RHEUMATOLOGY RESEARCH JOURNAL

Vol. 7, No. 4, October 2022
Webpage: http://rheumres.org/
Email: rheumres@gmail.com
ISSN:2476-5856
doi: 10.32592/RR.2022.7.4.111

Case Report Open Access

Guillain–Barré syndrome in a patient with Wegener's granulomatosis: A rare case

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Peripheral neuropathy may be reversible or permanent. Hence, we would like to present a 38-years-old man without any previous medical history admitted to ShahidSadoughi Hospital in Yazd in terms of reduced force of all four limbs, shortness of breath, cough and hemoptysis. Electromyography and nerve conduction velocity study was performed after the para-clinical examinations and Guillain–Barré syndrome was considered first diagnosis. Regarding the pulmonary symptoms, sinusitis and bilateral alveolar opacities, and positive anti-neutrophil cytoplasmic antibodies, Wegener's granulomatosis was suggested as a potential diagnosis and more steps were performed. This patient has been verified to have concurrent increasing limb weakness, also known as Guillain-Barré syndrome.

Keywords: Anti-neutrophil Cytoplasmic; Wegener's granulomatosis; Guillain–Barré syndrome; Vasculitis

Introduction

Peripheral neuropathy affects nerves beyond brain and spinal cord [1]. Neuropathy affects motor, sensory, or autonomic nerves resulting in different symptoms. Multiple nerves may be involved simultaneously. The peripheral neuropathy duration ranges from days to years. Probable etiologies are diabetes, leprosy [2,3], nutrient deficiency, celiac disease, medications, such as metronidazole or ciprofloxacin, trauma, ischemia, radiation therapy, excessive alcohol consumption, some viruses and autoimmunity [4,5]. Guillain-Barrésyndrome (GBS) is an acute immune mediated polyneuropathy leading to rapidly progressive ascending weakness and can be diagnosed via the neurological examinations, such as nerve conduction [6]. Wegener's granulomatosis is an uncommon multisystem illnessis characterized by widespread localized necrotizing vasculitis and granulomatous inflammation of the upper and lower respiratory tract. Respiratory symptoms and the presence of antineutrophil cytoplasmic antibodies (ANCA) point to the diagnosis of Wegener's granulomatosis, while the lack of ANCA does not exclude the diagnosis [7]. The occurance of these two uncommon diseases together is considered a rare event and and challenging.

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Received: 17 July 2022; **Accepted:** 03 Augest 2022

Case presentation

A 38-years-old man without any previous medical history was admitted to ShahidSadoughi Hospital in Yazd on February 7, 2021 in terms of shortness of breath, cough, hemoptysis and reduced force of all four limbs. Examinations revealed that the patient had a positive polymerase chain reaction (PCR) for Covid-19 two months ago and was treated with antibiotics, steroids, Remdesivir (8 doses), and 14 vials of mesenchymal stem cells. During hospitalization and in terms of the reduction of lung noise in both clinical examinations, Chest X-ray (Figure 1) and computed tomography (CT) scan (Figure 2) were requested for the patient. A left pleural effusion and widespread bilateral alveolar opacity were also seen on the CT scan. In response, a left side thoracostomy and pleural fluid diagnostic test were carried out. Analysis of the pleural fluid revealed both acute and long-term inflammatory cells. The echocardiography was normal. We also focused on the reduction of force in all four limbs that was progressive, and ascending and was confirmed in physical examination. Therefore, an electromyography and nerve conduction velocity (EMG-NCV) study was performed for the patient. Based on the patient history, physical exam and EMG-NCV findings (acute axonal type sensory-motor polyradiculo-neuropathy) GBS was considered first the diagnosis. Other paraclinical finding to find out the cause of the patient's neuropathy was done. Brain MRI was normal. Lumbar puncture was done, and result showed normal finding. (WBC = 0, RBC = 0, GLU = 98, LDH = 33, Protein = 55). A paranasal sinus CT scan (Figure 3) was carried out on the patient since he also had a history of sinusitis. Due to the patient's significant bilateral alveolar opacity and the findings of the CT scan, which showed evidence of increased thickness in the sphenoid and ethmoid sinuses and raised the possibility of inflammatory autoimmune illnesses, vasculitis was investigated in the patient. The results were interestingly amazing, c-ANCA was positive twice with high level, ESR = 108, and FANA = 1/160. Ceratine phosphokinase and aldolase were in normal range. Anti-double stranded DNA was negative. Based on hematuria (RBC: 15-20 in high power field) and proteinuria inthe patient's urine analysis, the urine was collected in 24 hours and the amount of proteinuria was measured as 826 mg. Thus, based on the results obtained, Wegener's granulomatosis was proposed as a possible diagnosis. Finally, the patient was diagnosed with GBS and Wegener's granulomatosis and treatment was started. The induction therapy was performed with 16 sessions of plasmapheresis, three consecutive daily intravenous (IV) pulses of methylprednisolone (1 gr) and one pulse of cyclophosphamide (1 gr). Maintenance therapy was performed with prednisolone (1 mg/ kg/day), and monthly IV cyclophosphamide (1 gr) pulse.

Discussion

Granulomatosis with polyangiitis (GPA) [8-12], is a granulomatous vasculitis affecting small and medium-sized vessels, predominantly upper respiratory tract, kidneys and lungs [13]. Involvement of nose and paranasal sinuses is very rare [14]. Cavitary nodules, interstitial infiltrates, alveolar hemorrhage and bronchial stenosis can be in terms of lung involvement in GPA. Neurologic involvement in GPA occurs in 22–54% of patients, including cerebritis, cranial nerve palsy, peripheral neuropathy and cerebro-vascular events [15].

Diagnosis is aided by the detection of ANCA, although positive results are not conclusive and

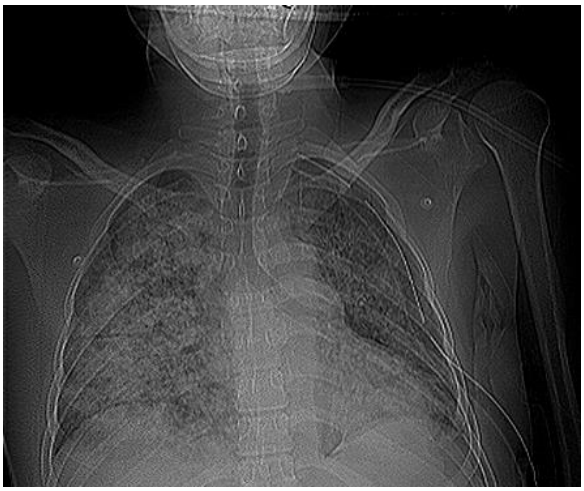

Figure 1: Chest X-ray shows bilateral alveolar opacity is present, left side chest tube is seen

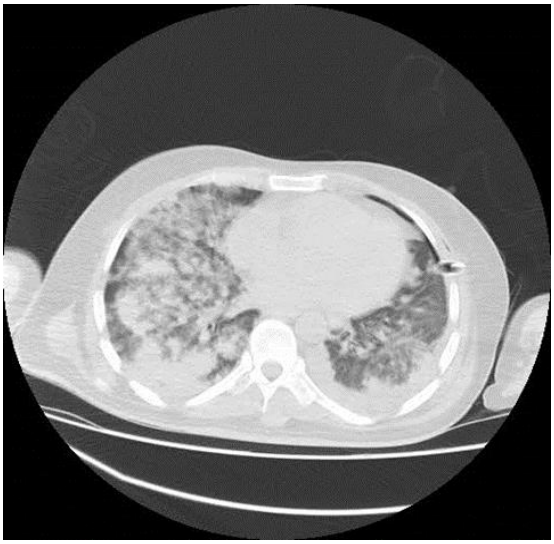

Figure 2: Chest CT scan, lung and mediastinal view, bilateral alveolar consolidation with air-bronchogram is noted. Left side plural effusion and chest tube is seen. Left side pneumothorax is seen)

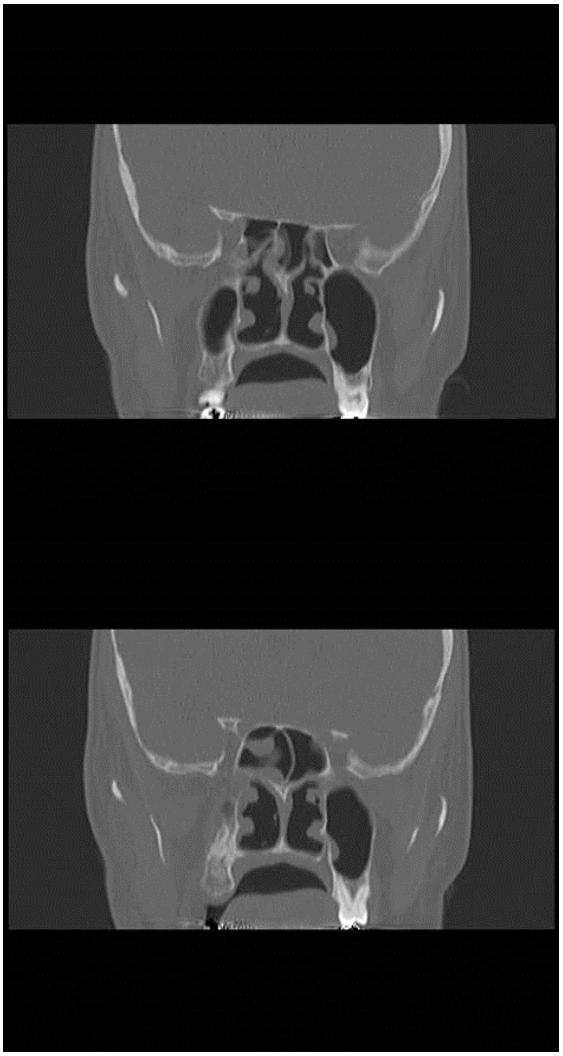

Figure 3: Paranasal sinuses CT scan. In coronal view shows mucosal thickening in ethmoidal air cells and sphenoid sinuses)

negative results do not completely exclude the disease. More than 90% of GPA patients are ANCA-positive [17]. ANCAs that target neutrophil proteinase 3 are linked to Wegners disease [18]. Hence, given that we are currently in an epidemic caused by Covid-19 and based on the patient's history and initial symptoms, and CT scan images, the first diagnosis that came to mind was re-infection with Covid-19 or complications from a previous infection. In addition, due to the fact that the patient was treated with mesenchymal stem cells, the complication of this type of treatment were considered a possibility. Hence, Sairam Atluri et al., Syed Shadab Raza et al., Jiajia Chen et al., report that treatment with MSCs infusion exhibited an excellent safety [19-21],Belmont et al. believed that injections of cellular products cause bvascular inflammation via the cytokines and finally lead to vasculitis [22]. Marsot-Dupuch et al. in 2002 reported trigeminal neuropathy as a rare complication of the disease [23]. Paul Dagum et al. in 1998 reported facial nerve palsy as a rare complication of the disease [24]. Belden et al. in 1993 reported optic neuropathy as neurologic finding of GPA [25]. Wayne P. Foster et al. in 1995 reported Optic neuropathy in Wegners' granulomatosis [26]. In the present case report, the patient who had only a previous history of sinusitis from the probable GPA findings, after the recent infection with Covid 19, developed peripheral polyneuropathy and presented with a clinical picture of weakness in 4 limbs. Concurrent progressive limb weakness, known as GBS, is a rare finding in Wegener's disease that was confirmed and treated in the present patient.The same 88-year-old woman was described by Sargios et al. as having low back discomfort, a persistent cough, and one episode of pinkish sputum. As the condition progresses, the woman develops respiratory failure. Finally, the patient had EMG-NCV, and GBS was verified when PANCA and proteinase 3 tests came back positive and highly positive, respectively [27]. Because of Covid-19 epidemic, the attention of mostly physician in the world is on this disease, and its complications and their treatment. This fact has reduced the diagnosis of ANCA-associated vasculitis [28], so it seems that this

issue, and the problems of delay in diagnosis should be given more attention.

Acknowledgment

Not applicable.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this study.

Funding

There is no funding.

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