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Severe Cholestatic Jaundice (Stauffer Syndrome) as a Rare Paraneoplastic Manifestation in Adrenocortical Carcinoma

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Introduction: Adrenocortical Carcinoma (ACC) is a rare and often aggressive malignancy arising from the adrenal cortex. Rarely, ACC can be associated with a paraneoplastic syndrome, such as tumor-associated hypoglycemia due to insulin-like growth factor-2 secretion, hyperreninemic hyperaldosteronism, erythropoietin-associated polycythemia,

and leukocytosis due to chemokine release from the tumor. Stauffer syndrome is a paraneoplastic syndrome characterized by reversible cholestasis in the absence of liver metastases, most frequently described in the context of renal cell carcinoma. Our case is the first reported association between Stauffer syndrome and ACC.

Clinical Case: A 38-year-old man presented with nausea, vomiting, painless jaundice, pruritus, and weight loss. Laboratory evaluation revealed an elevated total bilirubin of 8.7 mg/dL (N<1.3), the INR was elevated secondary to the cholestasis from vitamin K insufficiency. Computed tomography (CT) scan revealed a 20.4 cm, heterogeneous left adrenal mass with central necrosis concerning for ACC. There was no evidence of liver metastases. Additional laboratory workup showed elevated DHEA-sulfate of 1764.2 ug/dL (N<568.9), ACTH of 18 pg/mL (N<7-63), cortisol of 18.30 mcg/dL (N<19.50), androstenedione of 247 ng/dL (N<150), estradiol of 123 pg/mL (N<41), testosterone of 237 ng/dL (N<871), aldosterone of 11 ng/dL (N<=21), and plasma renin activity of 14 ng/mL/hr (N<4.3). Plasma metanephrines and catecholamines were normal. The patient's condition deteriorated rapidly with progressive renal failure and uremia requiring intensive care unit admission for continuous renal replacement therapy and critical care optimization preoperatively. The patient's total bilirubin increased to 34.5 mg/dL, which prompted liver biopsy. This revealed no evidence of cirrhosis or hepatic metastasis. Therefore, severe cholestasis was attributed to Stauffer syndrome, a paraneoplastic cholestatic jaundice from the underlying adrenal mass. The patient underwent exploratory laparotomy with left adrenalectomy, left nephrectomy, left ureterolysis, and wedge liver biopsy. Histopathology showed a necrotic ACC with tumor invasion into the adrenal capsule, no lymphovascular invasion, margins uninvolved, and Ki-67 index of 40%. Kidney parenchyma exhibited diffuse pigment casts. The liver specimen contained diffuse bile deposits and minimal chronic inflammation in the portal tracts, consistent with Stauffer syndrome. Postoperatively, the patient had rapid improvement in his renal function, and bilirubin level normalized after several weeks. Mitotane therapy was deferred until the complete recovery of the liver function.

Conclusion: Our case highlights a unique presentation of paraneoplastic hepatic dysfunction with jaundice associated with newly diagnosed ACC. Although Stauffer syndrome is one of the most characteristic paraneoplastic syndromes associated with renal cell carcinoma and other malignancies, paraneoplastic hepatic dysfunction associated with ACC has not been previously reported. The patient had rapid improvement of hyperbilirubinemia after surgical resection of the tumor. Stauffer syndrome should be considered in patients with ACC with liver dysfunction and jaundice without evidence of liver metastases.

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