



Wegener Granulomatosis Complicated by Brain Abscess Caused by *Nocardia* spp.

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ABSTRACT

Wegener granulomatosis is a multisystemic disease associated with high mortality rate and characterized by necrotizing granulomatous vasculitis predominantly in the respiratory tract and kidneys. Presence of kidney failure at the time of diagnosis describes a poor prognostic marker. We presented a Wegener granulomatosis case complicated with brain abscess caused by *Nocardia*. (*JAREM* 2016; 6: 126-8)

Keywords: Rapidly progressive glomerulonephritis, acute kidney failure, immunosuppression, infection

INTRODUCTION

Wegener's granulomatosis (WG) is a multisystemic disease that is associated with a high mortality rate and is characterized by necrotizing granulomatous vasculitis, predominantly in the respiratory tract and kidneys. The presence of kidney failure at the time of diagnosis describes a poor diagnostic marker. We present a case of WG complicated with brain abscess caused by *Nocardia* during the treatment.

CASE PRESENTATION

A 66-year-old male patient with complaints of fatigue, elevated fever, weight loss, hemoptysis, and left ankle pain was examined over the course of 2 months in the infectious diseases clinic of a hospital. His PPD test revealed that he was anergic, and no infection or malignancy focus was detected. The patient was admitted to the nephrology unit due to disruption observed in his kidney function tests. Verbal consent was obtained from the patient.

In his laboratory findings, leucocytes were 10,200/mm³, Hb was 9.1 g/dL, Hct was 25.3%, thrombocytes were 287,000/mm³, urea was 225 mg/dL, creatinine was 12.4 mg/dL, CRP was 52 mg/L, and sedimentation rate was 92 mm/h; proteinuria was observed in urinalysis, and plenty of erythrocytes were present in his urine sediment. Hemodialysis was started. He was positive (+) for anti-neutrophil cytoplasmic antibodies (c-ANCA), while complement levels were within the normal range. On observing his kidney size to be within the normal range in his ultrasonography, kidney biopsy was performed with the pre-diagnosis of "rapidly progressive glomerulonephritis". Along with the elevation of sedimentation, the presence of c-ANCA positivity, lower respiratory tract involvement, and the compatibility of kidney biopsy findings with Pauci-immune Type III crescentic glomerulonephritis, the patient was diagnosed with WG. He received 500 mg methylprednisolone (Prednol; Mustafa Nevzat Pharmaceutical, İstanbul, Turkey) pulse treatment for three consecutive days, followed by 750 mg intra-

venous (IV) cyclophosphamide (Endoxan; Eczacıbaşı-Baxter Hospital Products, İstanbul, Turkey) and five sessions of plasmapheresis. In the first week of treatment, creatinine levels (creatinine: 1.2 mg/dL) dropped down. The recommendation of oral prednisolone at a dose of monthly 750 mg IV cyclophosphamide+1 mg/kg was registered in his polyclinic follow-up. In the third month of his treatment, the patient was admitted with complaints of coughing, elevated fever, and left side pain. Bronchopneumonia was detected in his chest computed tomography (CT) (Figure 1). IV 2×1000 mg/day ceftriaxone (Desefin; Deva Holding, İstanbul, Turkey) and 500 mg/day levofloxacin (Tavanic; Zentiva Medical Products, Lüleburgaz, Turkey) were started, and the steroid dose was lowered. On the second day of his treatment, the patient developed fatigue and loss of balance, and his cranial magnetic resonance (MR) imaging showed a brain abscess (Figure 2). Surgical treatment was not recommended because the abscess size was larger than 1.5 cm. Samples were obtained via lumbar puncture for culture. Antibiotic treatment was expanded to meropenem 3×1000 mg/day (Meronem; AstraZeneca, Macclesfield, England) + TMP/SMX (Bactrim; Deva Holding, İstanbul, Turkey). Because *Nocardia* spp. reproduced in his cerebrospinal fluid culture, the treatment was planned as 8 weeks of parenteral 2×1000 mg ceftriaxone + 2×160/800 mg trimethoprim/sulfamethoxazole (TMP/SMX), followed by 6 months of oral TMP/SMX and monthly cranial MR controls. Immunosuppressive treatment directed at WG was limited to 16 mg/day methylprednisolone. At present, the controls are continued in the fourth month of the treatment, and the size of patient's brain abscess is decreasing and his renal functions (serum creatinine level: 1.3 mg/dL) are stable.

DISCUSSION

WG is a necrotizing granulomatosis vasculitis with an unknown etiology and a prevalence of 3/100,000; it is observed in the fifth decade regardless of sex (1, 2). Its main symptoms are coughing, dyspnea, and hemoptysis. More than 90% of patients have





Figure 1. Chest CT-bronchopneumonic infiltration
CT: computed tomography

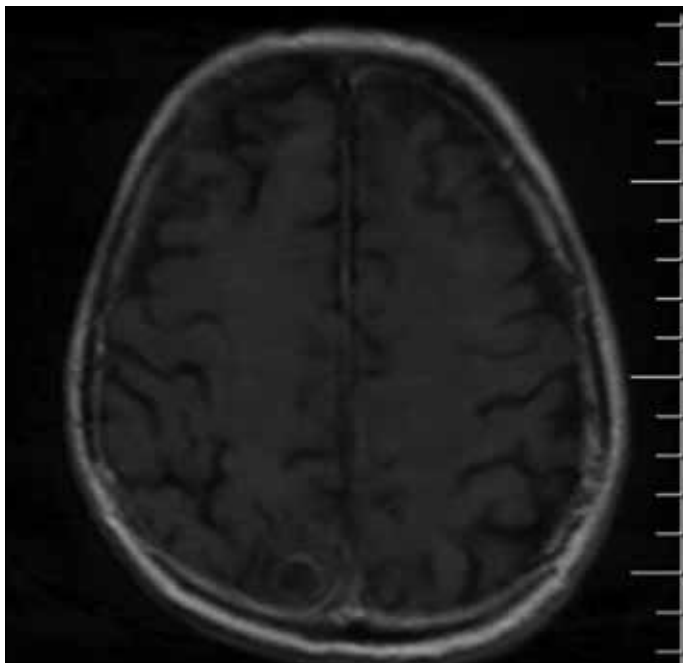


Figure 2. Cranial MR-brain abscess
MR: magnetic resonance

bilateral, multiple nodules and parenchymal infiltrations of varying sizes, and lung imaging displays abnormalities (3). Pulmonary hemorrhage along with kidney function disorder increases the mortality rate.

Glomerulonephritis develops in approximately 75% of WG patients. Histologically, focal necrotizing glomerulonephritis and crescentic glomerulonephritis are observed, and they can rapidly lead to kidney failure (4). Hematuria, proteinuria, and erythrocyte cylinders are observed as urinary findings. Two-thirds of patients

can have musculoskeletal complaints. Our patient had findings related to the kidneys, lungs, and skeletal system.

More than 90% of patients are c-ANCA positive (5, 6). In case of lung involvement accompanying hematuria/proteinuria, kidney or lung biopsy may need to be performed for a definitive diagnosis. Despite the risk of hemorrhage in our uremic patient, kidney biopsy was performed in order to plan for the immunosuppressive treatment, given his age. Patients who require dialysis have been reported to benefit from an aggressive treatment (7).

Fauci et al. (2) demonstrated in their study that a combination therapy of oral steroid + cyclophosphamide leads to 90% remission. However, because of the adverse effects of medicines, cyclophosphamide treatment is recommended to be administered parenterally (1, 3). We administered 0.5 g/m² single-dose IV pulse cyclophosphamide once a month for 3 days and 0.5 g/day IV methylprednisolone, followed by 1 mg/kg/day oral prednisolone follow-up treatment.

In a study comparing the effectiveness of plasmapheresis and pulse steroid in patients with kidney failure, the group that underwent plasmapheresis was reported to develop fewer late-stage kidney failures (8). Deaths are determined to occur within 3 months after the start of the treatment of this disease, which has a high mortality rate. In total, 54% of reported deaths are associated with infections. While the treatment of our case was ongoing with the diagnosis of lobar pneumonia that developed in the third month of immunosuppressive treatment, loss of balance occurred, which led to performing cranial MR that showed brain abscess in the parietal region. Reproduction of *Nocardia* spp. in the culture taken by lumbar puncture established the diagnosis. *Nocardia* is a rare, mortal, and opportunistic gram-positive bacterium that is particularly observed in cases with cellular immunity disorder (9, 10). Sole central nervous system involvement is very rare (9%) (9, 10). Slow growth in culture may result in mortality due to delayed diagnosis and treatment.

CONCLUSION

Patients who receive immunosuppressive treatments must be closely followed up as high-risk groups with regard to opportunistic infections, and the treatment of infections must be performed energetically.

Informed Consent: Verbal informed consent was obtained from patient who participated in this case.

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