee MP. Adrenal pseudocyst in pregnancy. A case report. J Reprod Med. 1996;41(3):195–197. [PubMed] [Google Scholar]
[6] Sell J, Williamson MR, Boutin RD, Igel BJ. CT demonstration of a giant adrenal endothelial cyst of the angiomatous subtype. Comput Med Imaging Graph. 1994;18(6):469–471. [PubMed] [Google Scholar]
[7] Davenport M, Pollard K, Smith SE, MacMahon MJ. Adrenal cysts–report, review and classification. Postgrad Med J. 1988;64(747):71–73. [PMC free article] [PubMed] [Google Scholar]
[8] Incze JS, Lui PS, Merriam JC, Austen G, Widrich WC, Gerzof SG. Morphology and pathogenesis of adrenal cysts. Am J Pathol. 1979;95(2):423–432. [PMC free article] [PubMed] [Google Scholar]
[9] Uretzky G, Freund H, Charuzi I, Luttwak EM. Cysts of the adrenal gland. Eur Urol. 1978;4(2):97–99. [PubMed] [Google Scholar]
[10] Foster DG. Adrenal cysts. Review of literature and report of case. Arch Surg. 1966;92(1):131–143. [PubMed] [Google Scholar]
[11] Thompson AG, Jacobson SA. Pseudocyst of the adrenal gland. Can Med Assoc J. 1966;94(2):90–91. [PMC free article] [PubMed] [Google Scholar]
[12] Esquivel E, Jr, Grabstald H. Giant adrenal cyst. J Urol. 1965;94(6):635–638. [PubMed] [Google Scholar]
[13] Elliott GB, Walker RH, Wright AS, Elliott KA. Adrenal giant cyst: hemangioma of medulla with osmotic pseudocyst formation. Ann Surg. 1964;159:275–278. [PMC free article] [PubMed] [Google Scholar]
[14] Stewart JO, Lester JG. Ruptured adrenal cyst. Postgrad Med J. 1962;38(446):709–710. [PMC free article] [PubMed] [Google Scholar]
[15] Armitage HV. Benign cyst of the adrenal gland. AMA Arch Surg. 1959;79(1):148–151. [PubMed] [Google Scholar]
[16] Ellis FH, Jr, Dawe CJ, Clagett OT. Cysts of the adrenal glands. Ann Surg. 1952;136(2):217–227. [PMC free article] [PubMed] [Google Scholar]
[17] Abeshouse GA, Goldstein RB, Abeshouse BS. Adrenal cysts; review of the literature and report of three cases. J Urol. 1959;81(6):711–719. [PubMed] [Google Scholar]
[18] Kasperlik-Załuska AA, Otto M, Cichocki A, Rosłonowska E, Słowinska-Srzednicka J, Zgliczyński W, et al. 1,161 patients with adrenal incidentalomas: indications for surgery. Langenbecks Arch Surg. 2008;393(2):121–126. [PubMed] [Google Scholar]
[19] Cantisani V, Petramala L, Ricci P, Porfiri A, Marinelli C, Panzironi G, et al. A giant hemorragic adrenal pseudocyst: contrast-enhanced examination (CEUS) and computed tomography (CT) features. Eur Rev Med Pharmacol Sci. 2013;17(18):2546–2550. [PubMed] [Google Scholar]
[20] Friedrich-Rust M, Schneider G, Bohle RM, Herrmann E, Sarrazin C, Zeuzem S, et al. Contrast-enhanced sonography of adrenal masses: differentiation of adenomas and nonadenomatous lesions. AJR Am J Roentgenol. 2008;191(6):1852–1860. [PubMed]