Adrenocortical Carcinoma and Sudden Death

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INTRODUCTION

Adrenocortical carcinomas (ACCs) are rare tumors that have a generally poor prognosis (1). The clinical course is usually associated with complications arising from active hormone secretion or metastatic disease. The following case is presented to illustrate an exceptionally rare but devastating complication of ACC, that of massive pulmonary thromboembolism arising from direct infiltration of tumor into the inferior vena cava (IVC) via the renal vein.

CASE REPORT

A 26-year-old male had been diagnosed with a left ACC approx 1 month prior to his death. He had presented with a 2-year history of intermittent gynecomastia with recent onset of fever, night sweats, and abdominal distension, and imaging studies had shown a tumor of the left adrenal gland with extension into the IVC with metastases to the liver and to the thoracic and lumbar spine. Biopsy revealed an ACC, confirmed immunohistochemically, for which he had just completed his first cycle of chemotherapy. The deceased collapsed unexpectedly at his home and could not be resuscitated.

At autopsy, the major finding involved his left adrenal gland, which was completely effaced by a large tumor mass measuring 190 × 130 × 80 mm and weighing 1264 g. There was no connection between the tumor and the adjacent left kidney. The cut surface of the tumor was lobulated with a tan color and focal areas of hemorrhage and necrosis (Fig. 1). A tongue of reddish-colored gelatinous tumor and thrombus had extended into the left adrenal vein and left renal vein, and then into the IVC (Figs. 2,3). The right adrenal gland was moderately atrophied. Histological examination of the tumor revealed aggregates of large, irregular eosinophilic cells with hyperchromatic nuclei and scattered mitotic figures in keeping with an ACC (Fig. 4). Deposits of metastatic tumor were identified in the liver and vertebrae and were also confirmed histologically.

The lungs were edematous with a combined weight of 1628 g. A large coiled saddle pulmonary thromboembolus (maximum diameter of 20 mm) was found occluding the pulmonary outflow tract, with smaller more peripheral pulmonary thromboemboli. No tumor deposits were identified in the thromboemboli. The thromboemboli had arisen from a tongue of tumor that had grown through the left adrenal vein into the inferior vena cava. Despite a high rate of angio-invasion there are very few reports of sudden death resulting from this phenomenon in patients with adrenocortical carcinoma.

Key Words: Forensic pathology; adrenocortical carcinoma; sudden death; pulmonary thromboembolism.

Abstract

A 26-year-old man who presented with a 2-year history of intermittent gynecomastia with recent onset of fever, night sweats, and abdominal distension was found to have a left-sided adrenocortical carcinoma with metastases to the liver and spine. Sudden death occurred 1 month after his presentation. At autopsy a saddle pulmonary thromboembolus was found occluding the pulmonary outflow tract, with smaller more peripheral pulmonary thromboemboli. No tumor deposits were identified in the thromboemboli. The thromboemboli had arisen from a tongue of tumor that had grown through the left adrenal vein into the inferior vena cava. Despite a high rate of angio-invasion there are very few reports of sudden death resulting from this phenomenon in patients with adrenocortical carcinoma.

DISCUSSION

ACC is a rare and aggressive tumor that is almost universally fatal, with a 5-year survival without treatment of only 3 to 12% (1,2). The clinical manifestations in patients with ACC may be the result of hormonal effects in actively secreting tumors, with virilization, feminization, or manifestations of Cushing’s syndrome, or to mass effects on surrounding tissue and organs from tumor bulk (2–4). Tumors of the adrenal cortex can arise either sporadically or in association with certain diseases with genetic bases such as Li-Fraumeni syndrome, type 1 multiple endocrine neoplasia (MEN 1), Carney complex, or Beckwith-Wiedemann syndrome (5). Determination of the prognosis of ACCs from histopathological or DNA analyses has proven difficult (3,6).