“A Case Report on T2dm With Systemic Amyloidosis Leading To Non Diabetic Renal Disease.”

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I. INTRODUCTION

The name AMYLOID is attributed to the pathologist Virchow, who in 1854 thought such deposits in autopsy liver were cellulose because of their peculiar staining reaction with iodine and sulphuric acid. Amyloidosis is a term applied to a heterogeneous group of rare diseases characterized by extracellular deposition of amyloid (insoluble fibrillary proteins) causing target-organ dysfunction and a wide range of clinical symptoms. These symptoms depend on the organ involved, and include nephrotic syndrome, hepatosplenomegaly, congestive heart failure, carpal tunnel syndrome, gastrointestinal (GI) symptoms and macroGLOSSIA.

It is estimated that about 20%-40% of patients with type 1 or type 2 diabetes will develop diabetic nephropathy (DN), which contributes to most end-stage renal disease worldwide. Diagnosis of DN is mostly clinically based on a long history of diabetes, proteinuria, hypertension, and a progressive decline in renal function. This diagnostic approach is inconclusive, due to the fact that non-diabetic renal disease (NDRD) has been found in type 2 diabetes mellitus (T2DM) patients. The prevalence of NDRD in type 2 diabetic patients with renal involvement varies from 20-80%. The diagnose of NDRD in diabetic patients has an obvious prognostic and therapeutic importance. The common NDRD includes glomerulonephritis, vascular nephropathy, cholesterol microembolism and so on. Systemic amyloidosis is a rare disease with an incidence of 4.5 per 100000 person-years. The pathological production of fibrillar proteins can deposit in numerous tissues and cause organ dysfunction including kidney, liver, heart, lung, spleen, gastrointestinal tract and bladder. Here, we describe a rare case of type 2 diabetes presenting with massive proteinuria due to primary systemic amyloidosis.

II. Case report

A 60-year-old female presented with chief complain of b/l pitting oedema of legs, generalized weakness and frothy urine for 3 weeks. She was diagnosed type 2 diabetes on routine testing 14 years and was on regular treatment (OHA). There was no history of diabetic retinopathy or neuropathy. She denied family history of diabetes or systemic disease.

On Examination :- Her general condition was unsatisfactory his initial blood pressure in left arm supine position was 130/90 and pulse rate of 88/min. Pallor was present, icterus, cyanosis, Lymph node absent, b/l lower limb pitting oedema present. Her random blood sugar was 142mg/dl by glucometer.

CNS:-WNL Patient was conscious orientated to time, place, person
CVS:- S1S2+, No added sound, no murmur was heard
R/S:- B/L AE+, normal vesicular breath sound,
P/A:- Soft, Non-tender, liver palpable 1-2cm

Her initial investigation revealed microcytic, hypochromic anemia (Hb 8.2 method photometry, Wbc 7.6 method electrical impedance), RFT (B.urea 42, method urease with Indicator dye, S.cret 1.2method enzymatic, Na134 method Direct ISE, K 4.40 method direct ISE) LFT (bil 1.2 method azobilirubin/dyphylline, sgot 52 sgpt 78 method kinetic with pyridoxal phosphate , alp 78 method method pnpp/am buffer , total protein 4.2 method biuret(alkaline cupric sulphate), albumin 1.8 globulin 2.4(calculated)

Urine routine/Microscopy revealed pulse as 2-3/HPF, RBC 4-5/HPF, Epithelial Cell 1-2/HPL, Albumin ++++, Sugar+, method Dipstic Reflectance Spectrophotometry/Microscopy. Viral markers were negative. 24 hour urine protein 4.8 gm. USG (W/A) was done which was suggestive of hepatomegaly and rt kidney enlarged in size with maintained cmd and echotexture.

Abdominal fat pad aspiration was done which came out to be positive for amyloid.
III. DISCUSSION

Diabetic nephropathy is a complication of diabetes associated with the kidney which could progressively lead to end-stage renal diseases. The diagnosis of DN is frequently based on clinical characterization exclusively. Actually, a variety of NDRDs are often overlooked in diabetic patients, which have significant impacts on prognosis and treatment. The challenges still exist to differentiate NDRD from DN in diabetic patients. The definitive diagnosis of amyloid is usually based on the typical histopathological findings of amyloid in the affected tissue. Therefore, renal biopsy is the only tool to diagnose NDRD. However, the renal biopsy cannot be used as a routine diagnostic test in type 2 diabetic patients with proteinuria because of the invasiveness. Therefore, the main indication for renal biopsy was clinically thorough suspicion of NDRD. The common indications for NDRD in type 2 diabetes are short duration of type 2 diabetes, acute or rapidly progressive renal failure, glomerular hematuria, absence of diabetic retinopathy, nephritic syndrome with normal renal function. As in our case, despite the patients having T2DM for more than 10 years, high level of proteinuria without retinopathy suggests a diagnosis of NDRD other than DN. Thus, biopsy was considered in our patient for the precise diagnosis (abdominal subcutaneous fat pad aspiration).

Abdominal subcutaneous fat pad aspiration is a non-aggressive technique with good sensitivity, minimally invasive, outpatient based, quick, easy, reproducible and repeatable procedure and widely used as a screening test in diagnosis of systemic amyloidosis. A wide spectrum of NDRNs reported in patients with type 2 diabetes mainly include glomerular and tubulointestinal lesions. Our case, we diagnosed the systemic amyloidosis which is an infrequent cause of non-diabetic renal disease in patient with type 2 diabetes based on positive Congo red staining.

Systemic amyloidosis is a rare and severe disease which involves several organs including kidney, liver, heart, lung, spleen, gastrointestinal tract, bladder and endocrine system and leads to a high mortality rate. Without treatment, the median survival is less than 6 months. For all newly diagnosed cases of amyloidosis, an assessment of the specific type of amyloid is critical in ensuring proper therapy. Renal disease as a frequent manifestation of the systemic amyloidosis usually cause proteinuric renal failure in the context of normal or low blood pressure.

References