

Virilizing Adrenal Cancer and Bail-out Nephrectomy

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ABSTRACT

We report a rare case of virilizing adrenal cancer with tumorous invasion of the left renal vein in which a retroperitoneal adrenalectomy with bail-out nephrectomy was performed. A tumor thrombus infiltrated the wall of the left adrenal vein and extended into the left renal vein. Initially, a kidney sparing procedure with partial tangential excision of the involved renal vein wall was performed. After positive vein margins were confirmed with intraoperative histology, the indication for nephrectomy was made. To the authors' awareness, this is the first report of a virilizing adrenal cancer with a tumor thrombus infiltration of the renal vein and surgical tendency for kidney preservation.

Key words: adrenal gland, tumor, thrombosis, virilization

Introduction

Malignant tumors of the adrenal cortex account for 0.05–0.2% of all cancers, with an approximate prevalence of two new cases per million per year in the general population^{1,2}. Hormonally active adrenocortical cancers are present in 30–40% of patients, with Cushing's syndrome being their most frequent clinical presentation³. Virilization is very rare and occurs in 6–8% of adults with adrenocortical carcinoma. Preoperative recognition of a venous tumor extension is of considerable importance in planning the surgical procedure. A poor prognosis in this neoplasm most frequently affecting young people makes its early detection pivotal, as the disease is surgically curable only at early stages.

Case Report

A 35-year-old female, mother of two healthy children, presented with a history of progressive weakness over the previous six months and oligomenorrhea. Excessive hirsutism on the patient's face, neck and shoulders, delicate maculopapulous rash on the facial skin and a moon face were observed. Abdominal and gynecological examinations did not yield any abnormalities. The patient was normotensive and normoglycemic. Her renal biochemical parameters were within normal ranges. ACTH was 8 ng/L (normal range: 10–100 ng/L), urinary 17-ketosteroids 83.5–119.5 $\mu\text{mol/d}$ (normal range: 17–52

$\mu\text{mol/d}$), androstendion 96.05 nmol/L (normal range: 2.1–6.4 nmol/L) and serum testosterone 12.1 nmol/L (normal range: 0.14–2.8 nmol/L). Urinary vanillylmandelic acid and aldosterone were within normal limits. Abdominal ultrasonography revealed a solid mass growing from the upper pole of the left kidney. Computerized tomography (CT) scan of the abdomen showed a 68x101 mm left adrenal expansion with a tumor thrombus extension through the adrenal vein into the left renal vein. The thrombus did not invade the inferior caval vein or induce retroperitoneal lymphadenopathy (Figure 1). Opposite adrenal gland was hormonally suppressed and hypoplastic on CT scan. Lumbotomy through 10th intercostal space and extra peritoneal extirpation of adrenal gland with tumor and tumor thrombus were undertaken. The renal hylus was clamped, the renal vein incised and its wall partially excised. The tumor thrombus was completely removed en block with tumor (Figure 2). The renal vein was sutured in warm ischemia lasting 7 minutes. Kidney reperfusion was excellent. As intraoperative histology examination revealed that the tumor infiltrated the resected renal vein margins, nephrectomy was performed. Pathologically, the cancer was found to be adrenal cortical with the endocrine tumor cell differentiation. Similar tumor cells were also found in renal vein margins. Kidney and lymph nodes were tumor free. Serum testosterone level fell to its normal value within 20 days after the surgery. Glucocorticoids

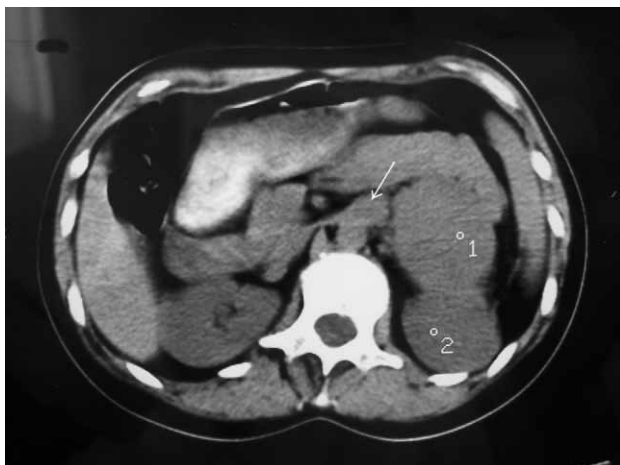


Fig. 1. Computer tomography (CT) scan. Large left adrenal tumor (1) with the arrow pointing on the tumor thrombus and the kidney (2).

were gradually withdrawn over the first postoperative month. The first postoperative abdominal CT was done three months after the surgery and was normal. Seven months after the surgery, the patient's serum testosterone values rose again. A control CT scan revealed multiple hepatic metastases and retro peritoneal lymphadenopathy. Mitotane therapy was started. The patient died 13 months after the surgery.

Discussion

Virilizing adrenal cancer is extremely rare tumor with a high malignant potential. In the present case, the authors' view was that a thrombotic tumor extension into the renal vein was not necessarily associated with nephrectomy. However, intraoperative histological examination after a successful kidney revascularization presented an indication for nephrectomy. Reviewing the literature reports on 12 women with virilizing adrenal cancer, none of them was found with a renal vein tumor thrombus. A positive dexamethasone test is useful in de-

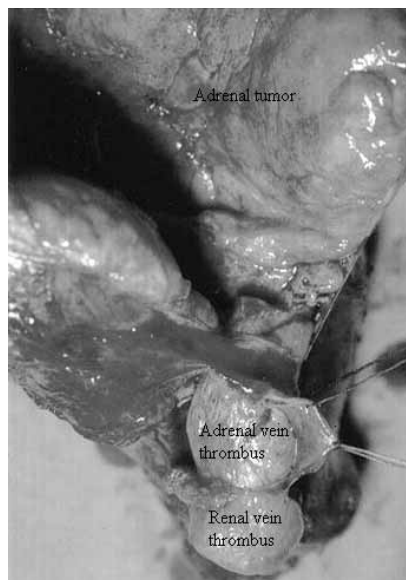


Fig. 2. Adrenal tumor with the tumor thrombus extension and infiltration of adrenal vein (on surgical sutures).

tection of virilizing adrenal cancer³ during the preoperative evaluation of hirsute women with serum cortisol, testosterone and androstendion elevations. Among current radiological diagnostic methods, MRI distinguishes among adrenocortical tumors the most accurately; it compares the ratio of individual signal intensities of each type of the adrenal mass to that of the liver⁴, and provides information about vascular invasion of the tumor. In the present case, CT scanning was used in diagnostic blood vessel evaluation. In our patient, a radical surgery could not prevent vascular spread of the disease into the liver. Mitotane has been used in altering extratumoral cortisol and androgens metabolism. In this patient, it proved ineffective in prolonging survival. Various studies combining mitotane and cytotoxic chemotherapy did not show significant prolongation of survival^{5–7}. Therefore, in patients with adrenal neoplasms, surgical removal of the tumor at early stages gives these patients the only chance for survival.

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VIRILIZIRAJUĆI KARCINOM NADBUBREŽNE ŽLIJEZDE I NEIZBJEŽNA NEFREKTOMIJA

S A Ž E T A K

U ovom radu je prikazan vrlo rijedak slučaj virilizirajućeg karcinoma nadbubrežne žlijezde s invazijom tumora u lijevu renalnu venu, kod kojega je učinjena adrenalectomija s neizbježnom nefrektomijom. Tumorski tromb je infiltrirao stjenku lijeve nadbubrežne vene i proširio se u lijevu bubrežnu venu. Isprva je učinjena poštedna operacija s prezervacijom lijevog bubrega na kojem je učinjena tangencijalna ekscizija zahvaćenog dijela stjenke bubrežne vene. Indikacija za nefrektomiju je postavljena nakon učinjene intraoperativne patohistološke analize, kada je potvrđen pozitivan venski rub. Prema saznanjima autora, ovo je prvo izvješće o virilizirajućem tumoru nadbubrežne žlijezde s tumorskom infiltracijom bubrežne vene, kod kojeg se nastojao sačuvati bubreg.