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GOODPASTURE'S SYNDROME: TREATMENT AND PROGNOSIS UPDATE

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ABSTRACT

Goodpasture syndrome or anti–glomerular basement membrane disease is one of most infrequent autoimmune disorder that mainly affect pulmonary and renal system that led to severe pulmonary haemoptysis and renal failure. The objective of the present study is to determine the effectiveness on treatment modalities and prognosis of Goodpasture syndrome. Studies were identified by searching (via Pub Med, Science Direct, CrossRef, Research Gate, Google Scholar) data base, only paper published in English are included. After reviewing literature, it is concluded that in Good Pasture Syndrome, if renal system and both pulmonary system is involved then immunosuppressive drugs, corticosteroid and plasmapheresis are considered as therapeutic regimens shows remark effectiveness along with supportive managements like venovenous extracorporeal membrane oxygenation (VV ECMO) and dialysis or renal transplantation. Goodpasture Syndrome prognosis depends on early diagnosis and treatment.

Keywords: Goodpasture syndrome, immunosuppressive therapy, Plasmapheresis, corticosteroid, haemodialysis, venovenous extracorporeal membrane oxygenation (ECMO) and prognosis.

INTRODUCTION

Goodpasture syndrome (GPS) and anti–glomerular basement membrane disease words is used interchangeable which specifically affect lungs and kidneys via the means of autoimmune cause. In this, our body immune system i.e., antibodies mistakenly attack the lung and kidney which cause haemorrhage of lung, glomerulonephritis and renal failure. The client suffering from GPS normally shows joint aches, chills, malaise, fever, fatigue and weight loss generally. If lung is involved - chest pain, haemoptysis, dyspnoea is shown. Whereas if kidney is involved, it shows haematuria, oedema on limbs and face, protein in urine, hypertension and uraemia symptoms were depicted.

American pathologist Ernest Goodpasture of Vanderbilt University was a one who first described the disease in 1919 when he described the case of 18 years old man who died with lung haemorrhage and glomerulonephritis. In 1958, clinical picture of pulmonary renal syndrome

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described by Stanton and Tage and named after Dr. Pasture. Anti–glomerular basement membrane antibodies

Goodpasture's disease is extremely rare condition. Each year in European white and Asian populations, occurrence ratio out of million population, 0.5 to 1.8 cases are diagnosed with Goodpasture syndrome. The disease predominantly affects the age group between 20 to 30 years and 60 to 70 years old in white population. In India, as per case report by Nitin Kumar Dumeeret.al, a 28-year-old male died following massive haemoptysis at Nizam′s Institute of Medical Sciences, Hyderabad in 2006. Furthermore, in Case series study of Prabhakar D et. al conducted retrospective study for one and half year in 2017 at a tertiary centre in North India. Overall, he took 17 clients with Goodpasture's syndrome [2]. After two years later, Srinivasrao Vavilapalli, et.al, performed a clinicomorphological retrospective study for 4.5 years on Anti-glomerular basement membrane disease in Telangana. He took 16 cases of anti-GBM glomerulonephritis in which 2 cases were notes as Goodpasture syndrome [3].

BACK GROUND:

Though Goodpasture syndrome is a rare case, there is different treatment modalities are used in order to treat affected person such as immunosuppressive therapy, Plasmapheresis, corticosteroid, haemodialysis, venovenous extracorporeal membrane oxygenation (ECMO).

Immunosuppressive therapy is mainly use to decrease the activation or efficacy of the immune system. The combination of glucocorticoids plus cyclophosphamide , methylprednisolone, etc were used. Plasmapheresis is a procedure that separate plasma from blood cells by remove harmful antibodies present in plasma and cleanse the blood, thereby protecting the body. Haemodialysis is process in which a machine is used to filters wastes, salts and fluid from blood when person kidney is not working properly. Venovenous extracorporeal membrane oxygenation (ECMO) is a transient mechanical assistance device that permits delivery of sufficient pulmonary support when gas exchange is significantly compromised.

METHODOLOGY:

Studies were identified by searching (via Pub Med, Science Direct, CrossRef, Research Gate, Google Scholar) data base, only paper published in English are included.

The Search term that are used were: Goodpasture's syndrome various treatment modalities such as immunosuppressive therapy, Plasmapheresis, corticosteroid, haemodialysis, venovenous extracorporeal membrane oxygenation (ECMO) and prognosis.

Jae Il Shin et.al conducted a study on pathogenesis then plasma exchange to Imlifidase on Goodpasture Disease. A blended method of glucocorticoids, therapeutic plasma exchange and cyclophosphamide appraised as general management, even with early introduction of treatment, clients who were having terrible prognosis depends on dialysis. Out of fifteen samples, ten patients had bad prognosis become free from dialysis in six months [4].

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Suheyla Apaydin conducted study on apheresis therapy as the management of anti-neutrophil cytoplasmic antibody correlate with advancing Goodpasture syndrome and Glomerulonephritis. When Vasculitis along with antineutrophil and anti–glomerular basement membrane disease which accelerate to inflammation of glomeruli of kidney, plasmapheresis is most important part of the therapies. In advanced stage, the Plasmapheresis is not effective. That's why early treatment should be initiated [5].

However Antoine Huart et.al, conducted a nationwide cohort-based study at French Society of Hemapheresis regarding the consequences of anti–glomerular basement membrane disease. Total one hundred twenty-two clients were selected, out of which twenty-eight were renal involvement, five were pulmonary involvement cases and remaining had both conditions along with Goodpasture syndrome. Each one of them got therapeutic plasma apheresis. One hundred one samples received blended immunosuppressive medication with plasma exchange. Twelve patient received combination of corticosteroid with apheresis and remaining two acquired with cyclophosphamide. With the above treatment, eighty-six percentage of clients showed one year survival rate, although seven clients died with infectious disease. With the help of serum creatinine level, Goodpasture disease clients with renal involvement stay around one year into most effective expect. In addition to checking out new immunosuppressive therapies, are warranted[6].

Jagoda Stojkovikj, et.al, published a case record on anti–glomerular basement membrane disease who initial symptoms appeared and detected after 15 years later. anti–glomerular basement membrane disease is an autoimmune disorder which mainly attack glomerulus membrane of kidneys and air sac of lungs that impaired their functions. Previously, death rate is more than ninety percentage. Due to advancement in therapeutic regimen, blended method of plasma exchange, cortisteroid and immune-suppressive medications boost up the viability up to fifty percentage. Pulmonary involvement of anti–glomerular basement membrane disease is occasional [7].

Antonio Greco, et.al, conducted a clinical on Goodpasture's syndrome. According to the researchers, there were no effective management for the disorder due to infrequency of disease and late detection. For correct treatment, it is very important to diagnose correctly. In Goodpasture's syndrome, body own antibodies strike against kidney and lung. That's why strong corticosteroid and cyclo-phosphamide were opt for definitive management. Especially in advanced stage, plasma apheresis showed great significance [8].

Lorenz Balke, et.al, performed a study on survival rate of Goodpasture's disease with extreme ARDS by using Extracorporeal Life support. Extracorporeal Membrane Oxygenation was inserted. Heparin was utilised. Moreover, daily plasma exchange, immunosuppressive medication and steroid was initiated. Over next ten days, titre of antibodies decreased rapidly without any side effects. Seven days later, Venovenous extracorporeal membrane oxygenation was stopped and twelve days afterward, client was weaned off from ventilator. Subsequently thirty days after hospitalization, client radiograph shown no pulmonary infiltration. Although Extracorporeal Life support guaranteed adequate supply of oxygen in absence of lung damage, given opportunity to manage client's sickness explicitly [9].

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Abdallah Dalabih, et.al, carried out case study of nine years old girl detected with recent emergence of Goodpasture's disease accompanied by acute lug bleeding and failure. Client was well managed by Extracorporeal life support through which they have capable to offer dialysis and plasma exchange. Child released from hospital with adequate pneumonic capability [10].

DISCUSSION:

In Good Pasture Syndrome, if renal system and both pulmonary system is involved then combination management comprising immunosuppressant medications, steroids and plasma exchange showed remark effectiveness along with supportive management like ventilator and dialysis. A similar study was conducted by Franco Dammacco et. al, reviewed 10 anti–glomerular basement membrane disease clients. 6 clients showed both renal and pulmonary involvement. Whereas other 4 cases depicted kidney insufficiencies alone. While analysing all cases, antibodies against body have been detected. Out of 4 cases who received dialysis due to development of glomerulonephritis, 2 patients died after six and eight months later. Ultimately 6 cases received blended therapy of immunosuppressive medications, steroid and plasma apheresis that led to notable rapid healing of kidney physiology with 1 year survival rate. The clinical severity of Goodpasture disease was detected from titre of circulating anti-GBM antibodies. On time detection and acceptance of blended therapy boost up patient condition seventy to ninety percentage of 1 year survival [11].

Though in Goodpasture syndrome, pulmonary system involvement is rare but venovenous life support improved pulmonary function of these client. In support of this, David G Herbert, et.al, performed case study on anti–glomerular basement membrane disease associated with lung distress and kidney failure. Client received extracorporeal life support for long period in absence of anticoagulant therapy. The researchers concluded that client acquired no anticoagulant therapy continuously for twenty days and did not cause any side effects. The client healed completely in absence of lung insufficiencies, machine technical problem, clotting mechanism or dialysis dependent [12].

Anti–glomerular basement membrane disease prognosis depends on early diagnosis and treatment. Similar study carried out by Tobias Lahmer and Uwe Heemann, performed a review study on Goodpasture's syndrome. The researcher found out initial detection of disease led to effective treatment and better outcome. Subsequently, diagnosis of disease in starting stage along with immediate management were vital in this case. Some clients who were on haemodialysis for prolonged period or require renal transplantation also [13].

CONCLUSION:

In Good Pasture Syndrome, if renal system and both pulmonary system is involved then blended therapy comprising of immunosuppressant medications, steroid and plasma exchange shows remark effective along with supportive managements like venovenous extracorporeal membrane oxygenation (VV ECMO) and dialysis or renal transplantation. Goodpasture Syndrome prognosis depends on early diagnosis and treatment.

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