

Relapse of Granulomatosis with Polyangiitis: A Case Report

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Abstract: *Granulomatosis with polyangiitis (GPA) is a systemic necrotizing vasculitis, which affects small- and medium-sized blood vessels and is often associated with cytoplasmic ANCA (antineutrophil cytoplasmic antibody) or c-ANCA. It was formerly called Wegener's granulomatosis (WG). It mainly involves the upper and/or lower respiratory tract and kidneys. Renal involvement is characterized by rapidly progressive necrotizing glomerulonephritis. It affects multiple organs and organ systems and results in debilitation. Interventions in several areas are required to tackle GPA. They include interventions in healthcare professional and patient education, prescribing practices, diet, medication-taking behavior and lifestyle. With adequate monitoring of patient, morbidity and mortality among GPA patients can be reduced. Continuous learning programmes on GPA may be conducted for both professionals and practitioners for augmenting knowledge related to GPA. Here is a case of a relapse episode of GPA in a 64-year-old patient who developed acute renal failure, respiratory problems and edema and steroid-induced hypertension and oral candidiasis due to corticosteroid use.*

Keywords: granulomatosis with polyangiitis, Wegener's granulomatosis, edema, renal failure, medical renal disease, candida

1. Introduction

Granulomatosis with polyangiitis (GPA) is a systemic necrotizing vasculitis, which affects small- and medium-sized blood vessels and is often associated with cytoplasmic ANCA (antineutrophil cytoplasmic antibody) or c-ANCA. It was formerly called Wegener's granulomatosis (WG). It mainly involves the upper and/or lower respiratory tract and kidneys. Renal involvement is characterized by rapidly progressive necrotizing glomerulonephritis. Limited forms of GPA predominantly affect the upper respiratory tract, whereas generalized forms of GPA include renal manifestations and/or alveolar hemorrhage and/or vital organ involvement with an altered general condition. The combination of immunosuppressant drugs and corticosteroids has converted this typically fatal illness into one in which 80% of patients achieve remission. However, despite considerable therapeutic progress over the last decades, relapses remain frequent (50% at 5 years), and maintenance treatment is now the main therapeutic challenge.¹ The aetiology of GPA is linked to environmental and infectious triggers inciting onset of disease in genetically predisposed individuals. c-ANCA are pathogenic and play an important role in the pathogenesis of this disease, although ANCA positivity is not essential for a clinical diagnosis of granulomatosis with polyangiitis.² Here, we will discuss a rare case of GPA with renal manifestations and respiratory problems.

2. Case Report

A 64-year-old female patient presented with complaints of difficulty in breathing and cough with yellow sputum for 2 days. She also complained of puffiness of face and swelling of both lower limbs for past 1 year. She was on steroids for 1 year. The breathing difficulty was progressive even when at rest. She added that she also had headache. The patient had similar episodes a year ago. She was a known case of Wegener's granulomatosis. She was also diagnosed with hypertension induced by glucocorticoid use. During her

current admission, her blood pressure reading was 140/80 mmHg. c-ANCA testing was performed and it showed positive result. Her USG report confirmed grade I medical renal disease. The findings were indicating acute renal failure. The patient's respiratory and renal conditions, edema and c-ANCA testing result were suggestive of a recurrence of granulomatosis with polyangiitis (GPA) or Wegener's granulomatosis (WG). Her creatine phosphokinase (CPK) level was 37 U/L and total WBC count upon admission was 17620/cu.mm.. Initial treatment comprised of Inj. Ceftriaxone (1g, BID), Tab. Azithromycin (500mg, OD), Nebulizations Salbutamol and Budesonide, Inj. Furosemide (20mg, BID), Tab. Prednisolone (20mg, OD) and Tab. Paracetamol (500mg, TID). Tab. Mycophenolate Mofetil (50mg) was prescribed SOS. Inj. Insulin was initiated 4 days after her admission when her fasting blood glucose (FBG) level rose to 317 mg/dl. 3 days after her admission she developed oral candidiasis, which was attributed to budesonide. Hence, budesonide was withdrawn. A day after her admission, she was diagnosed with hypertension due to steroid-dependence based on her history. Prednisolone was withheld on the second day for this reason and Tab. Doxofylline and Syp. Ambroxol were started. For the oral candidiasis, she was prescribed fluconazole mouthwash. Her symptoms eventually improved and she was discharged on the eighth day with Tab. Prednisolone (5mg, OD), Tab. Mycophenolate Mofetil (500mg, BID), Tab. Pantoprazole (40mg, OD), Tab. Telmisartan (40mg, BID) and Tab. Hydrochlorothiazide (12.5mg, BID) to be taken for a month.

3. Discussion

Between 70 and 90 percent of patients with GPA attain clinical remission with initial immunosuppressive therapy. They are then treated with maintenance immunosuppressive therapy, for a period that is typically 12 to 18 months. However, relapses are common, and some patients have frequent relapses. In addition, manifestations of the relapse may be different from the manifestations of the initial presentation or prior relapses.³ Our case is that of a relapse.

Volume 8 Issue 9, September 2019

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Constant follow-up and patient education are necessary to ensure that the patient is ready for relapse episodes and to reduce the frequency of relapse episodes. Prednisolone can cause sodium retention, resulting in dose-related fluid retention.⁴The principal mechanism of corticosteroid-induced hypertension is the overstimulation of the mineralocorticoid receptor, resulting in sodium retention in the kidney. This results in volume expansion and a subsequent increase in blood pressure. Corticosteroid-induced hypertension may respond to diuretic therapy.⁵ The smallest effective dose and shortest duration of steroid therapy should be used in order to decrease the development of this adverse effect. Apart from the disease itself, the edema could be attributed to prolonged prednisolone use. This sheds lights on the importance of dose-tapering of corticosteroids and regular monitoring of vitals while a patient is on steroid therapy. Telmisartan was prescribed keeping in mind the sodium and water retention properties of corticosteroids apart from edema due to renal failure. Oral candidiasis is commonly observed among patients on corticosteroid nebulization. Regular examination of oral health may prevent diseases in the oral cavity. During the course of therapy, not only total WBC count but also procalcitonin levels should be checked daily to confirm the status of infection. Based on culture-sensitivity reports, the antibiotic prescription should be streamlined. Our patient had a fresh diagnosis of hyperglycemia. This necessitates special interventions in diet, lifestyle and daily activities (including exercise). GPA was once a condition with high mortality rate. However, due to the presence of advanced diagnostic tools and professionals having sound knowledge of its manifestations, the numbers have improved.

4. Conclusion

GPA is a progressive rare disease with high incidence of relapses. It affects multiple organs and organ systems and results in debilitation. Interventions in several areas are required to tackle GPA. They include interventions in healthcare professional and patient education, prescribing practices, diet, medication-taking behavior and lifestyle. With adequate monitoring of patient, morbidity and mortality among GPA patients can be reduced. Continuous learning programmes on GPA or rare diseases may be conducted for both professionals and practitioners for augmenting knowledge related to GPA.

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