Use Of Noninvazive Positive Pressure Ventilation in a Case of Diffuse Alveolar Hemorrhage Due to Goodpasture's Syndrome

Goodpasture Sendromu Nedeniyle Alveolar Hemoraji Gelişen Bir Olguda Noninvaziv Pozitif Basınçlı Ventilasyon Kullanımı

Alveolar Hemoraji ve NPBV / Alveolar Hemorrhage and NPPV

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Özet

Goodpasture sendromu alveolar hemoraji, progresif glomerulonefrit ve antiglomeruler bazal membran antikorları ile kendini gösterir. Sıklıkla mortal olabilen bir hastalıktır. 18 yaşında erken tanı ile remisyon sağlanan bir erkek olguyu bildiriyoruz. Vaka acil servise atipik pnönomi belirti ve bulguları ve balgamında kan varlığı ile başvurdu. Göğüs grafisi yama tarzında alveolar opasiteler gösterdi (Figür A). Profilaktik makrolid grubu antibiyoterapi ve bronkospazm için bronkodilatör tedavi verildi. Vakada Goodpasture sendromun (GPS)'dan şüphelenildi. Anti-glomerüler bazal membran (AGBM) antikor testi gönderildi. Atipik pnömoni rezolüsyon döneminde olguda massif alveolar hemoraji gelişti. Laboratuar incelemesi proteinüri 20 mg/dl, Anemi 8 g/dl, Hemotorit %25, mikroskobik hematüri 350/HPF gösterdi. AGBM antikor testi pozitif bulundu. GPS tanısı konuldu. Olguya erken dönemde metilprednizolon ve siklofosfamid ile immunsupresif ve plazmaferez tedavisi başlandı. Şiddetli hipoksemi için noninvaziv pozitif basınçlı ventilasyon (NPBV) uygulandı. Plazmaferez tedavisi sürerken hemolitik anemi ve trombositopeni gelişti. Erken başlanan tedavi remisyonla sonuçlandı. Sonuç olarak GPS erken tanı ve NİPBV 'u içeren uygun tedavilerle iyi bir prognoza sahip olabilir.

Anahtar Kelimeler

Goodpasture Sendromu; Noninvaziv Pozitif Basınçlı Ventilasyon

Abstract

Antiglomerular basement membrane antibody disease is manifested by progressive glomerulonephritis, intraalveolar hemorrhage and antiglomerular basement membrane antibodies. It is frequently characterized by mortality. We present a case of a 18 year-old young showing remission by early diagnosis. The patient was admitted to emergency department with symptoms and findings of atypic pneumonia with bloody sputum. Chest radiography detected patchy alveolar opacities (Figure A). An ampric antibacterial treatment was given including macrolide, and bronchodilators because of bronchospasm. The patient was suspected for goodpasture's syndrome (GPS). Anti-glomerular basement membrane (AGBM) antibodies test was send. He developed massive alveolar haemorrhage in the resolution phase of atypic pneumonia. Laboratory examination revealed proteinuria of 20 mg/ dl, anemia Hb of 8 g/dl, hematocrit of 25%, microscopic hematuria of 350 erythrocite /HPF. AGBM antibodies was found as positive. GPS was diagnosed. Early immunosuppressive treatment with pulse methylprednisolone and cyclophosphamide and plazmaferez was started. Noninvasive positive pressure ventilation (NPPV) was used for severe hypoxemia. Haemolytic anemia and thrombocytopenia developed under plasmaphresis treatment. Early treatment resulted with remmission. In conclusion, the current case showed that Goodpasture's syndrome may have a favorable prognosis with early diagnosis and proper treatments including NPPV.

Keywords

Goodpasture's Syndome; Noninvasive Positive Pressure Ventilation

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Introduction

Antiglomerular basement membrane antibody disease is manifested by progressive glomerulonephritis, intraalveolar hemorrhage, and antiglomerular basement membrane antibodies [1-4]. It is frequently characterized by mortality. Immunosupression and plasmaphresis are the mainstay of treatment [2,5]. We report a case of Good Pasture syndrome referred to our pulmonary unit at a university Hospital in August of 2011 and discuss how to manage this kind of difficult cases.

Case Report

After swimming, an 18 – year- old man presented with symptoms or signs of atypic pneumonia upon admission in a hospital, complaining of dyspnea, cough, and sputum with blood. Physical examination revealed bilateral ronchi and rare rales on the chest. Chest radiography detected patchy alveolar opacities (Figure 1A). An ampric treatment was given including bronchodilators that were used for bronchospasm and macrolide. On

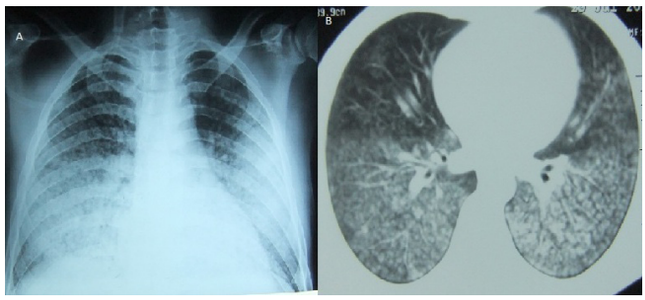


Figure 1. Chest radiography and computed tomography detected bilateral patchy alveolar opacities which were consistent with alveolar hemorrage(A,B).

the 10th hospital day, a sudden onset of pulmonary hemorrhage developed. A computed tomogrophy of the chest showed bilaterally alveolar infiltrates that were consistent with alveolar hemorrage (Figure 1B). Laboratory examination revealed anemia Hb of 8 g/dl, hematocrit of 25%, microscopic hematuria of 350 erythrocite /HPF, proteinuria of 20 mg/ dl. Our patient's findings were clearly positive for anti-glomerular basement membrane(AGBM) antibody. Thus, the patient was diagnosed as Goodpasture's syndrome. The treatment was started with intravenous high dose methylprednisolone and plasma exchange to remove AGBM antibody. After the treatment with pulse methylprednisolone, oral prednisolone (80 mg/day) and plasmaphresis, the pulmonary hemorrhage improved transiently. But the patient started to get worse again 4 days later that corresponds to the 14th day of hospital. The condition of patient deteriorated due to hypoxemia. Saturation of O2 decreased to 70% and respiratory rate increased to 40/min. Noninvazive positive pressure ventilation (NPPV) was started with positive end-expiratory pressure (PEEP) of 5 mm. After implementation of NPPV, the patient's oxygenation promptly returned to normal. Cyclophosphamide pulse therapy was administered. After this treatment, the patient's pulmonary manifestations and pulmonary hemorrhage improved. However, progressive deterioration of renal function (blood urea nitrogen of 67 mg/dl, serum creatinin of 3 mg/dl), thrombocytopenia (98000/ mm3), prolonged PT, and progressive hiperbilirubinemia developed under plasmaphresis. After completing plasmaphresis for 10 days, renal function and hiperbilirubinemia returned to normal levels. The patient was discharged from the hospital on the 35th day of hospital in a good condition.

Discussion

AGBMA disease is manifested by progressive glomerulonephritis, intraalveolar hemorrhage, and AGBMA[1-3]. AGBMD is a rare form of autoimmune glomerulonephritis often accompanied by lung haemorrhage and characterized by circulating and deposited antibodies that bind basement membrane type IV collagen antigenes in the glomerulus and lung alveolus [1]. We reported a case of AGBD which developed under observation and treatment for atypic pneumonia. AGBMD is one of the most important and rare diseases resulting in massive alveolar hemorrhage. Exposure to environmental factors such as viral infections, hydrocarbons, and tobacco smoking may precipitate the disease [5]. Our patient had had both exposure to atypic microorganisms and smoke.

In our case AGBMD was suspected when alveolar hemorrhage developed with the presence of hemoptysis, bilateral interstitial infiltrates on the chest x – ray. AGBMA in serum was positive. Thus, diagnosis was confirmed as Goodpasture's syndrome. Left untreated, patients with anti-GBM disease have a poor outcome, with increased morbidity and/or mortality linked to renal failure or pulmonary hemorrhage. The introduction of combined treatment with corticosteroids, cyclophosphamide, and plasmapheresis in the 1970s changed the approach to therapy and altered what had previously been poor outcomes in patients with the disease [6]. Thus, treatment was based on the corticosteroid, immunsuppressive agents, cyclophosphamide and azathioprin, in addition to plasmaphresis. NPPV was also required for severe hypoxemia due to alveolar hemorrhage. Two studies [7,8] that enrolled various type of patients included patients with Acut Lung Injury/Acut Respiratory Distres Syndrome and reported their results by subgroups. The number of patients in both studies was extremely small (7 and 15, respectively), and no sign suggested NPPV effectiveness. Despite an interesting recent cohort study that suggested that NPPV can be safe inselected patients with ALI/ARDS [9].

These findings show that therapy of cyclophophamide pulse administiration, azathioprin maintenance, and NPPV in case of severe lung injury associated with hypoxemia is effective against Goodpasture's syndrome with alveolar hemorrhage. According to the American Society for Apheresis, plasmapheresis is currently considered a standard and accepted as primary treatment modality for anti-GBM disease (category I indication) [10]. Prolonged plasmaphresis may lead to hyperbilirubinemia and anemia. So physicians should be aware of side effects of plasmapheresis..

In conclusion, the current case showed that Goodpasture's syndrome may have a favorable prognosis when the patients get the respiratory support with NPPV and intensive immunosuppressive treatment from an early stage. In addition, plasmaphresis should be monitorised for possible complications such as hemolytic anemia and trombocytopenia. According to the clinical and laboratory findings, plasmaphresis may be shortened.

Competing interests

The authors declare that they have no competing interests.

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