A case of a testosterone-secreting oncocytic adrenocortical carcinoma

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Abstract

Oncocytic neoplasms are most rarely found in the adrenal gland. They are usually benign and non-functioning. We present a case of a testosterone-secreting oncocytic adrenocortical carcinoma in a 19-year-old female who presented with acne, hirsutism and irregular menses. Clinical investigations revealed an elevated testosterone and DHEA-S and a 4x5 cm left adrenal mass. The tumour was successfully excised. The histology showed the tumour to be comprised of oncocytic cells with granular, eosinophilic cytoplasm, features consistent with an oncocytic carcinoma. This is the first case presented of a testosterone-secreting oncocytic adrenocortical carcinoma.

Oncocytomas are neoplasms that are characterised histologically by the appearance of epithelial cells with an abundant eosinophilic, granular cytoplasm. Most oncocytic neoplasms are non-functioning, however there have been seven reported cases of functioning oncocytic adrenocortical neoplasms.\textsuperscript{2,5–7,9,10} We report a case of a functioning oncocytic adrenocortical carcinoma in a 19-year-old female.

Case report

A 19-year-old female presented with recent onset hirsutism, acne affecting the face and shoulders, and irregular menses after stopping the oral contraceptive pill.

Biochemistry revealed an elevated testosterone of 12 nmol/L (0.5–2.7) and DHEA-S of 17.5 µmol/L (0.5–12). A dexamethasone suppression test revealed a 0800 plasma cortisol of 108 nmol/L (<50 nmol/L) indicating incomplete suppression. She had normal post synacthen 17-hydroxypregesterone, electrolytes, creatinine, liver function tests, urinary catecholamines and preoperative 24-hour urinary cortisol level.

Abdominal ultrasound followed by CT scan, revealed a five-by-four centimetre left adrenal mass with central necrosis but no evidence of invasion into surrounding structures or lymphadenopathy.

The patient underwent a laparoscopic converted to open left adrenalectomy. There was no evidence of invasion into surrounding structures. Postoperative recovery was uneventful.

Six weeks postoperatively, the serum testosterone and DHEA-S levels had both returned to normal and she reported improvements in her acne, reduced hair growth and the return of regular menses. Follow-up chest/abdomen/pelvis CT scans performed at 3 and 9 months postoperatively showed no evidence of tumour recurrence. A repeat dexamethasone suppression test was also normal 9 months post discharge.
The mass weighed 67 g. Histological findings included oncocytic cells with granular, eosinophilic cytoplasm, highly pleomorphic nuclei and infrequent tumour mitoses. There was necrosis but no evidence of vascular or capsular invasion.

Based on the histological findings, the specimen fulfilled four of the Weiss criteria and therefore was a malignant adrenocortical neoplasm. The specimen was sent for a second opinion in Sydney, Australia and was confirmed to be an adrenal cortical carcinoma by means of advanced immunohistological staining using IGF2.

**Discussion**

Oncocytic neoplasms are most commonly found in the kidney, thyroid and salivary glands but are rare in other sites.

We have identified 42 cases of adrenal oncocytic neoplasms in the English literature comprising 18 cases of adrenocortical oncocytoma, 17 cases of oncocytic adrenocortical carcinoma and 7 cases of adrenocortical oncocytoma of unknown malignant potential (UMP).\(^1\)\(^{-10}\)

Of the 17 cases of malignant oncocytic neoplasm arising in the adrenal gland, the average tumour size was 11.9 cm with a mean weight of 628.4 g. The average age at diagnosis was 52 years and there were 8 male cases and 9 female cases.

Of the 14 cases that included information regarding follow-up,\(^2\)\(^,\)\(^3\)\(^,\)\(^4\)\(^,\)\(^10\) 7 had no evidence of recurrence, 6 had documented recurrence at an average of 26 months following surgery, and 1 succumbed to their disease at 58 months.

There have been three previously reported cases of tumour functionality in oncocytic adrenocortical carcinomas. They included two cases of cortisol secretion alone\(^2\)\(^,\)\(^9\) and a case of co-secretion of cortisol and aldosterone.\(^10\) Our case is the first that we are aware of involving elevated serum testosterone and DHEA-S in an oncocytic adrenocortical carcinoma.

The main histological criteria adopted for the differentiation of adrenal oncocytic neoplasms is the Weiss criteria. Its accuracy has been debated recently and Lin et al have proposed a modified system for use in oncocytic neoplasms.\(^1\)

In summary, we have presented a unique case of a functioning oncocytic adrenocortical carcinoma associated with elevated testosterone and DHEA-S levels. To our knowledge this is the only such case reported to date in the English literature. Following surgery, the patient’s clinical signs and symptoms resolved and the serum testosterone and DHEA-S levels returned to normal.

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