Adrenal ODPO27

An Unfortunate Case of Adrenocortical Carcinoma - Review of Treatment
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Adrenocortical carcinoma (ACC) is a neoplasm with an incidence of 1 case per million people every year, with a greater predilection for females. ACC may present as an incidental finding on imaging, abdominal pain due to mass effect and/or as an endocrinopathy (Cushing’s syndrome, hyperandrogenemia). Prognosis is poor with a 5-year survival rate ranging from 10-60% depending on stage. Treatment options are limited. A 53-year-old female presented to the office with the complaints of abdominal pain, facial flushing, hirsutism, and 40-pound weight gain over the past year. On examination she was found to have stage 2 hypertension and central obesity. A CT scan of her abdomen showed a left heterogenous adrenal mass measuring 8.5x8.5x8.0 cm. 24-hour urine cortisol levels were 10 times the upper limit of normal confirming Cushing’s syndrome for which she was started on ketoconazole. She underwent left adrenalectomy and pathology was consistent with ACC. The histological marker of proliferation, Ki67, was 20% and suggested high-grade disease. In the setting of hypercoagulability from Cushing’s syndrome and malignancy, she developed a saddle pulmonary embolus for which she was treated with anticoagulation. A PET CT scan showed metastasis to the spleen, paraaortic lymph nodes, and mesentery. Upon multi-institutional expert review, adjuvant mitotane was initiated, albeit delayed due to poor patient follow up. Despite therapy, she suffered compression fractures from worsening metastatic disease. Eventually, the patient opted for hospice care and passed away. Initial therapy of ACC includes complete surgical resection. However, metastasis is typically present at time of presentation thus resection is rarely curative. Mitotane is often used as adjuvant therapy as several retrospective studies have shown a reduction in disease recurrence in patients with resected stage I to III ACC. However, there is mixed data regarding disease-free and overall survival. Though mitotane benefit has been seen at low doses (1 to 3 g/day), individual tumor metabolism variably affects therapeutic plasma levels. Thus, some protocols are based on achieving therapeutic levels (14-20 mcg/ml) which require additional monitoring. Prior to mitotane therapy, hypercortisolism is treated with medications like ketoconazole that block cortisol synthesizing enzymes. Mitotane has adrenolytic properties, so concomitant glucocorticoid replacement is required. Chemotherapy without mitotane has not been deemed effective, and it is uncertain whether chemotherapy in combination with mitotane is superior to mitotane alone. Few studies exploring immunotherapy with pembrolizumab following mitotane or chemotherapy have shown a small effect in prevention of cancer progression. Adjuvant radiation usefulness is unclear, and it is primarily utilized palliatively. Progress in treatment has been limited by rarity of ACC, metastasis at time of diagnosis, and poor response to therapy, as exemplified by this case. Robust data from more clinical trials is needed to improve ACC treatment and outcomes.

Presentation: No date and time listed