STAUFTER’S SYNDROME WITH JAUNDICE, A PARANEOPlastic MANIFESTATION OF RENAL CELL CARCINOMA: A CASE REPORT

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Summary.- OBJECTIVE: To report an infrequent case of Stauffer’s Syndrome with jaundice as a paraneoplastic syndrome of a metastatic renal cancer.

METHODS: We describe the set up of cholestatic jaundice without neoplastic liver infiltration in a patient with a metastatic renal cell carcinoma, which turned back with surgery and systemic treatment.

RESULTS: Proper treatment of baseline disease enables turn back paraneoplastic signs and symptoms of Stauffer’s Syndrome.

CONCLUSIONS: Reversible cholestatic jaundice without evidence of hepatic disease is an infrequent form of the Stauffer’s syndrome. This paraneoplastic syndrome is associated particularly with renal carcinoma but was described in lymphoproliferative diseases, prostate cancer and broncogenic tumours.

This paraneoplastic entity is characterized by elevated alkaline phosphatase, erythrocyte sedimentation rate and gamma-glutamyl transferase without liver neoplastic infiltration.

Keywords: Stauffer’s syndrome. Jaundice. Cholestasis. Renal cell carcinoma.

INTRODUCTION

Renal cell carcinoma is associated with a variety of paraneoplastic manifestations that can be the main presenting symptoms even before the kidney tumor diagnosis. Stauffer’s syndrome or cholestatic liver dysfunction is one of them.

CASE REPORT

A 51 year-old man was admitted to the hospital because of a 8-month history of hyporexia, weight loss, several episodes of rinitis and sinusitis with intermittent fever and malaise.
Past medical history included non treated hypertension. He did not consume alcohol or use illegal drugs. He smoked 15 cigarettes daily for 30 years.

On physical examination, the patient appeared chronically ill but without physical findings.

Laboratory data revealed leukocytosis 12,500/mm$^3$, thrombocytosis 568,000/mm$^3$, erythrocyte sedimentation rate 120 mm/h, creatinine 2.3 mg/dl, alkaline phosphatase 7860 U/l (normal <360), gamma glutamyltransferase at 897 U/l (normal < 55), aminotransferases and serum bilirubin were normal.

A diagnosis of Wegener disease was thought on the basis of his history for rhinitis and sinusitis with elevated creatinine and several laboratory results. Immunologic tests revealed no evidence of those kind of disorders (rheumatoid factor, antinuclear and antimitochondrial antibodies were absent).

Ultrasound of the abdomen revealed inespecific hepatomeglay and a 4cm in diameter solid mass in the left kidney. An abdominal tomography (TC) revealed no evidence of metastatic disease and confirmed the results of the ultrasonography.

Radionuclide bone scan showed bone metastases.

After few days the patient developed a jaundice with serum Bilirubin 13,6 mg/dl.

The kidney was surgically excised, an esplenectomy was done and an excisional liver biopsy was performed.

The biopsy revealed a renal cell carcinoma of clear cell type with a sarcomatoid component. Spleen was normal. Liver had an intrahepatic cholestasis.

After the surgery hepatic function got better but not normal.

He received radiotherapy on his lumbar spine cord because of bone metasateses and interferon as systemic therapy (5mill IU subcutaneous three times a week). After two months of treatment repeated laboratory studies revealed normalization of his liver chemistries.

DISCUSSION

Tumors can produce signs and symptoms at a distance from the primary lesion or its metastases. These are collectively referred to as “paraneoplastic syndromes” or “remote effects” of malignancy (1).

Renal cell carcinoma is associated with an up to 20% prevalence of para-neoplastic syndromes, which may be precursors of primary or recurrent disease (2).

Non-metastatic nephrogenic hepatic dysfunction syndrome without jaundice has often been described in patients with hypernefroma- This syndrome, described by Stauffer in 1961 (3), has been also found in prostatic cancer, bronchogenic carcinoma and malignant lymphoproliferative diseases (4,5).

Stauffer’s syndrome is a rare paraneoplastic entity that is characterized by elevated alkaline phosphatase, erythrocyte sedimentation rate, alpha-2 globulin, and gamma-glutamyl transferase, thrombocytosis, prolongation of prothrombin time, and hepatosplenomegaly, in the absence of hepatic metastasis (6).

It is associated with the production of cytokines by the tumor, specially the overexpression of interleukin-6 by the neoplastic cells (7).

Paraneoplastic cholestatic jaundice is extremely uncommon as a variant of Stauffer’s syndrome and few cases are published in the literature (8,9).

Our patient presented with the rare variant of icteric cholestasis. The absence of liver metastases and the presence of the typical intrahepatic cholestasis was confirmed by biopsy. Because he had bone metastases the laboratory normalization was not total till systemic treatment was indicated and objective response was obtained.

CONCLUSION

Our case, with confirmation by liver biopsy of the paraneoplastic cholestasis, is consistent with the icteric variant of Stauffer’s syndrome.

Physicians should recognize paraneoplastic syndromes as rare manifestations of common carcinomas and their usual improvement after the primary tumor treatment, specially in kidney cancer, in which the patognomonic triade of hematuria, polyglobulia and pain hardly appeared at the moment of the diagnosis.

REFERENCES AND RECOMMENDED READINGS

(*of special interest, **of outstanding interest)


