Paediatric T cell Lymphoma with Nephrotic Syndrome: A Rare Association

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Abstract Renal involvement is frequent in hematologic malignancies especially Hodgkin's lymphoma. Renal complications in children with malignancies primarily arise from tumour lysis syndrome, malignant infiltration or obstruction of the urinary tract, deposits of immunoglobulin fractions or crystals, renal infiltration by malignant cells, paraneoplastic or storage glomerulopathies. Nephrotic syndrome has been described in B cell type Non Hodgkin's lymphomas. There are very few reports of association of T cell lymphoma and nephrotic syndrome in pediatric patients. We present a case of peripheral T cell lymphoma with nephrotic syndrome in a 10 year old boy.

Keywords Nephrotic syndrome · Pediatric Lymphoma · PTCL

Introduction

Nephrotic syndrome as a rare paraneoplastic manifestation of extrarenal malignancy was first described by Galloway in 1922 [1]. Hodgkin's lymphoma has been the most commonly associated tumour, but cases have also been described in

connection with other haematological diseases and carcinomas [2, 3]. Non Hodgkin lymphoma (NHL) has been associated with nephrotic syndrome but mainly in adults and is often characterized by impaired renal function. A majority of such cases have occurred as a result of direct infiltration of lymphoma cells into the kidney. Nephrotic syndrome associated with pediatric T cell lymphoma has rarely been described.

Case Report

A 10 year old boy presented to the outpatient department in July 2010 with a history of left cervical lymphadenopathy of 1 year duration. He had no history of fever, weight loss or night sweats. Clinical examination revealed multiple left cervical lymph node enlargement, largest measuring 3×3 cm. There were no other lymph nodes or hepatosplenomegaly. Biopsy of the lymph node revealed Peripheral T cell NHL-unspecified. Immuno histochemistry showed CD3 and CD8 positivity and Alk1, CD15 and Epithelial Mebrane Antigen (EMA) negativity. Staging work up included computed tomography of chest, abdomen and pelvis and bone marrow examination; which were negative. Patient was diagnosed with Stage IA Non Hodgkin's Lymphoma, Peripheral T cell type.

He was started on chemotherapy with BFM 90 protocol and received the first cycle. He developed febrile neutropenia after the first cycle from which he recovered uneventfully. Unfortunately, patient defaulted for further chemotherapy. He reported back after 3 months with complaints of facial puffiness, decreased urine output and abdominal distension. At that time, he had persistent lymphadenopathy in the cervical region. Blood urea was 90 mg/dl and serum creatinine 1.2 mg/dl. 24 h urinary

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Fig. 1 a Sections shows single linear core of kidney biopsy with normal light microscopic histology of glomerular, tubular, interstitial and vascular compartments. Masson Trichrome stain, $\times 40$.

b Sections show glomeruli with normal capillary membrane thickness and cellularity and unremarkable tubules, PAS stain, $\times 400$

protein was 3.1 gm/day. Serum total cholesterol was 420 mg/dl, triglycerides 615, and LDL 61 mg/dl.

After excluding causes of ARF such as volume depletion, sepsis, uric acid nephropathy in association with tumour lysis syndrome, post-renal obstruction and, renal vein thrombosis, a renal biopsy was done which showed minimal change disease (Fig. 1a, b). He was started on prednisolone 60 mg/m² and diuretics. Subsequently urine output increased, edema came down and renal function normalised. Further he was continued on chemotherapy with CHOP regimen. Patient completed eight cycles of CHOP and is in clinical remission at the time of reporting.

Discussion

Many types of tumour have been reported to be associated with nephrotic syndrome, including various carcinomas, Hodgkin's lymphoma, other malignant lymphomas, chronic lymphocytic leukemia, and embryonal neoplasms. The glomerular lesions accompanying these tumors have been variable and range from membranous glomerulopathy to minimal change (lipoid) glomerulopathy [4, 5] and amyloid disease [6]. The most common type of lymphoma associated

with nephrotic syndrome is Hodgkin's lymphoma. It is clearly demonstrated that therapy of the neoplastic process causes remission of nephrotic syndrome [4].

NHL; mostly B cell NHL has been associated with nephrotic syndrome, mainly in adults. Association between T-cell lymphomas and nephrotic syndrome is less commonly described.

In some cases nephrosis is caused by invasion of the kidney by neoplastic cells [7], renal amyloidosis [8] or renal vein thrombosis. Amyloidosis cases during the course of NHL are usually of AL-type [8]. Neoplastic infiltration has been documented in about one-third of all lymphoma patients who underwent a post-mortem autopsy without signs of prior ARF during their lifetime [3]. In other cases an immunologic mechanism is likely. Some authors have suggested a possible pathogenetic link between T cell abnormalities and glomerular diseases, particularly minimal change disease and focal glomerulosclerosis [9].

In summary, we report a case of rare association of nephrotic syndrome in paediatric T cell lymphoma. It is extremely difficult to rule out other causes of nephrotic syndrome in this patient. Subsequent chemotherapy and supportive measures resulted in recovery of renal function and patient achieved a complete response at the end of the treatment.

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