Adrenocortical carcinoma with extension in to inferior vena cava and right atrium - report of 3 cases
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Background: Adrenocortical carcinoma is a rare highly malignant tumour. At the time of diagnosis, the majority of patients have local lymph node/visceral invasion (stage III) or distant metastases to liver, lung, bone and brain (stage IV). We report three unusual cases of adrenocortical carcinoma with extension into the inferior vena cava and the right atrium.

**Case 1:** A 31 yr old lady presented with weight gain, hirsutism and amenorrhoea of two years duration. On examination she appeared cushingoid with centripetal obesity, supraclavicular fat pads and proximal myopathy. Hormonal assessment was consistent with Cushing’s syndrome of adrenal origin. CT adrenals showed a 10x10x 8.5cm left adrenal mass extending in to IVC superiorly to the junction with the right atrium. Left adrenalectomy and removal of tumor thrombus from IVC and right atrium was performed on cardiopulmonary bypass. Histology confirmed adrenocortical cancer. Postoperatively, she was commenced on hydrocortisone and mitotane therapy and her recent CT scan showed no local recurrence or distant metastasis. Her recent 24 hour urinary free cortisol levels were normal. She is currently stable with no recurrence 18 months latter.

**Case 2:** A 53yr old lady presented with hypertension and abdominal distension. Biochemistry revealed Cushing’s syndrome of adrenal origin. CT scan showed a 7cm right adrenal mass extending in to IVC. PET FDG scan revealed a focus of increased activity in the segment 4a of liver. Perioperative Trans Oesophageal echo disclosed tumour extension in to right atrium. She underwent right adrenalectomy with removal of tumour from atrium and IVC under cardiac bypass. Postoperatively, she developed an infrarenal IVC thrombus and was commenced on heparin. This triggered bleeding from liver metastasis which responded to selective embolisation of the right inferior hepatic artery. Postoperatively, she was commenced on hydrocortisone and adjuvant mitotane therapy. Her recent CT scan showed no local recurrence and the some of the hepatic lesions have resolved. Her recent 24 hour urinary free cortisol level were normal. Nine months in to mitotane chemotherapy she is stable with no local recurrence.

**Case 3:** A 24 yr old male presented with weight gain, hypertension muscle weakness and reduced exercise tolerance of only 20 yards. He appeared cushingoid and had a palpable mass in left flank. Biochemical investigations confirmed Cushing’s syndrome of adrenal origin. MRI showed a 19x18cm left adrenal mass extending in to IVC and right atrium with no hepatic metastases. Left adrenalectomy, left nephrectomy and splenectomy were performed as the tumour was very large with extensive adhesions. Tumour was removed from IVC and atrium under cardiopulmonary bypass. Postoperatively, he developed coagulopathy, acute renal failure and severe respiratory failure needing tracheostomy and prolonged respiratory support. Unfortunately, this patient died 17 days after surgery.

**Discussion:** Right atrial involvement is very rare in adrenocortical carcinoma (3%). Radical adrenalectomy with removal of the tumour extending to right atrium under cardiopulmonary bypass can improve the outcome of these patients. The presented cases illustrate that when appropriate expertise is available, atrial involvement should not be a limiting factor for surgery.