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CASE REPORT

ADULT CLEAR CELL SARCOMA OF THE KIDNEY WITH TUMOR EXTENSION INTO THE LOWER PART OF THE INFERIOR VENA CAVA

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INTRODUCTION

Sarcomas of the kidney are rare and constitute 1 to 3 percent of all malignant renal tumors[1]. Clear cell sarcoma of the kidney is an aggressive paediatric neoplasm now thought to be pathologically distinct from Wilms tumor. The mean presenting age is 36 months. Although there have been other case reports of clear cell sarcoma of the kidney in the adult, many are not well documented and did not undergo thorough pathological analysis to distinguish them from Wilms tumor.

Herein we report a case of clear cell sarcoma of the kidney with a vena caval tumor thrombus in an adult patient.

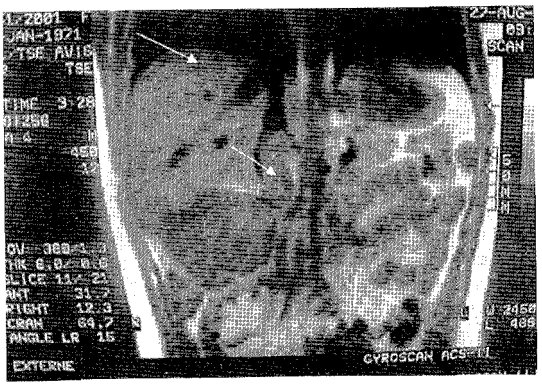

Fig. 1: Magnetic resonance imaging showing a heterogenous right renal mass with extension into the inferior vena cava (arrow)

OBSERVATION

A 30-year-old female presented with a 4-month history of right upper abdominal discomfort and 8 kg weight loss. The patient denied any history of renal colic passage of renal stones or overt urinary tract infection. Physical examination was normal. Abdominal computerized tomography showed an 7 x 10 cm renal mass with a tumor thrombus. Magnetic resonance imaging (MRI) confirmed the presence of a thrombus extending to the lower part of the inferior vena cava, well below the confluence of the hepatic veins (Fig. 1). Metastatic evaluation was negative. All other laboratory findings were within normal limits. The patient underwent right radical nephrectomy, hilar lymphadenectomy and removal of the vena caval thrombus through a subcostal incision and median sternotomy. Due to the fact that we do not have any experience with liver transplantation techniques for such cases, cardiopulmonary bypass and circulation arrest were used to remove the mass. The convalescence was uneventful and the patient was discharged home 10 days postoperatively.

The specimen weighed 850 gm (Fig. 2). Microscopy revealed cords of primitive cells within arborizing capillaries (Fig. 3). Stains for chromogranin, actin, cytokeratin, NSE and CD34 were negative while the stain for vimentin was positive. The diagnosis of renal clear cell sarcoma was made. The final pathological stage was T3bNoMo. No adjuvant chemotherapy was given after surgery. The 6-month follow-up visit showed a liver metastasis on CT scan of the abdomen. The patient subsequently received combination chemotherapy with actinomycin, vincristine and

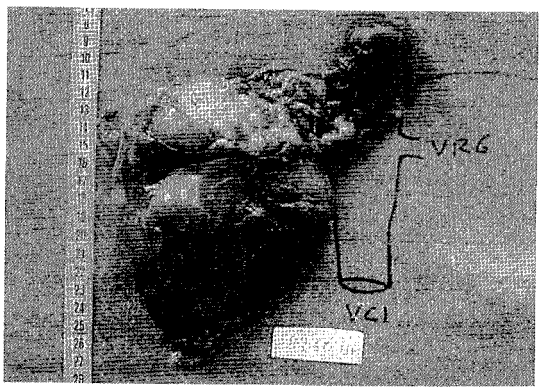

Fig. 2: Nephrectomy specimen with thrombus

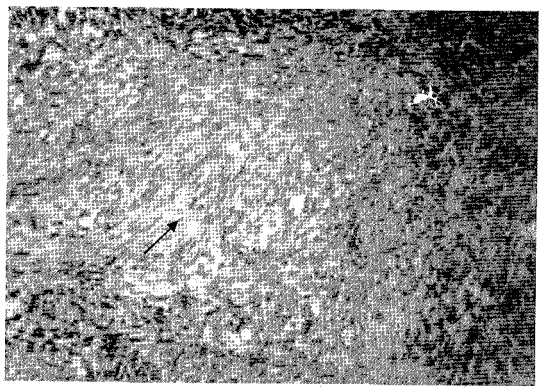

Fig. 3: Micrograph showing cords of discohesive cells with indistinct borders embedded in vascular stroma (H & E, x 250)

doxurubicin. with no response. Radiotherapy (5,000 cGy) was administered to the right hemi-abdomen to reduce the liver mass. The patient is still alive with recurrent disease.

DISCUSSION

First described in 1970, clear cell sarcoma of the kidney was initially thought to be a variant of Wilms tumor with an unfavourable prognosis. Since it has the propensity to metastasize to the bone, it was formerly termed bone metastasizing renal tumor of childhood[2]. However, as in most cases of soft tissue sarcoma, liver metastases were present in our case. It is unclear whether this tumor behaves differently in adults. A recent review of 351 patients of all ages revealed significant prognostic factors to be stage, age, presence of necrosis and treatment with Doxorubicin[3]. These factors primarily apply to paediatric patients, as adult cases have been extremely rare. Clear cell sarcoma of the kidney can have a variety of histological patterns, and immuno-histochemical analysis by experienced pathologists is necessary to confirm the diagnosis. The optimal treatment is unknown. Surgery, radiotherapy and chemotherapy are used alone but mostly in combination[4]. Doxorubicin has been reported to be effective, and incorporating it into chemotherapy regimens has resulted in a marked improvement in prognosis with 66% reduction in tumor related mortality[5]. To our knowledge we report on the third adult with clear cell sarcoma of the kidney and a vena caval thrombus.

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EDITORIAL COMMENT

This paper describes a rare tumor but in the sense this was a pathological curiosity, because we did not know what the histology was before the operation was carried out.
The best way to deal with inferior vena cava thrombi at this level is to use liver transplantation techniques to full immobilize the right lobe of the liver and to isolate the inferior vena cava at the supra hepatic level and the infra hepatic level rather than putting the patient on full cardio pulmonary bypass.

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