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Adult Adrenocortical Carcinoma Mass with Extension into the Inferior Vena Cava and Cushingoid Features: a Rare Case with Review of Literature

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ABSTRACT

Adrenocortical carcinoma (ACC) is an uncommon disease that accounts for only 0.02% of all cancers. Herein, we report a case of adult ACC with extension into the inferior vena cava and Cushingoid features. A 67-year-old woman was admitted to the surgery service for surgical resection of adrenal mass which was found on hypertension work-up. On physical examination, she was obese and cushingoid with ecchymoses on her skin. The patient was hypokalemic at the time of admission. Spiral CT with contrast of the abdomen and pelvis was done and showed solid mass measuring 10x11 cm, superior to the right kidney and posterior to IVC (inferior vena cava) with anterior displacement of IVC. Microscopic diagnosis was adrenocortical carcinoma with vascular invasion and necrosis (20% of surface area). In conclusion, hypertension can be the most common complaint in ACC patients and the presence of both hypertension and hypokalemia in a case can probably increase suspicion of ACC. Future studies are required to address these complaints.

Keywords: Adrenocortical carcinoma , Cushingoid, Inferior vena cava

INTRODUCTION

Adrenocortical carcinomas (ACC) are uncommon adrenal cortex tumors that follow an aggressive clinical course[1]. ACC can be functional or nonfunctional with regard to hormone synthesis and clinical features[2]. About 60% of ACCs secrete hormones (functional ACC) and the steroid profile often displays a wide variety of steroids in ACCs, which may be used as tumor markers to detect metastastic disease[3]. Patients with functional ACC can present with Cushing's syndrome[4]. The non-functioning tumors may present with pain or fullness in the abdomen due to mass effect, weight loss, weakness, fever and myalgias[5]. The distinction between ACC tumors and other cancers may sometimes prove difficult due to overlapping clinical, morphologic and even immunophenotypical features[1]. The aim of this study was to evaluate a case of adult ACC with extension into the inferior vena cava and Cushingoid features in West of Iran.

CASE REPORT

A 67-year-old woman was admitted to the surgery service on April 30, 2016 for surgical resection of adrenal mass which was found during hypertension work-up. Her past medical history included uncontrolled hypertension since last year with drug history of methyl dopa, metoral, hydrochlorothiazide, labetalol, spironolactone and furosemide. She had undergone an unidentified eye surgery seven years ago. On physical examination, she was obese and cushingoid with ecchymoses on her skin. The patient was hypokalemic and hypertensive at the time of admission. The lab data were: potassium (K) =2.6 (reference range=3.8-5 mEq/L), urinary free cortisol =578 (ref=110-138 nmol/24h), urine metanephrine =145 (ref=74-297 µg/24h) and urine vanillyl mandelic acid (VMA) =4 (ref=1.5-7.5 mg/24h). Sonography on March 3 demonstrated a hyperechoic well-demarcated mass measuring 9.7x11.2 cm adjacent to the right kidney with internal flow. A heteroechoic well-demarcated lesion was noted in the right hepatic lobe, measuring 58x55 mm, suggesting an adrenal origin with recommendation of abdomino-pelvic CT scan with contrast.

Spiral CT with contrast of the abdomen and pelvis was done on March 6 and showed a solid mass measuring 10x11 cm, superior to the right kidney and posterior to IVC (inferior vena cava) with anterior displacement of IVC. It seemed attached to the right liver lobe and upper right renal pole. The mass was heterogenous with probable origin from the right adrenal gland (carcinoma or pheochromocytoma). Right kidney was displaced inferiorly. A few tiny stones were observed in lower calyces of kidneys. Lymphadenopathy was not present. On May 2, right adrenalectomy was performed. The pathologist reported a piece of encapsulated tan/brown mass measuring 7x10x11 cm with creamy hemorrhagic cut surface compatible with ACC (**Figure 1**). There were vascular invasion and necrosis (20% of surface area). Immunohistochemical (IHC) staining for inhibin (**Figure 2**), melan-A (**Figure 3**) and synaptophysin were positive, whereas chromogranin and epithelial membrane antigen (EMA) were negative. Furthermore, cytokeratin (CK) showed focal positivity in favor of diagnosis.

DISCUSSION

We report an adult ACC attached to kidney and liver with extension into the IVC. ACC is an uncommon disease that accounts for only 0.02% of all malignant tumors[6]. The IHC results (positive reactions for vimentin, CD56, inhibin, melan-A, synaptophysin, Bcl-2, calretinin) confirm the diagnosis of ACC[7]. One study on 40 ACC patients showed that positive staining was observed for inhibin-α (92.5%), synaptophysin (72.5%), melan-A (65%), cytokeratin (22.5%) and chromogranin A (5%)[1];in our case,

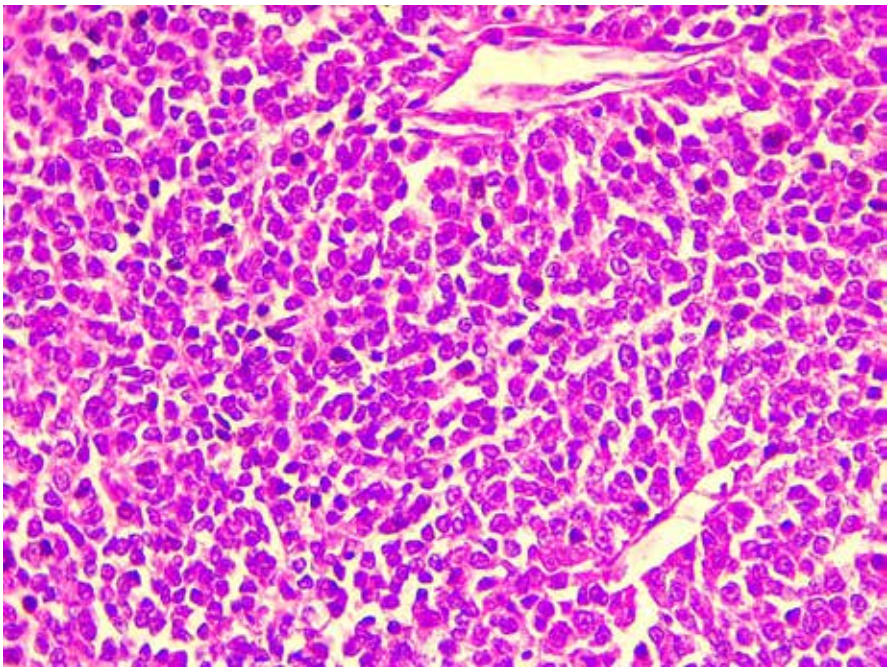

Figure 1: Hematoxylin and eosin staining, Adrenocortical carcinoma x400

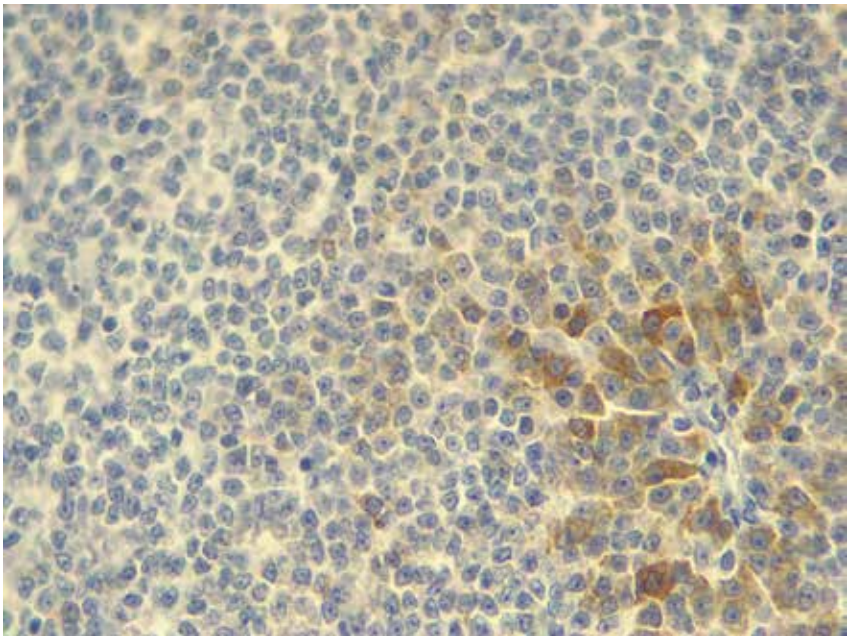

Figure 2: Immunohistochemical staining, Inhibin x400

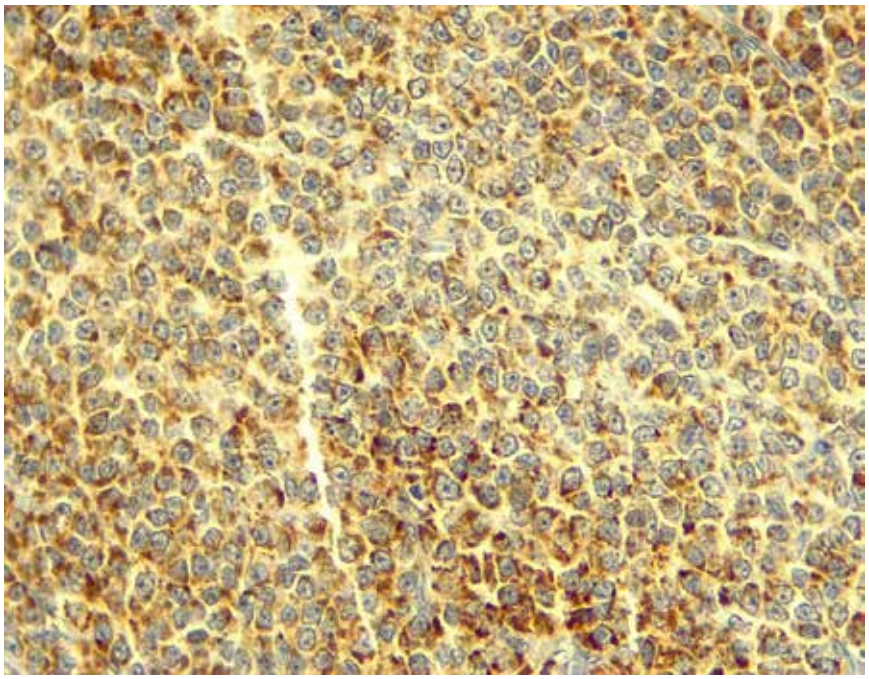

Figure 3: Immunohistochemical staining, Melan-A x400

the first four of these markers were positive while the fifth was negative. **Table 1** shows a number of cases with adult ACC. The prevalence of ACC covers both genders and sides and all ages. Therefore, ACC can develop at any age in either sex[8]. The most common complaint on the first visit was hypertension; out of seven cases with reported K levels, four had hypokalemia. Therefore, attention to hypertension and hypokalemia can be helpful in the primary diagnosis of ACC. About 60% of functional ACC tumors may secrete hormones such as excessive glucocorticoids (Cushing's syndrome), androgens (virilizing tumor) or estrogens (feminizing tumor)[9]. Aparna et al.,[10] similar to our case, reported features of Cushing's syndrome in their cases. Cushingoid features include central obesity, hoarseness, hirsutism, and hypertension[10]. Extension of adrenal cortical carcinomas into the IVC is rare. Similar to our case, four cases[7,8,11,12] have been reported with extension to the IVC.

CONCLUSIONS

Our study presented a rare case of ACC with extension into the IVC and Cushingoid features. Hypertension can be the most common complaint in ACC patients and the presence of both hypertension and hypokalemia in a case can probably increase suspicion of ACC; future studies are required to address these complaints.

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Table 1: Reported cases with adult adrenocortical carcinoma

Reference	Sex	Age	Hyper-tension	Hypo-kalemia	Other clinical signs	Lat-erality	Tumor size (cm)
2	M	69	Yes	-	Chest pain	Right	10
3	F	59	Yes	Yes	-	Right	6.5
4	M	61	No	No	Dyspepsia	Left	9
7	M	27	-	-	Diabetes, Intensifying stomach fullness, pain, nausea	Left	17 × 12
8	F	22	Yes	Yes	Loss of consciousness, Weakness, Tachycardia	Right	6×5.5×1.5
10	F	22	Yes	-	Abdominal pain, Lump, Seizures, Central obesity, Hoarseness, Hirsutism	Right	9×8
11	M	62	-	-	High grade fever, Weight loss, Lumbago	Left	-
12	M	33	-	-	Gynecomastia, Abdominal distention	Right	18×15
13	F	47	No	No	Left lower back pain	Left	13.5 × 8.6 × 4.2
14	F	30	-	-	Left flank pain, Lower urinary tract symptoms or gross hematuria	Left	17 × 13 × 12
15	F	54	Yes	-	Bilateral pitting leg edema, Diabetes, Hyperlipidaemia, Rheumatoid arthritis	Both	3×5
16	F	28	Yes	-	Palpitation, excessive sweating, flushing, cold extremities, dizziness	Right	16.5 × 6.5 × 8.7
17	M	34	Yes	-	Headache, chest pressure	Both	Right: 4.3 Left: 2.8
18	M	54	No	-	Swelling, Left flank pain, Weakness, Loss of appetite, Pallor, Edema feet	Left	20 x 14 x 10
19	F	36	-	-	Weight loss	Left	12×10.3×9.7
20	M	41	-	-	Fading, Pale, Hemorrhagic shock due to spontaneous rupture, Intense loin pain	Left	8
21	M	43	Yes	-	Abdominal pain, Vomiting	Right	9.9x 6.8
22	M	70	Yes	Yes	Generalized weakness, moon face, central obesity	Right	10
23	M	34	-	-	Abdominal pain	Right	-
24	F	51	-	No	Dyspepsia, Ascites, Peripheral edema	Right	8
The present case	F	67	Yes	Yes	Ecchymoses on her skin	Right	10x11

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