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CASE REPORT OPEN ACCESS

Misdiagnosis of Granulomatosis with Polyangiitis: An Unknown Disease for Pakistan

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Abstract

This case represents the misdiagnosis of granulomatosis with polyangiitis (GPA) as pulmonary tuberculosis (TB). This describes the severe consequences of misdiagnosing GPA, which led to this patient acquiring ESRD and requiring dialysis. Greater awareness of granulomatosis with can prevent mistreatment, as inappropriate treatment of this disease with anti-TB drugs can lead to severe consequences. In conclusion, c-ANCA testing is recommended for all patients showing signs and symptoms of TB, who have positive chest X-ray results but a negative sputum test for acid-fast bacilli, to eliminate GPA, as this disease can be fatal for the patient if it remains undiagnosed.

Key words: GPA, Misdiagnosis, Tb, ANCA.

INTRODUCTION

This case represents the misdiagnosis of granulomatosis with polyangiitis (GPA) as pulmonary tuberculosis (TB). Literature suggests that GPA is no more "an unknown disease" in Pakistan.[1-2] Greater awareness of granulomatosis with polyangiitis (also known as Wegener's granulomatosis) can prevent mistreatment, as inappropriate treatment of this disease with anti-TB drugs can lead to severe consequences. Antineutrophil cytoplasmic antibody (ANCA) tests should be conducted for patients with ambiguity before GPA leads to kidney failure.[3] Several cases have been reported as being misdiagnosed between pulmonary TB and granulomatosis, in which the patients were diagnosed with GPA instead of TB.[4-6] However, the opposite occurred in the present case; the patient was misdiagnosed with TB and was treated for TB for six months. The actual disease, GPA, was not diagnosed or treated; thus, the disease became complicated and the patient developed end-stage renal disease (ESRD).

Case Report

This case is presented in 3 stages

Stage 1

A 21-year-old female pharmacy student, weighing 52 kg, presented with a three-month history of recurrent nasal obstruction, catarrh, epistaxis, multiple joint pain and persistent dry cough with vomiting and diarrhea. The patient had a fever of 102°F and a heart rate of 150 bpm. A complete blood count (CBC) showed elevated platelets (748000/µL) and a white blood cell (WBC) count of 19000/µL. Other lab values, including renal function, liver function and serum electrolytes, were within limits. Her apparent health condition indicated tuberculosis (TB)-like signs and symptoms. A chest X-ray revealed slight opacity in the right lung, but the Mantoux test and sputum culture results were negative for TB. Despite these negative results, the patient was prescribed the anti-tuberculosis drug, Myrin-P Forte

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(a fixed dose combination of 275 mg ethambutol, 150 mg rifampicin, 75 mg isoniazid and 400 mg pyrazinamide) at 2 tablets once daily for six months, along with 50 mg vitamin B6 once daily for six months. The patient was advised to continue this treatment for the full 6 months. During treatment, the patient's weight dropped to 45 kg.

Stage 2

The patient's symptoms reappeared after 5 months and 10 days while she was still taking the anti-tuberculosis medication. Lab tests, including cytoplasmic ANCA (c-ANCA), were performed again. The WBC and platelet counts were 14000/μL and 387000/μL, respectively. Vascular lining was prominent on the chest X-ray and pus cells showed 25–27 cells/HPF on the urinalysis. Ultrasound of the renal system demonstrated increased echogenicity of the parenchyma, reduced corticomedullary differentiation and free fluid in the abdomen. Creatinine was 7.54 mg/dl, the estimated glomerular filtration rate (eGFR) was 7 ml/min and urea was 205 mg/dl. The c-ANCA was greater than 100. These results confirmed GPA. The patient was treated with 15 mg prednisolone three times daily for ten days, then 10 mg three times daily for the next 10 days, 20 mg esomeprazole twice daily for 20 days, 10 mg domperidone maleate twice daily for 20 days, 400 mg moxifloxacin once daily for 5 days and metoclopramide injections as needed. The patient's weight increased to 68 kg.

Stage 3

The patient was admitted in emergency condition having advanced renal failure, severe metabolic acidosis and hypercalcemia and was initially managed by hemodialysis and blood transfusion. Further investigations showed bilateral fluffy shadows on the chest X-ray and c-ANCA and anti-glomerular basement membrane (GBM) were positive (double-positive). The patient was treated for GPA and received 1 g salmeterol for 3 days and an immediate 750-mg injection of cyclophosphamide. The patient exhibited drastic clinical improvement (i.e., the chest X-ray became clear). The patient currently has grade III ESRD with c-ANCA1 and creatinine at 7.3 mg/dl. She was advised to undergo routine hemodialysis twice weekly and was prescribed 1000 mg mycophenolic acid in the morning and 500 mg in the evening and 10 mg prednisolone twice daily with ossein mineral complex upon discharge.

DISCUSSION

Friedrich Wegener first distinguished granulomatosis with polyangiitis (also known as Wegener's granulomatosis) from autoimmune systemic vasculitis in 1936.[7] He established that this 'rhinogenic granulomatosis' preferentially exhibits granulomatous lesions in the respiratory tract in addition to vasculitis. Clinical and histopathological studies have revealed that GPA follows a two-phase course. In most patients, the upper and lower respiratory tract is affected first (initial predominantly granulomatosis phase). If untreated, this limited form can progress into a generalized vasculitic disease leading to necrotizing crescentic glomerulonephritis, pulmonary capillaritis and associated constitutional symptoms.[8] When full-blown, the disease is life-threatening and requires highly aggressive immunosuppressive treatment. Histological overlap exists between GPA and tuberculosis. GPA is easier to misdiagnose, but it can now be diagnosed and distinguished from other respiratory tract diseases by widespread awareness of the c-ANCA test.[3]

This case describes the severe consequences of misdiagnosing GPA, which led to this patient acquiring ESRD and requiring dialysis. If proper diagnoses guidelines had been established, this misdiagnosis may have been avoided and the patient would have been diagnosed with GPA when the pulmonary TB results were negative. Although the patient had TB-like signs and symptoms, the disease was misdiagnosed by performing chest X-rays followed by sputum testing for acid-fast bacilli. A standard procedure is needed where if a patient has TB-like signs and symptoms with positive chest X-ray results, but the sputum test is negative for acid-fast bacilli, the patient must undergo c-ANCA testing to eliminate GPA before starting TB treatment. Additionally, undiagnosed GPA leads to ESRD, as occurred in this patient, causing permanent kidney damage. Patient survival is only possible if the patient undergoes multiple hemodialyses monthly.

Obtaining the patient's past medical, family and social histories is necessary before conclusively diagnosing TB. Patients having no past medical, family or social history of TB are unsusceptible to TB, even if they show signs and symptoms and the chest X-ray shows opacity in the lungs.

CONCLUSION

In conclusion, c-ANCA testing is recommended for all patients showing signs and symptoms of TB, who have positive chest X-ray results but a negative sputum test for acid-fast bacilli, to eliminate GPA, as this disease can be fatal for the patient if it remains undiagnosed.

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CONFLICT OF INTEREST

Authors declare no conflict of interest.

ABBREVIATIONS

ANCA: Antineutrophil cytoplasmic antibody; **TB:** Tuberculosis; **ESRD:** End-Stage Renal Disease; **CBC:** Complete blood count; **WBC:** White blood cell; **eGFR:** Estimated glomerular filtration rate.

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