Adrenal

ADRENAL CASE REPORTS II

Metastatic Adrenocortical Carcinoma in Remission After a Single Dose of Gemcitabine

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Background

Adrenocortical carcinoma (ACC) is a rare tumor with an incidence of one per million population per year. Overall prognosis is poor especially in the presence of distant metastasis where five-year survival rate is 10%. Aside from chemotherapy along with mitotane, second-line regimens are not well-established, and therapeutic options remain limited. Here we present a case of metastatic ACC with rapid and complete remission after salvage therapy with gemcitabine (GEM).

Clinical Case

A 28-year-old man presented with night sweats, 10-lb weight loss and abdominal pain for three months. An abdominal CT showed a 13x14 cm complex mass partially effacing the inferior vena cava (IVC) and a chest CT showed multiple pulmonary micronodules. Biochemical evaluation was unremarkable, confirming a non-functioning adrenal tumor.

The patient underwent right adrenalectomy, hepatectomy, right diaphragm resection and cavotomy of IVC. Pathology confirmed ACC with extra-adrenal and vascular invasion. He then underwent adjuvant intensity-modulated radiation therapy with no evidence of disease on imaging thereafter. However, he was lost to follow-up for six months and returned with new enlarging pulmonary nodules.

He was started on chemotherapy (cisplatin, etoposide, adriamycin) with mitotane which were discontinued after two months, due to intractable vomiting despite high dose glucocorticoids and anti-emetics. He was then enrolled in a clinical trial using immunotherapy with nivolumab and ipilimumab which led to a significant decrease in the size of pulmonary metastases. His clinical course was complicated by the development of type 1 diabetes mellitus and proliferative glomerulonephritis related to immunotherapy.

After one year of immunotherapy, repeat imaging showed disease progression with new pulmonary nodules. These agents were discontinued, and he was then given a single dose of GEM. Subsequent imaging with CT chest, abdomen and pelvis three months later showed complete response with resolution of lung nodules. He has no evidence of disease one year post therapy and continues to undergo active surveillance.

Discussion

GEM-based chemotherapy has shown to be a modestly active regimen based on a multicenter study which demonstrated a partial response or stable disease in 4.9% and 25% of cases, respectively, with a median duration of 26 weeks. Median progression-free survival is 12 weeks (range 1 to 94). From the available data, GEM remains an important option for salvage therapy however, to date, there are no reliable predictive biomarkers to potentially identify responsive patients.

Conclusion

This is an extremely rare case of metastatic ACC achieving complete response after definitive surgery and a single dose of gemcitabine, despite failing first-line chemotherapy and immunotherapy. The unusual positive response may lead to the consideration of its use as a second line regimen.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

Bradycardia as a Presentation of Malignant Pheochromocytoma

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SUN-902

Introduction

Pheochromocytoma usually presents with tachycardia, hypertension and other symptoms related to catecholamine release. Bradycardia has been reported in up to 10% of patients and is due to an unusual reflex to cathecolamine surge.

Case Report:

57-year-old morbidly obese female with a history of COPD, hypertension, paroxysmal bradycardia, and urinary incontinence was referred to endocrinology clinic after an adrenal mass was found incidentally as part of a work-up for microscopic hematuria and urinary incontinence. CT abdomen showed unilateral 3.3 x 3.9 cm left lobular adrenal mass. Family history was negative for malignancies or pheochomocytoma. Patient denied substance abuse or ethanol use. Her medications include amlodipine, hydrochlorothiazide and albuterol. Physical exam was remarkable for a BMI of 51.6 and bradycardia of 50 bpm and well controlled blood pressure. On further review of her history, patient had prior visits to outpatient and emergency department for chronic headaches, insomnia, palpitations and sweating. Patient had a previous admission 13 years prior for chest pain and hypertensive urgency associated with headache and junctional bradycardia.

Laboratory workup was significant for serum metanephrine levels of 749 pg/ml (normal <42), serum normetanephrine 185 pg/ml(normal <145), serum chromogranin A 130 ng/L(normal <39) and 24hr urine metanephrine of 1340 ug/24hr (normal <96) and urine normetanephrine of 357 ug/24hr (normal <80). Previous ECG showed sinus bradycardia. Echo-cardiogram revealed normal ejection fraction with left atrial enlargement.

MIBG showed large focal area of intense radio-tracer accumulation in left mid abdomen and small focal lesions in the liver. Diagnosis of malignant pheochromocytoma was made and patient was referred to another institution for further management.

Conclusion:

Malignant pheochromocytoma is a rare neuroendocrine tumor that usually presents with tachycardia and hypertension. 10% of patients with malignant pheochromocytoma can debut with bradycardia due to sinus arrest. Although malignant and benign pheochromocytoma share the same clinical and histological features, the presence of metastasis is the main clue to differentiate them. Surgery remains the main treatment option even if complete