Bilateral Adrenocortical Carcinoma Presenting as Large Palpable Lumps: A Case Report

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ABSTRACT

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Adrenocortical carcinomas are rare aggressive tumours and bilateral ACC are still rarer. They may be sporadic and unilateral or bilateral and associated with many syndromes. Awareness about the existence of such lesions is needed to clinically suspect these rare lesions. Here we present a case of bilateral adrenal enlargement and discuss the types of presentations and clinical associations.

Keywords: Bilateral adrenal masses, Adreno-cortical carcinomas, Adrenal syndromes

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INTRODUCTION

Adrenocortical carcinomas are rare aggressive tumors. Their annual incidence is approximately one to two per million among the population of the United States of America. Primary malignant tumors originating from the adrenal gland include adrenocortical carcinomas, primary adrenal lymphomas, and malignant pheochromocytomas; however, the incidence of these tumors is low.[1-4] Most adrenal tumors are sporadic and unilateral, but 2% to 6% of adrenal tumors are bilateral and associated with Li-Fraumeni syndrome, type I Multiple Endocrine Neoplasia, Beckwith-Wiedemann syndrome, and Carney complex, principally in children.[1-4] When bilateral adrenal masses are detected, an effort is undertaken to find other primary malignant foci or the kidney injury secondary to renal vein thrombosis.

There are a few reported cases of bilateral primary adrenocortical carcinomas.[1] Here we report one such case of large bilateral adrenocortical carcinoma.

CASE REPORT

A 48-year-old female was referred to our OPD from Surgical Gastroenterology department with bilateral adrenal masses that were detected incidentally by contrast enhanced computer tomography (CT) for evaluation of generalized abdominal pain. Pain was continuous, nonradiating and dull aching in nature. She also had history of loss of appetite and weight loss. No history of any comorbidity. Contrast-enhanced computer tomography (CT) scan revealed bilateral adrenal mass, 17x13x12 cm on left side & 7x7x6 cm on right side with heterogenous enhancing lesion with areas of necrosis & calcification replacing the adrenals **(figure 1, 2 & 3).** Absolute washout of contrast on left side was 41% and on right side was 56%. Relative washout on left side was 23% and on right side was 36%. The results of adrenal function tests were within normal ranges.

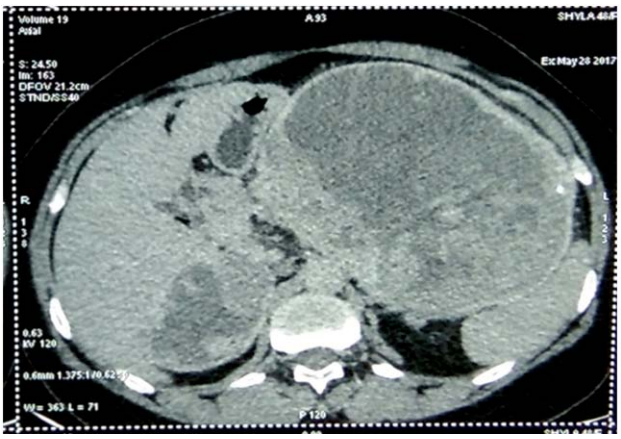

Figure 1. **Abdominal computed tomography (CT) examination of the patient. This abdominal CT image shows bilateral adrenal mass (left > right)**

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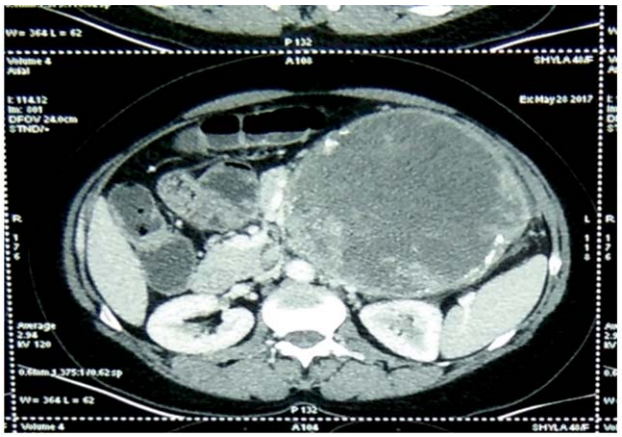

Figure 2. **Contrast enhanced CT abdomen showing heterogenous enhancing lesion with areas of necrosis & calcification replacing the adrenals**

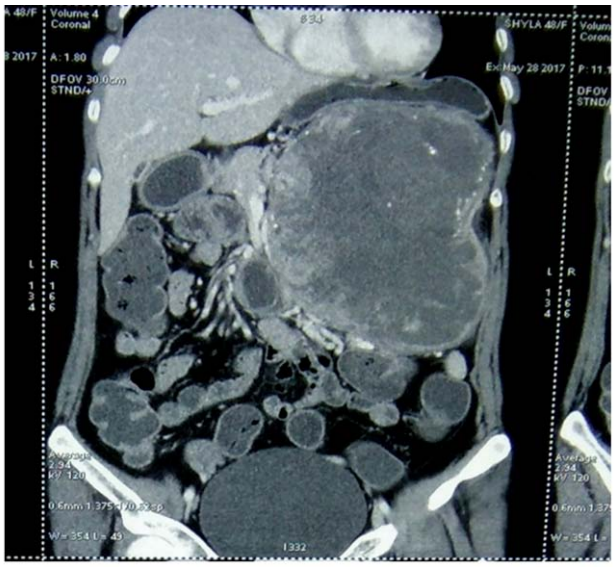

Figure 3. **Sagittal view of abdominal computed tomography (CT) examination of the patient. This abdominal CT scan shows a large adrenal mass displacing the left kidney inferiorly and the spleen superiorly.**

DISCUSSION

Neoplastic involvement of the adrenal gland may result from primary tumors originating from the adrenal cortex of the adrenal gland. Primary malignant tumors originating from the adrenal gland include adrenocortical carcinomas, primary adrenal lymphomas, and malignant pheochromocytomas. Adrenal glands are more frequently the site of metastatic disease caused by primary carcinomas. Any primary cancer can spread to the adrenal glands; lymphomas, lung cancer, melanomas, leukemia, renal carcinoma, and ovarian carcinoma account for the majority of adrenal metastases.[1,4]

The incidence of adrenocortical carcinoma is estimated to be 0.4/100,000. Adrenocortical carcinoma increases with tumor size to 25/100,000 (median diameter >6 cm).[4] Bilateral manifestations of adrenocortical carcinoma occur in only 2-6% of the cases reported and are associated with Li-Fraumeni syndrome, type I multiple endocrine neoplasia, Beckwith-Wiedemann syndrome, and Carney complex, principally in children as shown in **table 1.**

Table 1. Syndromes associated with Adrenocortical carcinoma

Syndromes	Genetic Changes	Characteristics
Li Fraumeni syndrome	CHECK2 & TP53	Breast cancer, osteosarcoma, soft tissue sarcoma, leukemia and Adrenocortical carcinoma
MEN 1 syndrome	MEN 1 gene	Hyperparathyroidism, pancreatic islets cell tumors, pituitary tumors, Adrenal tumors
Beckwith-Wiedemann syndrome	IGF-2 gene (11p15)	Macroglossia, macrosomia, midline abdominal wall defect, hepatoblastoma, Wilm's tumor, ACC, thyroid carcinoma
Carney complex syndrome	PRKA-R1A gene	Spotty skin pigmentation, cardiac myxomas, ACC, Cushing's syndrome

In contrast to our patient, many patients with adrenocortical carcinomas present with clinical symptoms of endocrine excess. Indeed, hormone-functioning tumors account for 26% to 94% of adrenocortical carcinomas[3,4] Most patients with adrenocortical carcinomas are diagnosed at an advanced stage of disease with large primary tumors (median tumor size at diagnosis, >10 cm) and invasion to adjacent organs. The main clinical symptoms, such as abdominal discomfort or back pain, are related to the mass effect of a large tumor.[3,4]

All adrenal tumors detected have to be diagnosed for malignancy potential and hormonal activity to render timely and curative treatment. Imaging studies using CT, MRI, and FDG-PET to demonstrate adrenal mass size and appearance have been used to distinguish between benign and malignant lesions. Differentiation between malignant and benign adrenal lesions can be performed by using 18-FDG-PET with >95% accuracy.[4,5] The Weiss criteria, developed in 1984, were established to distinguish benign from malignant adrenal tumors using nine pathologic features. The classification system is based on tumor structure, cytology, and invasion as shown in **box 1.** The presence of three or more of the

Box 1. Weiss pathologic criteria of differentiating benign and malignant adrenal tumour

High nuclear grade (Furman grade 3 to 4)
High mitotic rate (greater than 5 mitoses per high-power field)
Presence of atypical mitoses
Character of cytoplasm (low percentage of clear cells)
Diffuse architecture of tumor cells
Presence of necrosis
Invasion of venous structures
Invasion of sinusoidal structures
Invasion of tumor capsule,
High nuclear grade (Furman gr

Weiss criteria is associated with malignancy, with a sensitivity of 100% and a specificity of 96%. In addition, 18-FDG-PET plays an important role in evaluating treatment response and residual masses.

In general, surgery involving adrenal tumors should be considered in patients with functioning cortical tumors and clinical symptoms.[4,6,7] Regarding nonfunctioning tumors, recommendations regarding treatment mainly refer to the tumor size. In general, clinically silent lesions <3 cm without any criteria of malignancy are not resected and should be followed closely by CT or MRI scans every 6 or 12 months.[4,6,7] The indications for an adrenalectomy are a definitive or presumed diagnosis of primary adrenocortical carcinoma and circumstances technically obstructive to a minimally invasive approach. In the case of any intraoperative features of malignancy, conversion to an open approach should be performed to enable extensive radical compartment resection.[3,6,7]

The differentiation between benign and malignant adrenal lesions is based on macroscopic and microscopic features.[3,8] The Weiss score is the most widely used classification for microscopic characteristics suggestive of a malignant tumor. Three or more histologic criteria are necessary to establish the diagnosis of adrenal carcinoma.[9]

END NOTE

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Conflict of Interest: None declared

Editor's Remarks: These are rare but highly malignant extensive tumours that have a variety of clinical and radiological findings. The case report is published so that its clinical presentation is familiar to all clinicians.

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