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ANCA-Associated Vasculitis Overlaps with Acute Cerebral Infarction: A Case Report

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Abstract

Antineutrophil Cytoplasmic Antibody (ANCA) Associated Vasculitis (AAV) is a group of multisystem autoimmune small vessel diseases. We report a case of a 68-year-old woman who initially presented with bilateral numbness of her hands and feet over past 20 days and cranial MRI before hospitalization showed acute cerebral infarction in left basal ganglia, so was admitted to the department of neurology. Her symptoms persisted and blood tests indicated heart failure, renal impairment and evidence of an inflammatory response. Chest Computed Tomography (CT) scan and ANCA testing confirmed a diagnosis of AAV renal injury and then was transferred to the department of nephrology. She was treated with blood purification for four sessions, glucocorticoid and cyclophosphamide therapy at which time her kidney function had improved. This case study illustrates that the clinical manifestations of AVV are complex, varied, and prone to misdiagnosis.

Keywords: ANCA; Vasculitis; ANCA-associated vasculitis; Acute cerebral infarction

Introduction

Anti-Neutrophil Cytoplasmic Antibody (ANCA)-Associated Vasculitis (AAV) affects small and medium-sized arteries and can progress to both kidney and lung involvement due to production of autoantibodies against the antigens Myeloperoxidase (MPO) and Proteinase 3 (PR3) [1]. AAV comprises Microscopic Polyangiitis (MPA), Granulomatosis with Polyangiitis (GPA) and Eosinophilic Granulomatosis with Polyangiitis (EGPA), characterized by necrotizing inflammation of the small blood vessels [2]. Renal involvement is a common and severe complication of AAV as it can cause requirement of Renal Replacement Therapy (RRT), End-Stage Renal Disease (ESRD) or death [3,4].

Case Presentation

A 68-year-old female patient was admitted to our hospital for bilateral numbness of her hands and feet. Over 20 days previously, the patient experienced bilateral numbness of her hands and feet and unstable walking, without dizziness, headache, limb weakness, speech disorder, blurred vision, and disorders of consciousness, and visited a local clinic. She was treated with Chinese Medicine, however her symptoms continued to worsen and developed unsteadiness walking. At 2 days before admission, she visited the local hospital and the cranial MRI examination revealed acute cerebral infarction in left basal ganglia. Therefore, she was referred to our hospital and admitted to the neurology department. Review of her medical history showed that she had hypertension for last 25 years and had been prescribed once daily reserpine 0.25 mg. Besides, the patient had hyperthyroidism for 8 years and were treated with 50 μg/d of levothyroxine sodium. And she had a history of rheumatoid arthritis for five months not received treatment and anemia for two months being prescribed ferrous sulfate 600 mg and folic acid 12 mg daily.

On physical examination, her body weight was 65 kg, height 160 cm, Body Mass Index (BMI) 25.4 kg/m^2. Her body temperature was 36.4°C, systolic/diastolic blood pressure was 135/85 mmHg, pulse rate was 78 beats/min and oxygen saturation decreased (70-90%). On the first day of admission, she developed presented with fever, accompanied by cough and sputum. The maximum body temperature was 38.2°C. A physical examination did not show any skin rash and superficial lymph nodes were not palpable. The lungs were clear to auscultation bilaterally with rales, and the heart rate (78 beats/min) was regular without murmurs, gallops, or rubs.

The abdomen was soft without tenderness or rebound pain and the liver and spleen were not palpable. The lower extremities had pitting edema. Laboratory studies showed the following:

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Proteinuria (1+), a urine RBC count of 291 cells and the WBC count of 72 cells per high-power field, no casts in the urine, a blood WBC count of 13.52×10^9/L, a hemoglobin level of 75 g/L, a platelet count of 328×10^9/L, a serum albumin level of 25.25 g/L, a blood urea nitrogen level of 10.42 mmol/L, a SCr level of 181.0 μmol/L, a B-type Natriuretic Peptide (BNP) level of 1016.00 pg/ml, a high-sensitivity cardiac troponin I level of 23.6 pg/ml, a C-reactive protein level of 112.57 mg/L, and an erythrocyte sedimentation rate of 88 mm/h. Laboratory findings for anti-hepatitis B antibody, and hepatitis C virus antibody were negative. Her rheumatoid factor level was 142.00 IU/ml, anti-nuclear antibody was 1:1000 (cytoplasmic fiber type) and MPO-ANCA antibody was positive. PR3-ANCA and anti-GBM antibody were negative. Chest CT showed multiple patchy high-density shadows and a little pleural effusion in both lungs, and an abdominal ultrasound B-scan revealed diffuse changes in both kidneys. Based on the medical history, symptoms, signs and auxiliary examination results, the patient was suspected to have ANCA-associated vasculitis affecting multiple organs, such as the heart, lung, kidney and nervous systems four days after admission and immediately transferred to the department of nephrology. During the four days, she developed presented with fever, accompanied by cough and sputum. Receiving symptomatic and supportive treatment of diuresis, anticoagulation, anti-infection and albumin supplementation.

On hospitalization in department of nephrology, laboratory tests revealed the follows: Phase-contrast microscopy analysis showed 80% of urine RBC morphology was normal, a blood WBC count of 14.04×10^9/L, a hemoglobin level of 77 g/L, a platelet count of 312×10^9/L, a serum albumin level of 23.34 g/L, a blood urea nitrogen level of 12.80 mmol/L, a SCr level of 316.1 μmol/L, a NT-pro BNP level of 31308.2 pg/ml. Laboratory findings for MPO antibody was strong positive and P-ANCA was positive. PR3 and c-ANCA were negative. Her complement C3 level were 0.7 g/L (normal range: 0.9-1.8 g/L) and complement C4 level was normal. Serum Immunoglobulin (Ig) G level was 18.60 g/L (normal range: 7-16 g/L). IgA and IgM antibody levels were normal.

ANCA - associated vasculitis was further diagnosed based on past medical history, physical examination, laboratory tests, ultrasonography and CT. We commenced plasma exchange therapy, and 40 mg of methylprednisolone per day was administered intravenously considering serious lung infection. Two days later, serum creatinine increased to 319.7 μmol/L so that plasma exchange therapy was continued, the dose of methylprednisolone was increased to 500 mg per day intravenously for 3 days, Besides, she was treated with cyclophosphamide 0.6 g. Finally, her serum creatinine stabilized at 250 μmol/L and clinical symptoms significantly improved over the total 12-day treatment. She was discharged from the hospital under continued oral prednisone 35 mg/d and therapy and scheduled for follow-up outpatient review two weeks later.

Discussion

AAV is a group of multisystem autoimmune small vessel diseases. The conditions are characterized by formation of granulomas and inflammation of small arteries, arterioles, venules, and capillaries. Untreated, the disease can be fatal and it is often misdiagnosed because of its relative rarity and non-specific presentation [5]. The most commonly affected body systems include the upper airways, lungs, kidneys, eyes, and peripheral nerves, although almost any part of the body can be affected [6]. Indeed, multisystem involvement is often the clue for diagnosis [5].

Investigations should include laboratory assessments of inflammatory markers, kidney function and serological testing for ANCA [6]. ANCAs have a key role in the pathogenesis of AAV because they can directly or indirectly activate inflammatory cells such as neutrophils and promote the release of various inflammatory factors, which results in injury to small vessels [7,8]. In addition, a chest X-ray should be taken and CT or Magnetic Resonance Imaging (MRI) may be required to assess the chest, brain and head and neck structures in detail. It is recommended that a kidney biopsy should be considered to confirm the diagnosis [9].

The characteristics of the case presented here were in accordance with previous studies [10]. Initially, the patient presented with bilateral numbness of her hands and feet and unstable walking, all symptoms that could be attributed to acute cerebral infarction. The persistence of her symptoms prompted her to visit to our hospital for further tests. Her urinalysis was positive for protein, albumen and blood tests showed renal impairment and evidence of an inflammatory response. Renal biopsy was not attempted due to poor performance status of the patient. In conjunction with all symptoms, signs, radiographic evidence and the results of clinical laboratory tests, we confirmed diagnosis of AAV renal injury. Considering the severity of the patient's pulmonary AAV and heart failure, plasma exchange and was glucocorticoid therapy administered together with cyclophosphamide, and achieved ideal therapeutic effects. However, the patient still needs further disease assessment and follow-up treatment.

In summary, early diagnosis and treatment are vital for improving the prognosis of patients with AAV. The disease is prone to misdiagnosis because it often involves several body systems with specific clinical features. Early diagnosis and initiation of effective treatment will lead to be a better outcome for affected patients [11].

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