Sarcomatoid carcinoma of adrenal gland: a rare case report
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Abstract
Adrenal sarcomatoid carcinoma (ASC) are very rare and aggressive malignant tumors of adrenal glands containing both epithelial (carcinomatous) and mesenchymal (sarcomatous) components. ASC presents a diagnostic challenge due to its atypical symptoms and histological patterns which influence treatment. At the time of diagnosis, a large percentage of patients are already at the metastatic stage and succumb within a few months. Here, we report a case of functional sarcomatoid carcinoma in right adrenal gland in a 37-year-old female, who presented with new onset hypertension. The patient underwent right sided adrenalectomy and she was alive at the time of writing the current report. To the best of our knowledge, ASC reported in literature to date showed only few patient presented with endocrine hypersecretion and only one patient presented with both hypertension and endocrine dysfunction. So, our case is the rarest among the rare.

Keywords: adrenal gland, functional, sarcomatoid carcinoma.

Introduction
Sarcomatoid carcinomas, comprised of sarcomatous and carcinomatous differentiation components, are a notable kind of malignant tumor, commonly occurring in the digestive and respiratory tracts and breasts. These tumors are rarely observed in the adrenal glands. Primary adrenal sarcomatoid carcinoma, an extremely uncommon, aggressive malignant tumor, constitutes a subgroup of adrenocortical carcinomas. Patients can present with non-functional adrenal swellings with pressure symptoms by themselves or the result of their metastatic damage to other organs, as well as functional adrenal swellings with corresponding endocrine disturbances. The majority of reported patients succumbed to disease within a year, due to local recurrence or de novo metastases. Timely and accurate diagnosis and effective treatments are therefore required. The present study reports a case of primary adrenal sarcomatoid carcinoma with endocrine hypersecretion.

Case report
A 37-year-old female presented with 3-month history of gradual swelling of face and gaining of 10 kg weight. For last 1 month, she also had paroxysmal palpitation along with facial flushing, recurrent right sided upper abdominal pain, generalized weakness and lethargy. These problems compelled her to visit a physician with subsequent diagnosis of hypertension and she was put on amlodipine. Her menstrual cycle was normal and she had no history of taking steroid or oral contraceptive pills. The patient was obese with Cushingoid appearance, had hirsutism and dorso-cervical hump without any abdominal mass. Baseline biochemical investigations revealed diabetes mellitus, dyslipidemia and hypokalemia with metabolic alkalosis. Hormonal assay showed hyper-cortisolism (raised mid-night salivary cortisol and non-suppressive dexamethasone suppression test), hyperaldosteronism (raised serum and 24-hour urinary aldosterone), upper limit of 24-hour urinary metanephrine and normal
androgen level. Ultrasonography of abdomen identified an underlying right adrenal mass measuring 9 cm x 6.9 cm. A computed tomography (CT) scan of abdomen (with contrast) confirmed a rounded well defined mixed density mass of 10 cm x 8 cm having flecks of calcification at right adrenal region (Figure 1a & 1b). After intravenous contrast, moderate and heterogeneous enhancement with absolute washout 55% and relative wash out 20% suggested carcinoma or pheochromocytoma.

Hence, decision was taken for right adrenalectomy. The surgery was completed uneventfully. On microscopy, there was anaplastic adrenocortical cells, having abundant cytoplasm with pleomorphic hyperchromatic nuclei embedded within a sarcomatoid stroma. Areas of necrosis, calcification, osteoid differentiation and cells in mitosis were also seen (Figure 2a & 2b). Immunohistochemistry and positron emission tomography (PET) scan were planned. Initial PET scan was normal, but about 3 months later she developed resistant hypertension. This time PET scan showed metastasis to multiple sites. At the time of writing this report, 5 months post-operatively the patient remained alive and was taking 3rd cycle of chemotherapy.

Figure 1a & 1b Computed tomography images showing mixed density mass with flecks of calcification in right adrenal gland

Figure 2a & 2b Histopathological pictures of tissues from right adrenal gland showing epithelial and sarcomatous components
Discussion
Sarcomatoid carcinomas of the adrenal glands are extremely rare malignant tumors known with 1 year mortality of 100%. The age of presentation ranges from 29-79 years (mean 54.3 years). Although the number of cases reported is small, there is a female to male ratio of 1:1.4. Tumours tend to be very large at the time of presentation (mean size 13 centimeter). Usual presentation is with vague abdominal pain, abdominal lump in the flanks, loss of weight and generalized weakness. These carcinomas may be classified as either functional or non-functional tumors. If the tumor is functional, patients may present with the corresponding endocrine abnormalities like hypercortisolism causing Cushing’s syndrome, hyperaldosteronism causing severe hypertension, muscle cramps, testosterone secretion in women causing deepening of voice, acne, balding, hyperestrogenemia in males causing gynaecomastia, impotence and in females causing irregular menses, menorrhagia. Despite aggressive treatment, this variant of adrenocortical carcinoma has a poor prognosis, with the majority of patients succumbing to the disease within 3-12 months (mean 7 months) following surgical intervention. Limited surgical resection and presence of distant metastatic spread at time of presentation are the main culprit of early demise. Pre-operative diagnosis is elusive, which reduces treatment effectiveness and directly influences prognosis. Dynamic enhanced multidetector row CT scans may reveal tumor composition and blood supply. Histopathology and immunohistochemistry must be performed to confirm a diagnosis. Surgical excision, including complete metastatic resection, is currently the most effective treatment, while post-operative adjuvant chemotherapy may decrease relapse and metastasis rates. Doxorubicin based chemotherapy has been used. Adjuvant chemotherapy is ineffective in advanced disease; therefore, further research is warranted in order to develop specific therapy for the treatment of this unusual but devastating type of tumor.

Conflict of interest: Nothing to declare.

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