Case Report

Five-year girl with advanced lupus nephritis and insulin dependent diabetes mellitus

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INTRODUCTION

Insulin Dependent Diabetes Insipidus (IDDM) or Type 1 Diabetes (TID) involves a spectrum of renal diseases including diabetic nephropathy and non-diabetic renal disease. A number of renal diseases like membranoproliferative glomerulonephritis, immune complex glomerulonephritis and Goodpasture’s disease have been reported in TID.1–3 The association of TID with other organ-specific autoimmune disorders is well documented, but association with Systemic Lupus Erythematosus (SLE) is infrequently described in the literature.4–7 Lupus nephritis with TID has reported in literature mostly in adult population but rare in children.6

CASE REPORT

A five-year-old girl was presented with low grade fever, dryness of mouth and bilateral pain over knee joints over four months. She had facial puffiness, mild bilateral pedal edema, and ascites on admission. Her weight was 14.7 Kg (50th percentile) and height was 105 cm (50th percentile), heart rate 110/min and BP were 110/70 mm Hg (95th). On physical examination she had bilateral multiple non-tender cervical lymphadenitis and hepatosplenomegaly.

Investigations revealed she had mild anemia with Hb:10.1 gm%, ESR:134 mm/1st hour, TLC: 9.2 x 103/mm3, platelet count: 2.8 lakh/mm3, DC: N56, L40, M1, LDH: 923U/L(110-295U/L), blood urea: 40 mg/dl, serum creatinine: 0.8 mg/dl, serum electrolytes: Na:144 mEq/L, K: 4.6 mEq/L and iCa :1.07 mEq/L, Bone marrow aspiration showed normal, M:E= 5:1 ratio without any atypical cells, dsDNA: 69.86 IU/ml (normal range: <4 IU/ml), serum C3:55 mg/dl (normal range: 88-206 mg/dl), serum C4: <1 mg/dL (normal range:13-75 mg/dl), ANA hep-2:3+ speckled (positive), urine microscopy showed Albumin 3+, RBC: 10-15/high power field (HPF), Pus cells : 4/6/ hpf, Rheumatoid Factor: 228.6 IU/mL unit, CRP(Q): 4.4 mg/L. Serum Albumin 2.6 g/dl, serum cholesterol 229 mg/dl and
significant proteinuria (spot urine protein and creatinine ration 1.8).

Ultrasound of kidneys showed right kidneys of 8.1x4.0 cm and left kidney of 8.0x3.8 cm mild ascites. An ultrasound guided renal biopsy was conducted in view of hypertension, edema, significant proteinuria and microscopic hematuria which revealed; full house IF, LM (Figure 1), EM (Figure 2) suggestive of stage V lupus nephritis.

Initially she was treated with oral Prednisolone, MMF (Mycophenolate Mofetil), HCQ(Hydroxychloroquine) and Amlodipine to control blood pressure.

In subsequent follow up after six months, she was in remission with mild proteinuria but developed hyperglycemia (blood sugar 290mg/dl) and glycosuria (urine sugar 1.5 g%). She was diagnosed with type 1 DM and treated with sub cutaneous insulin with consultation with endocrinologist.

DISCUSSION

SLE and T1D are both autoimmune disorders of unknown etiology and rare in very young children. Inheritance of HLA-DR3 or DR4 haplotypes tenders an increased risk for the development of T1D.4, 7 HLA-DR3 and DR4 haplotypes also are important in disease predisposition for SLE.7 Furthermore, shared disease susceptibility alleles were found for SLE and T1D: STAT 4 8, 9 and TNFAIP3. 10 These imply common pathogenetic mechanisms in SLE and T1D.

The existence advanced stage of LN with IDDM, another autoimmune disease is rare in very young children. As in this case the child was found to have type 1 DM along with LN stage V, carry poor prognosis and she is having a high risk of CKD requiring close follow-up.

Figure 1: Light microscopy with H and E stain.

Light Microscopy stained with H and E, PAS, MT, silver methenamine and Congo red include renal cortical parenchymal core showing enlarged glomeruli and reveal diffuse mild increase in mesangial matrix and cellularity and diffuse thickening of capillaries which show membrane texture alterations with intramembranous “mottling” and few epimembranous "spikes" (Figure 1).

Three (13%) glomeruli had segmental endocapillary cellularity with intracapillary mononuclear cell/ neutrophil margination.

Figure 2: Electron microscopy.

Electron microscopy showing glomerular basement membrane thickness varies from 346.1 to 983.4 nm (mean 546.81 nm). Neutrophils are seen occluding lumina of few capillaries (Figure 2).

Scattered subendothelial, mesangial and many subepithelial/ intramembranous electron dense deposits are identified. The deposits do not show substructure on higher magnification. Visceral epithelial cell foot processes show significant effacement (about 60-70%) and focal microvillous transformation. Several tubuloreticular inclusions are identified in endothelial cell cytoplasm of glomerular as well as peritubular capillaries.

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