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A Rare Feminizing Adrenocortical Carcinoma: A Case Report

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ABSTRACT

Adrenocortical carcinoma (ACC) is a rare tumor that accounts for only 0.02% of all cancers. About 60% of patients present with symptoms of excessive hormone production. However, ACC is usually nonfunctional when it occurs in adults. Its presentation with only virilizing symptoms is extremely rare. We report a case of a huge functional, virilizing ACC of the right adrenal gland that measured 14 cm by 9 cm by 6 cm and weighed 600 gm, which was successfully extirpated.

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare tumor that accounts for only 0.02% of all cancers [1]. About 60% of patients present with symptoms of excessive hormone production. However, ACC is usually nonfunctional when it occurs in adults. Its presentation with virilizing symptoms is extremely rare. We herein report a case of a huge functional, virilizing ACC of the right adrenal gland that measured 14 cm by 9 cm by 6 cm and weighed 600 gm, which was successfully extirpated.

CASE REPORT

An 18-year-old male presented with complaints of dull, vague abdominal pain localized to the right upper quadrant for a 6-month duration. The pain was not associated with vomiting, fever, or any other systemic complaint. Other complaints were bilateral breast enlargement, striae over axilla, and breast, thigh, and facial puffiness for 4 years. A physical examination revealed hypertension with a typical Cushingoid appearance, including a plethoric moon face, truncal and centripetal obesity, and abdominal cutaneous striae (Figure 1).

An abdominal examination revealed a large lump of about 10 cm by 15 cm located in the right hypochondrium. It was smooth, firm to hard in consistency, and was not moving with respiration. Laboratory testing revealed no anemia, normal white blood cell counts, and normal differential counts. His fasting blood sugar was 98 mg/dL, and his serum creatinine 1.2 mg/dl, with normal serum electrolytes. Liver function tests were normal. Urinalysis was normal. Ultrasonography revealed a solid mass at the upper pole of the right kidney. The chest radiograph did not reveal any abnormality. Magnetic Resonance Imaging (MRI) of the abdomen showed the organ of origin was the right adrenal gland, which was heterogenous with hemorrhagic areas within (Figure 2).

The liver and the opposite adrenal were normal. Functioning of both kidneys was normal. His serum cortisol (42.08) and DHEA (832.7) levels were elevated. A 24-hour urinary catecholamine

KEYWORDS: Adrenocortical carcinoma, functional, virilizing

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case report
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Figure 1. Physical appearance at presentation.
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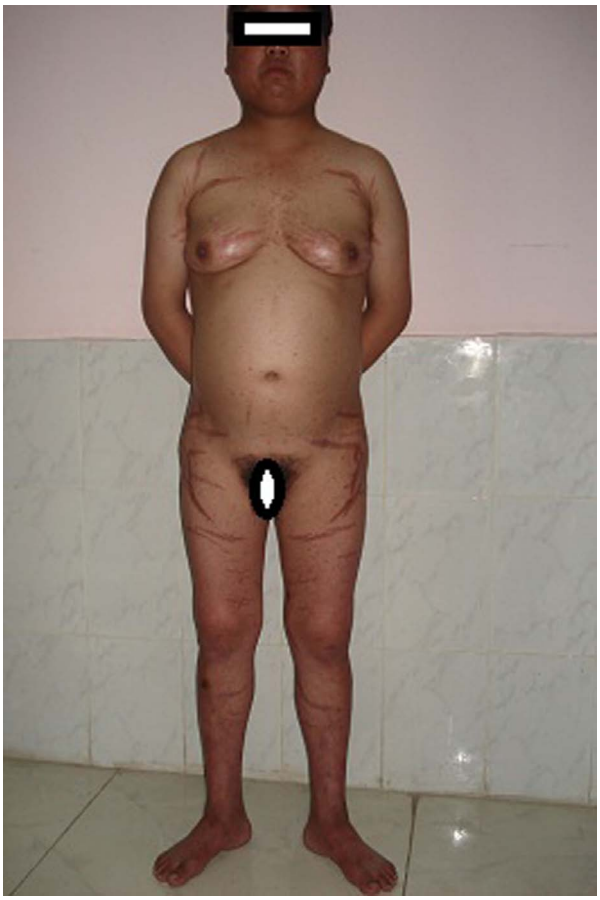

was normal. The patient was subjected to right radical adrenalectomy. The right kidney could be preserved. The tumor mass was abutting the IVC and there was involvement of the posterior surface of the liver. Histopathological examinations of the excised specimen confirmed the diagnosis of adrenocortical carcinoma (Figure 3) with a mitotic figure of 8-9/50hpf, an extracapsular extension, lymphovascular invasion, positive peripheral margins with tumor necrosis, cystic degeneration hemorrhage, and focal calcification. Postoperative recovery was uneventful. The Cushingoid features improved gradually within 3 months after the surgery. The patient is alive with no metastases 1 year after the surgery. His blood pressure and electrolyte levels remain normal.

DISCUSSION

ACC is a rare, aggressive tumor that affects only 1 to 2 persons per million [1]. It represents 0.02% of all cancers and has a poor prognosis, causing 0.2% of all cancer deaths. Most series show a female preponderance, but our case is an 18-year-old male. ACC can be functional or nonfunctional, depending on the production of corticosteroids, androgen, estrogen, or mineralocorticoids by the tumor [2]. The functioning tumors may secrete excessive glucocorticoids (Cushing's syndrome), mineralocorticoids and their precursors, including androgens (virilization) and/or estrogens (feminization), as well as aldosterone causing hypertension as in our case which had elevated cortisol and DHEA. Most of the carcinomas (60%) are functional, and the first manifestation is usually Cushing's syndrome. They are typically large, with imaging features of

Figure 2. An MRI showing a right adrenal mass.
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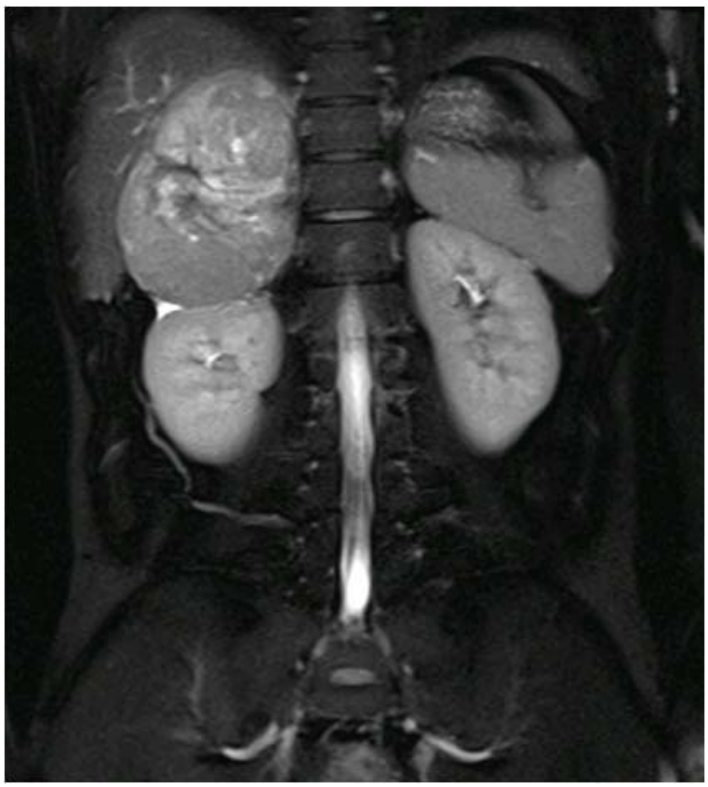

Figure 3. Microsections showing adrenocortical carcinoma.
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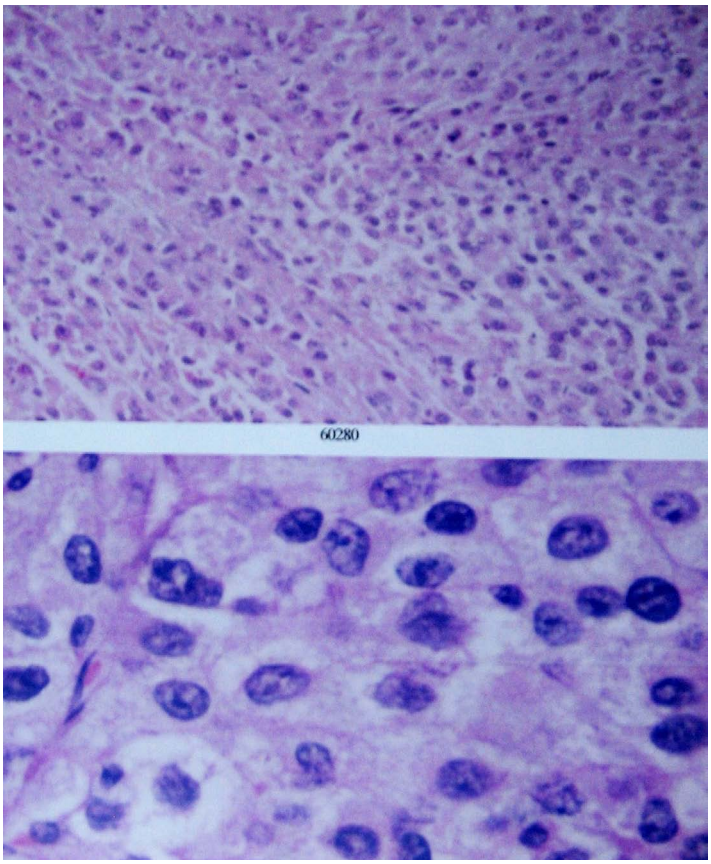

malignancy, including heterogeneous density, central necrosis, and calcification requiring histological evaluation. They can also be biochemically active, prompting imaging evaluation for mass detection.

Estrogen-secreting tumors, which can cause feminization, are rare. However, ACC with virilizing syndrome is a rare presentation that was seen in our case, and by looking at the presentation of the patient, combined with biochemical and imaging studies, ACC becomes the most suitable diagnosis without any differentials. Prompt regression of Cushingoid fascia and cutaneous manifestation were observed after the tumor extirpation. Such a large mass should be closely followed over a lifetime as chances of recurrence, after many years of treatment, are very high due to its characteristics. Mitotane (adrenocortical cytotoxin) is usually reserved for these cases and also for inoperable ones. There may be local tumor invasion, including the potential for tumor thrombus formation, which can embolize, or hormone excess syndromes and paraneoplastic syndromes upon follow-up.

The detection of tumors at an early clinical stage is crucial for curative resection, and total resection offers the only prospect for a cure. Patients with functional ACC may have a better prognosis because they present earlier, unlike patients with nonfunctional variants who invariably present when the tumors are very large or are associated with distant metastasis. Estimates of the overall 5-year survival rate are approximately 20 to 35%. For cases where total surgical resection is achieved, this rate is estimated to be approximately 32 to 47%. In those cases where total surgical extirpation has not been possible, the 5-year survival rates are 10 to 30%. Even after apparently complete surgical resection, local or distant relapse occurs in nearly 80% of cases. Patients who show no response to mitotane or who relapse are probably best served by a referral to a major cancer center where they can be enrolled in one of several ongoing combination chemotherapeutic/radiation and/or surgical resection protocols. The size of the adrenal tumor on CT or MRI is considered the best indicator of malignancy [3]. This case is reported due to the rarity of its presenting features.

REFERENCES

1. Norton, J.A. (2005). "Adrenal tumors." In: DeVita, V.T., Jr., Hellman, S., Rosenberg, S.A., eds. *Cancer: Principles and Practice of Oncology*, 7th ed. Philadelphia, PA: Lippincott Williams & Wilkins,1528-1539.

2. Schulick, R.D. and M.F. Brennan. (1999). "Adrenocortical carcinoma." *World J Urol* 17:26-34. PubMed ; CrossRef

3. Lal, G. and O.H. Clark. (2005). "Thyroid, parathyroid, and adrenal." In: Brunicardi, F.C., Andersen, D.K., Billiar, T.R., et al., eds. *Schwartz's Principles of Surgery*, 8th ed. New York, NY: McGraw-Hill,1458.