An Interesting Case of Hepatic Adrenocortical Carcinoma

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Figure 1.

Figure 2.

Figure 3.
Adrenocortical carcinoma (ACC) is a rare solid tumor with an incidence of 0.5 to 2 cases per million per year. It affects women more commonly than men with a ratio of 1.5:1. Ectopic ACC are considered to be extremely rare with no exact incidence data yet.\textsuperscript{1-6} We report an interesting case of hepatic ACC in a young woman with clinical signs of virilization.

A 21-year old Sundanese woman visited our endocrine clinic with progressive hirsutism over the face, body, and extremities starting 14 years previously. She had irregular, heavy periods when she was 7 years old. She also experienced pubertal development of her breasts. However, both menstrual cycle and breast development ceased when she was 8 years old. She noticed voice deepening and alopecia. Physical examination showed male-type alopecia and intense hirsutism. Tanner stage was 3 for breast tissue and 5 for pubic hair. (\textbf{Figure 1}) There was no galactorrhea. Body mass index was 21.4 kg/m\textsuperscript{2}. Hormonal evaluation revealed increased level of free testosterone (>1500 ng/dl; NV: 8.4-48.1 ng/dl), dehydroepiandrosterone sulfate (>1000 ug/ml; NV: 65.1-369 ug/ml), and estradiol (533.60 pg/ml; NV: 14-124 pg/ml), low level of LH (<0.07 mIU/ml; NV: 1.7-11 mIU/ml) and FSH (<0.30 mIU/ml; NV: 1.34-9.40 mIU/ml), slight increased in morning serum cortisol (26.61 ug/ml; NV: 4.3-22.4 ug/ml), normal serum thyroid stimulating hormone (3.2 mIU/l; NV: 0.34-4.25 mIU/l) and prolactin (14.70 ng/ml; NV: 3.30-15.80 ng/ml). Gynecological ultrasound and brain MRI examination showed no structural abnormality. Abdominal CT scan demonstrated contrast enhanced solid inhomogenous mass sized 11.6 x 14.2 x 15.6 cm in right liver lobe. (\textbf{Figure 2}) Neither suprarenal mass nor paraaortic lymphadenopathy was seen in the abdominal CT scan. Chromosomal examination revealed normal female karyotype (46, XX). Further liver biopsy showed morphology and immunohistochemistry (positive for CD 56, HEP 1, and NSE) consistent with adrenocortical carcinoma. (\textbf{Figure 3}) Surgical therapy with referral to other institution was offered to the patient as first line treatment. Meanwhile, the patient got spironolactone 100 mg OD.

Virilizing tumors are rare and few of them are androgen-producing adrenal tumors. Ectopic adrenal tumors are even rarer. Ectopic adrenal tissue can be found close to the adrenal glands, or along the path of descent or in association with gonad. Moreover, they have ever been reported in nervous system, stomach, gall bladder, and liver.\textsuperscript{4,5,7-9} There have been several case reports of adrenal rest tumor of liver; however, our literature review found no report of ectopic ACC of liver.\textsuperscript{10-12} Cortical tissue, embrologically derived from mesoderm, seems to be the sole component of the tumor which can undergo malignant transformation or become hormonally functional. Functioning tumors are more frequent in women. Our case demonstrated virilization as chief complaint. The ectopically located functioning tumors display the same clinical picture as tumors located in adrenal gland, with Cushing’s syndrome and virilization are the most frequent symptoms in order of frequency.\textsuperscript{11-14} The virilization, as shown in our case, is due to excessive androgen production of dehydroepiandrosterone sulfate and testosterone. The distinction of ACC from benign adrenocortical tumor is important. Since there is no previous report of hepatic ACC and surgery is the keystone of curative treatment modality for ACC at adrenal gland, we planned the patient for surgical resection.\textsuperscript{3,15,16} Adjuvant treatment with chemotherapy (mitotane and combination of cytotoxic drugs), irradiation might be considered in ACC treatment.\textsuperscript{2,3} To control androgen effects, spironolactone was administered in our patient. However, there was no significant improvement in symptoms.\textsuperscript{2,17}

In conclusion, we present the first reported case of hepatic ACC. A thorough history, physical examination, and appropriate laboratory, imaging examination are critical in evaluating virilized female patients. Elevated serum concentration of dehydroepiandrosterone sulfate and testosterone might direct clinician to functioning adrenal cortical tissue as etiology, with further investigation of exact tumor site.

\textbf{ETHICAL STATEMENT}

Informed consent was obtained from patient and family prior to publication of this case and accompanying image.
REFERENCES