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## A Fatal Case of Treatment-Related Adverse Effects in Granulomatosis with Polyangiitis (Wegener's Granulomatosis)

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### ABSTRACT

Well-studied therapies have proven to be effective in treating Granulomatosis with polyangiitis (formerly Wegener's granulomatosis) (GPA). There has been considerable improvement in survival of patients with GPA but treatment related morbidity and mortality remains still high, particularly in patients with renal disease. We describe a case of 64-year old woman with recent onset GPA, who responds well to the initial cyclophosphamide based therapy but latter develops a fatal stroke. Infectious complications should be considered in patients with GPA who are on adequate immunosuppression but develop symptoms that may mimic a relapse. Aggressive diagnostic interventions should be undertaken to discriminate between an infection and a relapse of GPA.

### KEYWORDS

Granulomatosis with Polyangiitis, Wegener's Granulomatosis, Cerebrovascular Accident, Infection, CNS Manifestation

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### References

- [1] B. V. Maramattom, C. Giannini, E. M. Manno and E. F. Wijdicks, "Wegener's," 2005. <http://www.ncbi.nlm.nih.gov/pubmed/15980626>  
<http://www.ncbi.nlm.nih.gov/pubmed/15980626>
- [2] H. A. Rula, R. Silverstein, G. Hoffman, L. Zhang, P. Imrey and C. Langford, "The Correlation between Circulating Microparticles and Platelet Aggregation and Disease Activity in Wegener Granulomatosis," *Arthritis and Rheumatism*, Vol. 62, Supplement 10, 2010, p. S856.
- [3] M. DeBandt, V. Ollivier, O. Meyer, C. Babin-Chevaye, F. Khechai, D. de Prost, J. Hakim and C. Pasquier, "Induction of Interleukin-1 and Subsequent Tissue Factor Expression by Anti-Proteinase 3 Antibodies in Human Umbilical Vein Endothelial Cells," *Arthritis Rheum*, Vol. 40, No. 11, 1997, pp. 2030-2038. doi: [10.1002/art.1780401116](https://doi.org/10.1002/art.1780401116)
- [4] P. A. Merkel, G. H. Lo, J. T. Holbrook, et al., "Brief Communication: High Incidence of Venous Thrombotic Events among Patients with Wegener Granulomatosis: The Wegener's Clinical Occurrence of Thrombosis (WeCLOT) Study," *Annals of Internal Medicine*, Vol. 142, No. 8, 2005, pp. 620-626.
- [5] G. S. Hoffman, G. S. Kerr, R. Y. Leavitt, C. W. Hallahan, R. S. Lebovics, W. D. Travis, et al., "Wegener Granulomatosis: An Analysis of 158 Patients," *Annals of Internal Medicine*, Vol. 116, No. 6, 1992, pp. 488-498
- [6] R. Seror, A. Mahr, J. Ramanoelina, C. Pagnoux, P. Cohen and L. Guillevin, "Central Nervous System Involvement in Wegener's Granulomatosis," *Medicine*, Vol. 85, No. 1, 2006, pp. 54-65.

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- [7] H. Nishino, F. A. Rubio and J. E. Parisi, "The Spectrum of Neurologic Involvement in Wegener's Granulomatosis," *Neurology*, Vol. 43, No. 7, 1993, pp. 1334-1337.
- [8] W. Zhang, G. Zhou, Q. Shi, X. Zhang, X. F. Zeng and F. C. Zhang, "Clinical Analysis of Nervous System Involvement in ANCA-Associated Systemic Vasculitides," *Clinical and Experimental Rheumatology*, Vol. 27, Supplement 52, 2009, pp. S65-S69.
- [9] J. U. Holle and W. L. Gross, "Neurological Involvement in Wegener's Granulomatosis," *Current Opinion Rheumatology*, Vol. 23, No. 1, 2011, pp. 7-11. doi:10.1097/BOR.0b013e32834115f9
- [10] J. M. Provenzale and N. B. Allen, "Wegener Granulomatosis: CT and MR Findings," *American Journal of Neuroradiology*, Vol. 17, 1996, pp. 785-792.
- [11] B. A. Cunha, "Nervous System Infections in the Compromised Host: A Diagnostic Approach," *Infectious Disease Clinics of North America Central*, Vol. 15, No. 1, 2001, pp. 567-590.
- [12] J. U. Holle, W. L. Gross, U. Latza, B. Noelle, P. Ambrosch, M. Heller, R. Fertmann and E. Reinhold-Keller, "Improved Outcome of 445 Wegener's Granulomatosis Patients in a German Vasculitis Center over Four Decades," *Arthritis & Rheumatism*, Vol. 63, No. 1, 2010, pp. 257-266.
- [13] R. Luqmani, R. Suppiah, C. J. Edwards, R. Phillip, J. Maskell, D. Culliford, D. Jayne, K. Morishita and N. Arden, "Mortality in Wegener's Granulomatosis: A Bimodal Pattern," *Rheumatology*, Vol. 50, No. 4, 2010, pp. 597-702.
- [14] G. S. Hoffman, Y. Drucker, M. F. Cotch, et al., "Wegener's Granulomatosis: Patient-Reported Effects of Disease on Health, Function, and Income," *Arthritis & Rheumatism*, Vol. 41, No. 12, 1998, pp. 2257-2262. doi:10.1002/1529-0131(199812)41:12<2257::AID-ART22>3.0.CO;2-K
- [15] E. Reinhold-Keller, N. Beuge, U. Latza, et al., "An Interdisciplinary Approach to the Care of Patients with Wegener's Granulomatosis: Long-Term Outcome in 155 Patients," *Arthritis & Rheumatism*, Vol. 43, No. 5, 2000, pp. 1021-1032. doi:10.1002/1529-0131(200005)43:5<1021::AID-ANR10>3.0.CO;2-J
- [16] K. De Groot, L. Harper, D. R. Jayne, L. F. Flores Suarez, G. Gregorinin, W. L. Gross, et al., "Pulse versus Daily Oral Cyclophosphamide for Induction of Remission in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis: A Randomized Trial," *Annals of Internal Medicine*, Vol. 150, 2009, pp. 670-680.
- [17] A. D. Booth, M. K. Almond, A. Burns, et al., "Pan-Thames Renal Research Group. Outcome of ANCA-Associated Renal Vasculitis: A Five-Year Retrospective Study," *American Journal of Kidney Diseases*, Vol. 41, No. 4, 2003, pp. 776-784. doi:10.1016/S0272-6386(03)00025-8
- [18] C. Charlier, C. Henegar, O. Launay, C. Pagnoux, A. Berezné, B. Bienvenu, P. Cohen, L. Mouthon and L. Guillevin, "Risk Factors for Major Infections in Wegener Granulomatosis: Analysis of 113 Patients," *Annals of the Rheumatic Disease*, Vol. 68, No. 5, 2009, pp. 658-663. doi:10.1136/ard.2008.088302
- [19] M. Little, P. Nightingale, C. A. Verburgh, et al., "Early Mortality in Systemic Vasculitis: Relative Contribution of Adverse Events and Active Vasculitis," *Annals of the Rheumatic Disease*, Vol. 69, No. 6, 2010, pp. 1036-1043. doi:10.1136/ard.2009.109389
- [20] A. Ibrahim, B. Spellberg and J. Edwards, "Iron Acquisition: A Novel Perspective on Mucormycosis Pathogenesis and Treatment," *Current Opinion in Infectious Disease*, Vol. 21, No. 6, 2008, pp. 620-625. doi:10.1097/QCO.0b013e3283165fd1
- [21] W.-F. Li, H. E. Chao, X.-F. Liu, S.-Y. Wang, J.-L. Qu and Z.-F. Lin, "A Diagnosis Neglected for 6 Years: Report of a Misdiagnosed Case of Pulmonary Mucormycosis and Review of the Literature," *Chinese Medical Journal*, Vol. 123, No. 17, 2010, pp. 2480-2482
- [22] M. F. Liu, F. F. Chen, T. R. Hsiue and C. C. Liu, "Disseminated Zygomycosis Simulating Cerebrovascular Disease and Pulmonary Alveolar Haemorrhage in a Patient with Systemic Lupus Erythematosus," *Clinical Rheumatology*, Vol. 19, No. 4, 2000, pp. 311-314. doi:10.1007/s100670070052
- [23] E. Nogueira, P. W. Ind, J. Friedland and A. D. Salama, "Mucormycosis May Mimic Disease Relapse in Wegener's Granulomatosis," *The Journal of Rheumatology*, Vol. 37, No. 6, 2010, pp. 1364-1365. doi:10.3899/jrheum.091423
- [24] J. D. Finkelstein, P. A. Merkel, D. Schroeder, et al., "Antiproteinase 3 Antineutrophil Cytoplasmic Antibodies and Disease Activity in Wegener Granulomatosis," *Annals of Internal Medicine*, Vol. 147,

No. 9, 2007, pp. 611-619.

- [25] R. J. Falk and G. S. Hoffman, "Controversies in Small Vessel Vasculitis," *Current Opinion in Rheumatology*, Vol. 19, No. 1, 2007, pp. 1-9.
- [26] H. P. McAdams, M. R. de Christenson, D. C. Stollo and E. F. Patz Jr., "Pulmonary Mucormycosis: Radiologic Findings in 32 Cases," *American Journal of Neuroradiology*, Vol. 168, No. 6, 1997, pp. 1541-1548.
- [27] J. H. Stone, P. A. Merkel, R. Spiera, P. Seo, C. A. Langford, G. S. Hoffman, U. Specks, et al., "RAVE-ITN Research Group," *The New England Journal of Medicine*, Vol. 363, 2010, pp. 221-232. doi: 10.1056/NEJMoa0909905