Abstract

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Antiglomerular Basement Membrane Disease and Goodpasture's Disease

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Abstract

Goodpasture's disease also known as anti-GBM disease associated with high level of circulating IgG autoantibodies to the Alpha 3 chain of type IV collagen [1]. In the Caucasian population, the occurrence of Good pasture disease is more prevalent, reaching 1 case per million per year. Anti-GBM disease with pulmonary involvement is more prevalent in males, around 80% usually occur in the 2nd decade, whereas isolated anti-GBM nephritis can also occur in older people without male preponderance.Many patients with acute symptoms of lung haemorrhage may have kidney disease or have symptoms of lung haemorrhage in isolation, including cough, dyspnoea, haemoptysis, and iron deficiency anaemia.Renal involvement in glomerulonephritis may be presented with dark and red urine, but progression to oliguria is so fast that this step is skipped. Glomerulonephritis occurs in one third or half of patients in the absence of lung haemorrhage. Further deterioration is usually rapid once significant renal impairment has occurred.

Hematuria, modest proteinuria, dysmorphic red cell and red cell cast in microscopy have often been seen in urine examination (even apparently associated with pulmonary disease).

Renal biopsy is important because it gives diagnostic and prognostic information, typical histological features are diffuse proliferative glomerulonephritis with variable degree of necrosis, crescent formation, glomerulosclerosis and tubular loss. Linear deposition of immunoglobulin along with glomerular basement membrane is pathognomonic. IgG and anti-GBM antibodies that are circulating are almost always present.

The titers of anti-GBM antibodies correlate with the intensity of nephritis. Shift in titer also represents treatment and relapses.Anti-GBM antibodies often associated with ANCA [2], especially ANCA myeloperoxidase, in such double positive patients may have a clinical course and response to treatment more usually vasculitis than Goodpasture disease. Recovery of renal function is likely even patient is dialysis dependent. For most patients, anti-GBM disease treatment should be begun as soon as possible. However, the chances of recovery and independent kidney function in patients with some clinical and pathological conditions are poor. Kidney function recovery is only 5 percent in patients who have a high percentage of crescent on kidney biopsy (85- 100 percent).Oliguria and/or advanced kidney failure that needs dialysis to begin, should consider before initiation of immunosuppressive therapy due to low chance of kidney recovery and the ability of the patients to withstand intense immunosuppression but treatment is appropriate for those patients who have pulmonary haemorrhage.

The purpose of the treatment is to suppress kidney inflammation, remove circulating pathogenic autoantibodies and suppressing autoantibody formation.This treatment can prevent ongoing kidney damage but is unable to reverse the chronic kidney damage already developed. In most patients treated with plasma exchange combined with immunosuppression, antibodies are eliminated within eight weeks [3].Plasma exchange removes anti-GBM antibodies from the bloodstream steadily and slowly (within several weeks) and usually must be achieved for two to three weeks before anti-GBM antibodies are eliminated completely [4].Cyclophosphamide for 3 months and eventually tapered corticosteroids fully removed within 6 months tend to be enough based on available clinical data[5]. After 3 months of cyclophosphamide, continuation of treatment with either Azathioprine or Mycophenolate is indicated in patients with persistent anti-GBM antibodies. Anti-GBM disease relapses are rare. After renal transplant, recurrence of anti-GBM disease maybe 50 percent for those who have detectable anti-GBM antibodies at the time of renal transplant [6]. It is not recommended to treat patients who do not have detectable anti-GBM antibodies for more than six months.

Biography

Syed Shabbar Musavi works as a professor at the University hospital of Kerry, Co. Kerry, Ireland.