

Short Communication

MembranoProliferative GlomeruloNephritis in Down Syndrome

Rozita Abolghasemi¹, Rozita Doosti², Mahdieh Taghizadeh³, , Maryamossadat Shahidi⁴

1. Division of Nephrology of Imam Hossein Hospital, School of Medicine, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
2. MS research Center, Sina Hospital, School of Medicine, Tehran University of Medical Science, Tehran, Iran.
3. Medical Genetic Department, Medical Tarbiat Modaress University, Tehran, Iran.
4. Student's research committee of Medical Tabriz University, Tabriz, Iran.

Corresponding Author email: mtaghizadeh362@gmail.com

ABSTRACT: Down syndrome patients have many complications that affect their survival. A variety of nephrological abnormalities have been reported in some of them. We report a case of 38-years-old man with Down syndrome who presented with right leg pain & erythema, pitting edema and mild fever and history of cardiac valvular disease. His laboratory tests showed Creatinine rising and proteinuria. Biopsy of kidneys showed Memberano Proliferative GlomeruloNephritis (MPGN). We conclude that early diagnosis and treatment of renal disease is necessary for increasing the survival of Down syndrome patients.

key Words: MPGN; Down syndrome; Proteinuria; Renal disease.

INTRODUCTION

Down Syndrome (DS) is the most common chromosomal abnormality with prevalence of 1-2 cases per 1000 births. DS patients have many complications that affect their survival. A variety of nephrological abnormalities such as glomerulonephritis (Gupta et al., 1991), immunotactoid glomerulopathy (Takemura et al., 1993), focal and segmental sclerosis or hypercalcemia with modularly calcinosis (Filler et al., 2001), Chronic Renal Failure (CRF) and needing Chronic Peritoneal Dialysis (CPD) (Kupferman et al., 1994) and renal transplantation (Baqi et al., 1998) have been reported in some of them.

In the cross sectional study in DS evaluated 69 cases and reported hyperuricemia (24.2%), hyperuricemia without gout (11.6%), CRF (4.5%), voiding disturbances (0.04%), mild proteinuria (0.04%), microscopic hematuria (0.03%) and hypertension (0.01%) (Malaga et al., 2005).

It seems that the incidence of these anomalies is high enough to encourage systemic screening of them.

Case presentation

In Jun 2011, a 38-years-old man with DS was admitted to Imam Hossein Hospital because of right leg pain & erythema, pitting edema and mild fever. He had history of cardiac valvular disease with 1week admission in 13years before. In his physical exam, we found normal Jugular venous pressure (JVP), grade 3 holosystolic murmurs in apex and left sternal border and s3 heart sound, with clear lungs. Also pitting edema from knee to 1/3 of lower leg with erythema and decrease range of knee motion seems clear, but pulses were full and symmetric.

Echocardiography reported 60% ejection-fraction with sever Mitral regurgitation (MR) and Aortic insufficiency (AI) with no vegetation for rulling out endocarditis. Radiography and sonography of right leg showed soft tissue swelling and normal arterial flow. So we started treatment of cellulitis by vancomycine and gentamycine.

Routine laboratory tests performed and showed increasing in serum Creatinine (Cr= 1.9 mg/dl), Blood Urea Nitrogen (BUN=156), Erythrocyte Sedimentation Rate (ESR=35mm/h), +3 proteinuria in Urine Analysis (U/A). Volume of 24 hours urine was 1000ml with 3000 mg protein and 1.83 mg Creatinine. Repeating 24h U/A was 2600 ml volume with 3072 mg protein and 1.35 mg Creatinine.

Sonography of kidneys showed increased cortical echo and differentiation between corticomedullary echo. Biopsy of kidneys suggested Membranoproliferative Glomerulonephritis (MPGN), so treatment with cyclosporine and corticosteroid was started. Outpatient follows up until now showed decreasing proteinuria less than 100 mg with resolution of Acute Renal Failure (ARF).

Our patient also presented subclinical hypothyroidism, so levothyroxine was started for him.

DISCUSSION

Cardiac valvular disease, diabetes mellitus, hypothyroidism and renal involvement are common in DS patients, But to our knowledge it seems to be first time of reporting MPGN in them.

Membranoproliferative Glomerulonephritis (MPGN) that called Mesangiocapillary or Lobular Glomerulonephritis is an uncommon lesion found to underlie clinical manifestations of glomerular disease. Due to the extreme heterogeneity of pathogenesis processes that can be responsible for the pattern of injury, it is best not to regard it as a specific disease (West, 1986).

Idiopathic type I MPGN is currently a rare disorder and a diagnosis of exclusion. (Hepatitis C virus infection, with or without mixed cryoglobulinemia , Mixed cryoglobulinemia not due to hepatitis C virus infection, Systemic lupus erythematosus , Monoclonal immunoglobulin deposition diseases, Hepatitis B virus infection , Sub acute bacterial endocarditis (SBE) or infection of a ventriculoatrial or ventriculojugular shunt for treatment of hydrocephalus) (Khatab et al., 2010).

Type II is also called dense deposit disease, because it is characterized by continuous, dense ribbon-like deposits along the basement membranes of the glomeruli, tubules, and Bowman's capsule (Alchi and Jayne, 2010). Type III is an immune complex disease, similar to type I However, subepithelial deposits are prominent in type III and there is complex disruption of the glomerular basement membrane with large lucent areas. How this occurs is not well understood. There is an inherited form of type III disease that is linked to chromosome 1q32, the locus for the complement receptor family (Donoso et al., 2010).

Therefore regular monitoring of DS patients for renal diseases from early infancy to adulthood has been recommended.

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