

Stauffer's Syndrome: A Rare Paraneoplastic Syndrome with Renal Cell Carcinoma

Abstract

An elderly male patient presented with cholestatic jaundice and weight loss. On evaluation, he was found to have left renal mass and hepatomegaly. Diagnosis of Stauffer's syndrome was confirmed based on his clinical history, biochemical evaluation, and liver biopsy. Resolution of jaundice was noted after removal of the renal mass.

Keywords: Cholestasis, jaundice, paraneoplastic, renal

Introduction

Nonmetastatic nephrogenic hepatic dysfunction syndrome (Stauffer's syndrome) is a paraneoplastic manifestation that often appears as the initial clinical presentation of renal cell carcinoma, bronchogenic carcinoma, leiomyosarcoma, and prostate adenocarcinoma.^[1-3] Although jaundice has rarely been described with this syndrome, a few case reports have highlighted a variant of the syndrome with deep icterus.^[4,5]

Case Report

A 63-year-old male presented with a history of yellowish discoloration of eyes and urine, generalized itching, and weight loss of 10 kg over last 1 month. On clinical examination, he was afebrile, deeply icteric, and had palpable liver. His investigations showed hemoglobin 8.4 gm%, total count of 7070/Cu.mm, and platelets of 191000/cu.mm. Serum bilirubin was 35.8 mg/dl (direct bilirubin – 32.10 mg/dl), aspartate aminotransferase – 42 IU/ml, alanine aminotransferase – 58 IU/ml, international normalized ratio – 1.14, and serum creatinine – 1.8 mg/dl. Serological tests for hepatitis B and C, HIV, and autoimmune liver diseases were negative. Serum ceruloplasmin levels and ferritin levels were within normal range. Contrast-enhanced computed tomography abdomen showed a 5.3 cm × 1 cm × 4.8 cm left renal mass, hepatomegaly, and no evidence of biliary dilatation or ascites. Liver biopsy showed

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mild portal fibrosis with hepato canalicular bilirubinomatosis and lobular inflammation. The possibility of cholestatic jaundice due to paraneoplastic manifestation of renal cell carcinoma was considered. The patient was managed by therapeutic plasma exchange for severe pruritus in the preoperative period, and he underwent laparoscopic left radical nephrectomy. The surgical specimen showed clear cell renal cell carcinoma with perinephric fat, hilar sinus fat, hilar vessels, and ureter free of tumor invasion (Fuhrman Nuclear Grade II). Postoperatively, he had gradual fall in the bilirubin values over a period of 4 weeks.

Discussion

Renal cell carcinoma accounts for 2% of all cancers and 2% of all deaths due to neoplasms.^[6] It is associated with multiple systemic and paraneoplastic manifestations which may hide the true diagnosis until full evaluation.^[7] Cholestasis is malignancies which may be related to compression of biliary tree by lymph nodes, hepatic infiltration by the malignant cells, metastasis, side effects of drugs, or rarely due to paraneoplastic syndromes. Stauffer's syndrome, originally described in 1961 by M. H. Stauffer, is characterized by elevated alkaline phosphatase, erythrocyte sedimentation rate, α -2-globulin, and γ -glutamyl transferase, thrombocytosis, prolongation of prothrombin time, and hepatosplenomegaly, in the absence of hepatic metastasis.^[8] Interleukin-6 overexpression by the primary tumor has

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been implicated for the syndrome.^[9] Cholestatic variants of the syndrome have been described in literature; authors recommend that renal cell carcinoma should be included in the differential diagnosis of both icteric and anicteric cholestatic liver disease.^[4]

Our patient presented with cholestatic jaundice, itching, hepatomegaly, and no radiological evidence of liver metastases. On histopathological analysis, there was no renal vascular involvement. His clinical improvement after surgery confirms that his cholestatic jaundice was secondary to renal cell carcinoma. This case highlights a rare paraneoplastic syndrome associated with renal cell carcinoma.

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Conflicts of interest

There are no conflicts of interest.

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