Adrenal Oncocytoma – A Rare Functional Tumor Presenting as Cushing Syndrome

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Abstract
Adrenal oncocytoma is very rare pathological variant of adrenal neoplasm. These are usually large and non-functional; however, rarely functional adrenal oncocytomas are also presented as Cushing's syndrome and pheochromocytoma. We report a case of adrenal oncocytoma in 38 year old female presented with symptoms of Cushing Syndrome.

Introduction
Oncocytomas are epithelial tumors composed of cells with abundant eosinophilic granular cytoplasm packed with mitochondria. Sites of origin for oncocytoma include kidney, salivary glands, parathyroid, lung, pituitary and ovary. However, the oncocytoma of the adrenal gland is very rare. There are nearly 50 case reports of adrenal oncocytoma in English literature. These are usually large and non-functional; however, rarely functional adrenal oncocytomas presenting as Cushing’s syndrome and pheochromocytoma have also been reported. We describe a case of 38 year old female with symptoms of Cushing syndrome, treated with laparoscopic adrenalectomy, histologically proved to be adrenocortical oncocytoma with immunohistochemistry marker positive for the same.

Case Report
38 year old female, married since 24 years, no co morbidities, presented with chief complaints of generalized swelling all over body and puffiness of face since 6 months, shortness of breath since 4 months, generalized weakness and palpitation since 2 months. She had noticed swelling on limbs (lower limbs> upper limbs) and then facial puffiness. She gives h/o oligomenorrrhoea since 6 months.

On examination, she was well nourished and adequately built. She was afebrile, pulse was 98/minute, and blood pressure was 150/90mm of Hg in right brachial artery. She had generalized edema, jugular venous pressure normal; she had violaceous striae on trunk, medial side of thighs, hyper pigmented knuckles, elbow and posterior shoulder blades.

On investigation, serum cortisol level at 8am-3.2mcg/dl (3.7-19.4), urine cortisol level per day- 508.25mcg/day (58-403), plasma free metanephrine level- 39.20pg/ml normal, plasma ACTH- 43.4 pg/ml which is normal.

CT scan of Abdomen and pelvis was done which revealed well defined 2.5 X 1.8cms mass lesion seen in the left adrenal gland, the lesion was solid and demonstrated enhancement. CT Renal Angiography was done was s/o there is evidence of well defined, solid, rounded lesion seen involving the left adrenal gland, measuring 2.3 X 2.2cms, shows significant enhancement and washout on the post contrast study. The relative and absolute enhancement is more than 70% suggestive of an adrenal adenoma.

Patient underwent left laparoscopic adrenalectomy, adrenal mass being well encapsulated weighing 6gm histologically proved to be adrenocortical oncocytoma, microscopic features of well encapsulated tumor comprised of solid sheets of polygonal cells with abundant granular, eosinophilic cytoplasm (Figure 1). On immunohistochemistry, tumor cells were positive for Melan A and negative for Inhibin A, EMA and chromogranin A with Ki-67 proliferation index is 8-10% in highest proliferating areas with nuclear pleomorphism (Figure 2).

Discussion
The adrenal oncocytoma is rare benign neoplasm, has been seen mostly

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in kidney, thyroid and salivary glands. Large size (> 5 cm), heterogeneous appearance and presence of necrosis can be confused with adrenal cortical carcinoma. These adrenal oncocytomas are often detected incidentally by imaging studies during the evaluation for unrelated problems and should be included in the differential diagnosis of adrenal incidentalomas, especially if large tumors are detected. It has been seen that approximately 22% patients have adrenal oncocytoma with malignant behavior. The pathogenesis of adrenal oncocytoma is not well established. Females are more affected than male with left side predominance.

Adrenal gland masses are best visualized on CT or MRI but still no definitive features can differentiate benign from malignant adrenal oncocytic neoplasm on imaging. There are some features which can differentiate the adrenal oncocytic tumor from adenoma, like non-homogeneous appearance, increased attenuation, fat-poor on CT scan and loss of signal intensity on oppose phase MR images, fat poor on MRI. Generally, fat concentration is useful to differentiate the majority of malignant and benign adrenal lesions because almost all malignant lesions are lipid-poor, whereas the most of benign lesions are lipid-rich and present lower attenuation on CT scan. Definitive diagnosis of oncocytoma can be made pre-operatively by fine needle aspiration cytology after careful exclusion of functionality of tumor and when the tumor outline is preserved along with no invasion to surrounding structure.

Histopathologically, Weiss system is the most popular for differentiating benign from malignant adrenocortical neoplasm. In rare situations, histopathology is unable to differentiate it from other adrenal masses so by doing immunohistochemistry like negative chromogranin A and absence of neurosecretory granules on microscopy will differentiate from pheochromocytoma. Adrenal oncocytoma demonstrate numerous mitochondria in tumor cell cytoplasm at electron microscopy and absence of mitochondria will give clue to the diagnosis of adrenocortical adenoma.

Laparoscopic adrenalectomy is the procedure of choice from small as well as large functioning and non-functioning adrenal masses. Even bilateral large functioning adrenal tumors can be treated safely by laparoscopically in single stage when the operator has sufficient experience. Laparoscopic approach was adopted in this patient, which has been proved to be an extremely reliable procedure for benign pathologies and isolated metastases. Laparoscopic surgery offers a safe alternative in confronting adrenocortical neoplasms, even when the biological behavior of the tumors cannot be pre-operatively evaluated in a definite way.

It remains challenging, however, to distinguish between benign and malignant adrenal oncocytomas. A combination of biochemical, histopathologic, radiologic, and clinical features can be used to guide appropriate management, but surgical resection remains the mainstay of treatment for lesions deemed to be malignant.

In our case also, laparoscopic adrenalectomy was done which was proven to be adrenocortical oncocytoma histologically and on immunohistochemistry, tumor cells are positive for Melan A and immunonegative for Inhibin A, EMA and chromogranin A. Ki-67 proliferation index is 8-10% in highest proliferating areas.

Conclusions

Adrenocortical oncocytomas are rare neoplasm of adrenal gland, majority of them benign and nonfunctional, but few can be functional and may present as Cushing’s syndrome. Laparoscopic adrenalectomy is the treatment of choice for adrenal oncocytomas.

References