

CASE REPORT

OPEN ACCESS

A peripheral neuropathy with unusual progression in Wegener's granulomatosis

Nayef Mohammed Kazzaz, Scott Louis Massien

ABSTRACT

Introduction: We present a case of Wegener's granulomatosis (WG) with unique sequence of progression into a rare presentation of peripheral neuropathy; previously there has been one similar case. However, instead of the two years lag after sinusitis development our case manifested with peripheral neuropathy in a quick fashion. Also we have observed a chronological sequence of progression from infectious process into Wegener's granulomatosis. **Case Report:** We present a case of a 66-year-old Caucasian male previously healthy who presented initially with symptoms of sinusitis and pneumonia, which in a span of three months presented with peripheral neuropathy advancing to left foot drop. **Conclusion:** In Wegener's granulomatosis, the symptoms of most prominence might not necessarily be the classical presentation of upper or lower respiratory involvement but could be neurological and does not always follow the course of chronic involvement before advancing to severe neurological consequence such as foot drop. In addition, the hypothesis that infections could have contributory role in development or

is a factor in Wegener's granulomatosis is observed here in the chronological sequence of events from pneumonia to manifestation of the disease and nonetheless the seroconversion of C-ANCA to positivity.

Keywords: Wegener's granulomatosis (WG), Granulomatosis with polyangitis, Small vessel vasculitis, Peripheral neuropathy

Kazzaz N, Massien S. A peripheral neuropathy with unusual progression in Wegener's granulomatosis. International Journal of Case Reports and Images 2013;4(7):341–344.

doi:10.5348/ijcri-2013-07-327-1

INTRODUCTION

We present a case of Wegener's granulomatosis (WG) with a unique sequence of progression into rare presentation of peripheral neuropathy, also an elegant chronological relation of WG with an infectious process. Wegener granulomatosis is a syndrome that involves the sinuses, lungs and kidneys; which are part of the diagnostic criteria in addition to special granulomatous changes on biopsy and C-ANCA. Our case qualifies for Wegener's granulomatosis based on the European Medicines Agency algorithm. Of importance, WG annual incidence has been reported to be 11.3 per million, with the typical age of presentation between age 35 and 50 [1].

First unique aspect was the rapid sequence of disease progression observed; in contrary to the usual slow progression of WG over a year before developing peripheral neuropathy passing through the common manifestation of WG. In span of three months,

Nayef Mohammed Kazzaz¹, Scott Louis Massien²

Affiliations: 1MD, PGY-2 Resident, Internal Medicine Department, University Hospitals Case Medical Center, 11000 Euclid Ave, Cleveland; 2MD, Scott Massien MD, ABIM, Clinical Instructor, Internal Medicine, University Hospitals Ahuja Medical Center, Kathy Risman Pavilion, 1000 auburn drive suite 110, Beachwood.

Corresponding Author: Nayef Kazzaz, Internal Medicine Department, University Hospitals Case Medical Center, 11000 Euclid Ave, Cleveland, OH44106; Ph: (216)262-5793; Email: nkazzaz86@gmail.com

Received: 10 September 2012

Accepted: 16 November 2012

Published: 01 July 2013

presentation of vasculitic peripheral neuropathy was evident as the main symptom of concern. There has been only one case in literature that shared the mononeuritis multiplex presentation but was associated with prolonged course of sinusitis over two years initially [2, 3].

Second point of importance, is that our case further supports the theory of infectious disease being a culprit in Wegener's pathogenesis. As the initial presentation, patient had lung involvement that was suggestive of infectious process that advanced quickly within three months span to WG full picture. Also worth mentioning, resolution of the cavitary lung lesion in a quick fashion by antibiotics supports the hypothesis. However, other explanation that the antibiotic used could have had anti-inflammatory abilities, yet still this is not a favorable explanation giving the pauci-immune process of WG in oppose to other autoimmune rheumatologic disease.

CASE REPORT

We present a case of a 66-year-old Caucasian male previously healthy, who presented initially with symptoms of cough, greenish sputum, dyspnea, rhinorrhea and maxillary pain with a chest computed tomography (CT) scan significant for bronchiectasis, centrally cavitary lung nodules and conglomerative infiltrate in the left lower lobe, mediastinal and left hilar adenopathy. Later he was treated with antibiotics as a case of community acquired pneumonia with symptomatology improvement.

After three months the patient sustained an avulsion fracture of the fifth toe then presented to us with foot drop. On further questioning, patient stated course started with bilateral tingling of the legs then shooting pain down the left leg. Also, History of fatigues, myalgia, and bilateral morning hand stiffness that lasts less than 30 minutes was obtained. Patient was started on low dose prednisone—although his initial ANCA was negative—with improvement of the latter constitutional symptoms but not the foot drop. Electromyography was consistent with diagnosis of mononeuritis multiplex.

The persistence of foot drop along with history of 15 pounds weight loss and investigations significant for leukomoid reaction of 38 thousand white cells—without left shift—warranted repeating CT scan and bronchoscopy to rule out chronic infectious process such as pulmonary tuberculosis and histoplasmosis. Repeat CT scan showed interval development of combined interstitial and ground-glass opacities in peripheral portions of the right upper lobe, tree-in-bud opacifications in the right middle and lower lobe, resolution of previously described bronchiectasis in the left lower lobe with cavitary lung lesion resolution.

Bronchoalveolar lavage washing for tuberculosis Ziehl–Neelsen stain, fungal cultures, viral cultures and cytology were negative. Of significance, Bronchoalveolar lavage returned bloody and cytology study had iron stain positive in 66% of alveolar macrophages, which indicate prior bleeding engulfed by the white blood cells

[4]. In the setting of these new studies especially the negativity of infectious process and the new ground glass opacities on CT scan warranted repeated WG work-up.

The new work-up revealed an ANA 1:320 speckled with positive 91 conversion of C-ANCA as 1:40 and acute kidney injury. Thereafter, sural nerve and gastrocnemius muscle biopsies were significant for lymphoplasmacytic vasculitis involving the epineurial arterioles and scant perivascular lymphocytic inflammation involving the perimysial blood vessels, respectively. Pulse steroid treatment for three days was started and orthotics was supplied to aid in patient daily mobility and activities. Patient was discharged with prednisone as treatment of his newly diagnosed ANCA vasculitis. On follow-up, the laboratory results of his renal function was significant for persistent raised blood urea nitrogen and creatinine around 35 and 1.47, respectively. In addition urinalysis was significant for ten red blood cells and positive 2 granular casts. Given history of sinusitis, cavitary lung nodules that resolved into ground glass opacities and bloody Bronchoalveolar lavage return, C-ANCA positivity, red cell casts in urine; patient was deemed WG and therefore starting cyclophosphamide, with later improvement of his foot drop, in addition patient regained weight [1, 4].

DISCUSSION

This case warrants attention in several aspects of its presentation; mainly the peripheral neuropathy as a symptom of main concern, most prominence and its early presentation in the course of the disease. Also we show a chronological relation of WG with infectious etiology especially given improvement of our patient's cavitary lung lesions and symptomatology with antibiotic treatment alone. Thereafter, the patient progressed to development of C-ANCA positivity and fully manifested his disease although in a subtle way.

In our review of literature, we found that there has been only one case report which shares the presentation of foot drop that was in a biopsy proven vasculitis that however does not share the quick progression; as their patient had history of chronic sinusitis two years prior to the peripheral neuropathy development [2]. Their case was consistent with the Luigi Cattaneo et al. study that declared in WG among other small disease vasculitis peripheral neuropathy usually develops after a prolonged course of sinusitis, lung manifestations or renal involvement priors to neurological disease. However, our case shares the rare feature of being severe neurological manifestations that is usually more likely with other vasculitic diseases rather than WG [3]. With this information, our report here should warrant physicians to consider in peripheral neuropathy and other neurological manifestations, the possibility of small vessel vasculitis and search for other subtle symptoms of these diseases as they might not be the primary concern for the patient but definitely aid in both

diagnosis and treatment.

The second point is more of an academic importance, as there has been hypothesis about infection being a triggering factor or even a causing entity in WG. This hypothesis has been speculated specially with studies making a connection between staph aureus nasal carriage and small vessel vasculitis [5, 6]. However, up to our review there has been no chronological sequence of infection development that led in a short span of three months to the conversion of C-ANCA to positivity. Cavitory lung nodules in the CT scan are consistent with WG presentation especially with resolution into ground glass opacity, as it has been reported in prior studies. In addition, the hemorrhagic staining of macrophages on the Bronchoalveolar lavage further support the diagnosis of small vessel vasculitis. So such resolution of pulmonary manifestation of WG achieved solely with antibiotics, might indicate the direct relation between infection and WG and even stir up the question of prophylactic antibiotic treatment in patients with family history of autoimmune disease or encourage development specific vaccinations for such a population [4, 7–10].

CONCLUSION

Our case is unique in the aspect that it is the first case that shows the quick progression from sinusitis and lower respiratory tract infection in three months to a rare presentation of peripheral neuropathy in the setting of Wegener's granulomatosis. In addition, we observed a chronological relation of infectious process culminating in the development of full clinical manifestations of Wegener's granulomatosis, which is further supported by conversion of C-ANCA to positivity from an initial negative test. Both aspects will assist us in understanding of Wegener's and will prompt physicians to consider such diagnosis early in the disease process and thereafter earlier initiation of treatment.

Author Contributions

Nayef Mohammed Kazzaz – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Scott Louis Massien – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

Copyright

© Nayef Mohammed Kazzaz et al. 2013; This article is distributed under the terms of Creative Commons Attribution 3.0 License which permits unrestricted use, distribution and reproduction in any means provided the original authors and original publisher are properly credited. (Please see www.ijcasereportsandimages.com/copyright-policy.php for more information.)

REFERENCES

1. Watts R, Lane S, Hanslik T, Hauser T, Hellmich B, Koldingsnes W, et al. Development and validation of a consensus methodology for the classification of the ANCA-associated vasculitides and polyarteritis nodosa for epidemiological studies. *Ann Rheum Dis*. 2007;66(2):222-7.
2. Peshin R, O'Gradaigh D. Mononeuritis multiplex as a presenting feature of Wegener granulomatosis: a case report. *Clin Rheumatol* 2007;26(8):1389-90.
3. Cattaneo L, Chierici E, Pavone L, Grasselli C, Manganello P, Buzio C et al. Peripheral neuropathy in Wegener's granulomatosis, Churg–Strauss syndrome and microscopic polyangiitis. *J Neurol Neurosurg Psychiatry* 2007;78(10):1119-23.
4. Ananthakrishnan L, Sharma N, Kanne JP. Wegener's granulomatosis in the chest: high-resolution CT findings *AJR Am J Roentgenol*. 2009;192(3):676-82.
5. Stegeman CA, Tervaert JW, Sluiter WJ, Manson WL, de Jong PE, Kallenberg CG. Association of chronic nasal carriage of *Staphylococcus aureus* and higher relapse rates in Wegener granulomatosis. *Ann Intern Med*. 1994;120(1):12-7.
6. Kallenberg CG. What is the evidence for prophylactic antibiotic treatment in patients with systemic vasculitides? *Curr Opin Rheumatol*. 2011;23(3):311-6.
7. Rodrigues CE, Callado MR, Nobre CA, Moura FE, Vieira RM, de Albuquerque LA et al. Wegener's granulomatosis: prevalence of the initial clinical manifestations--report of six cases and review of the literature. *Rev Bras Reumatol*. 2010;50(2):150-64.
8. Holle JU, Gross WL. Neurological involvement in Wegener's granulomatosis. *Curr Opin Rheumatol*. 2011;23(1):7-11.
9. Tejero-Delgado MA, Guerrero AL, Hernández E, Martín-Polo J, Martín-Serradilla JI, Gutiérrez F et al. a Peripheral neuropathy as a first sign of microscopic polyangiitis. *Neurologia*. 2011;26(5):312-4.
10. Mubashir E, Ahmed MM, Hayat S, Latif S, Heldmann M, Berney SM. Wegener Granulomatosis: A Case Report and Update. *South Med J*. 2006;99(9):977-88.

Access full text article on
other devices



Access PDF of article on
other devices

