ANTIGLOMERULAR BASEMENT MEMBRANE (ANTI-GBM) DISEASE SUCCESSFULLY TREATED WITH MYCOPHENOLATE MOPHETIL (MMF).

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Anti-GBM disease is a rare form of vasculitis affecting glomerular and/or pulmonary capillaries and is associated with the autoantibodies which are directed against the type IV collagen antigens situated in the basement membrane. Rapidly progressive glomerulonephritis along with diffuse alveolar hemorrhage are the clinical manifestations of this disorder. The rarity of this disease has resulted in the treatment methods not having changed for many years. Standard therapy consists of therapeutic plasma exchanges (TPE), glucocorticoids (GCS) and cyclophosphamide (CTX). There are only a few cases presenting efficacy of drugs other than CTX in literature available. MMF, medication which inhibits B-cells and T-cells proliferation and the formation of antibodies, can be potentially a less toxic alternative to CTX.

Case presentation:

An 18-year old woman displayed fatigue, malaise, non-productive cough, hemoptysis and renal insufficiency. Her history included oral contraceptive treatment and smoking 10 cigarettes per day for 2 years. Serum creatinine concentration at the time of admission to the hospital was 4 mg/dl and increased rapidly to the maximum value of 7,3 mg/dl, together with escalated oliguria. Therefore temporary hemodialysis treatment was necessary (3 sessions). Laboratory tests revealed hematuria, proteinuria (5,4 g/24 h), severe anaemia (Hgb 6 g/dl). Antinuclear antibodies and anti-neutrophil cytoplasmic antibodies were negative, C3 and C4 components of complement were normal, anti-glomerular basement membrane (anti-GBM) antibodies were positive at a very high level (827 U/ml, upper limit 10 U/ml). A chest X-ray revealed bilateral lung infiltrates and a high-resolution computer tomography of the chest revealed multifocal alveolar and ground glass opacities, which confirmed a diffuse alveolar hemorrhage (DAH). Therapeutic plasma exchanges (18 treatments) with immunoglobulin infusions, methylprednisolone (1 g i.v. on three consecutive days) followed by oral prednisolone (1mg/kg/day) and mycophenolate mophetil (1g twice daily orally) were introduced.

This treatment resulted in hemoptysis stopping, the normalization of chest X-ray, a gradual amelioration of renal insufficiency (serum creatinine concentration after 30 days of treatment was 1,7 mg/dl), the reduction of proteinuria and hematuria and also the normalization in the level of anti-GBM antibodies. Immunosuppression therapy was continued for 12 months in tapered doses. After this period serum creatinine was 1 mg/dl and urinalysis did not display any abnormalities.

Conclusion

Clinical remission of the severe anti-GBM disease in the young woman was reached using fertility-sparing treatment. Mycophenolate mophetil (2 g per day) together with conventional treatment (TPE, GCS) proved to be effective and without any major side effects in spite of unfavourable factors such as the high serum creatinine level, the necessity of dialysis at the time of diagnosis, the high anti-GBM level and the DAH.

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