

Wegener's granulomatosis

Wegener's granulomatosis (WG) is a systemic [vasculitis](#) that can affect a variety of organs including ear, nose and throat, lungs and kidneys. However WG is unusual in the pituitary and rare in the central nervous system.

A 56-year-old male with likely WG presented with polyuria and polydipsia despite six months of conservative medical management. MRI scanning revealed an enlarging heterogeneously enhancing pituitary gland. Following endoscopic transsphenoidal pituitary biopsy and debulking, final tissue pathology was diagnostic for WG in the pituitary gland.

Diagnosis remains difficult but most patients present with central [diabetes insipidus](#) (CDI) as well as varying degrees of [hypopituitarism](#) on a background of disease activity in other organs. Clinical judgment needs to balance the need for invasive surgical tissue diagnosis with increasing immunosuppressive therapy.

It is important to consider this rare complication of WG to ensure timely diagnosis and management ¹⁾

Pending classification

1: Song SY, Lan D, Wu XQ, Ding YC, Ji XM, Meng R. Clinical characteristics, inflammation and coagulation status in patients with immunological disease- related chronic cerebrospinal venous insufficiency. *Ann Transl Med.* 2021 Feb;9(3):236. doi: 10.21037/atm-20-4201. PMID: 33708863; PMCID: PMC7940939.

2: Triano MJ, Haberstroh WD, Lenka A, Whelton SA. Relapsed granulomatosis with polyangiitis with panhypopituitarism. *BMJ Case Rep.* 2021 Jan 28;14(1):e237774. doi: 10.1136/bcr-2020-237774. PMID: 33509867; PMCID: PMC7845712.

3: Mohamed H, Ce Ó Mathúna E, Elbardesy H, Shorten G. Anaesthetic management of a patient with eosinophilic granulomatosis with polyangiitis for internal fixation of a fractured femur. *BMJ Case Rep.* 2021 Jan 8;14(1):e239052. doi: 10.1136/bcr-2020-239052. PMID: 33419755; PMCID: PMC7798413.

4: Inaba R, Fuse Y, Kurimoto F, Suzuki S, Watanabe K. A Rare Case of Eosinophilic Granulomatosis with Polyangiitis Presenting as Ischemic Stroke and Splenic Infarction. *J Stroke Cerebrovasc Dis.* 2021 Mar;30(3):105539. doi: 10.1016/j.jstrokecerebrovasdis.2020.105539. Epub 2020 Dec 14. PMID: 33333478.

5: Munawar K, Nayak G, Fatterpekar GM, Sen C, Zagzag D, Zan E, Hagiwara M. Cavernous sinus lesions. *Clin Imaging.* 2020 Dec;68:71-89. doi: 10.1016/j.clinimag.2020.06.029. Epub 2020 Jun 18. PMID: 32574933.

6: Funayama Y, Watanabe G, Tsukita K, Suzuki H, Uenohara H, Suzuki Y. [MPO-ANCA- associated hypertrophic pachymeningitis with monoclonal gammopathy of undetermined significance: a case report]. *Rinsho Shinkeigaku.* 2020 Jul 31;60(7):500-503. Japanese. doi: 10.5692/clinicalneuro.60.cn-001411. Epub 2020 Jun 13. PMID: 32536662.

7: Celik O, Kalfa M, Celebi ARC, Cabuk B, Ceylan S. Granulomatosis With Polyangiitis as an Uncommon Cause of Panhypopituitarism. *J Clin Rheumatol*. 2019 Dec;25(8):e145-e146. doi: 10.1097/RHU.0000000000000766. PMID: 31764508.

8: Sharifi G, Mohajeri-Tehrani MR, Navabakhsh B, Larijani B, Valeh T. Idiopathic granulomatous hypophysitis presenting with galactorrhea, headache, and nausea in a woman: a case report and review of the literature. *J Med Case Rep*. 2019 Nov 16;13(1):334. doi: 10.1186/s13256-019-2276-4. PMID: 31731892; PMCID: PMC6858767.

9: Mur T, Ghraib M, Khurana JS, Roehm PC. Granulomatosis with Polyangiitis Presenting with Bilateral Hearing Loss and Facial Paresis. *OTO Open*. 2019 Jan 25;3(1):2473974x18818791. doi: 10.1177/2473974x18818791. PMID: 31236531; PMCID: PMC6572924.

10: Abushamat LA, Kerr JM, Lopes MBS, Kleinschmidt-DeMasters BK. Very Unusual Sellar/Suprasellar Region Masses: A Review. *J Neuropathol Exp Neurol*. 2019 May 20;nlz044. doi: 10.1093/jnen/nlz044. Epub ahead of print. PMID: 31233145.

11: Xu D, Xu H, Wang F, Wang G, Wei Q, Lei S, Gao Q, Zhang Q, Guo F. Multiple Distinctive Demyelinating Lesions Caused by Eosinophilic Granulomatosis With Polyangiitis: Case Report and Literature Review. *Front Neurol*. 2019 Mar 11;10:213. doi: 10.3389/fneur.2019.00213. PMID: 30930835; PMCID: PMC6423896.

12: Margoni M, Barbareschi M, Rozzanigo U, Chioffi F, Paolazzi G, Marangoni S. Isolated choroid plexus involvement in a case of granulomatosis with polyangiitis negative for antineutrophil cytoplasmic antibodies (ANCA). *J Neurol Sci*. 2019 Mar 15;398:128-130. doi: 10.1016/j.jns.2019.01.045. Epub 2019 Jan 26. PMID: 30708210.

13: Zhang X, Xing B, You H, Wu H, Zhong Y, Ma J. Severe ophthalmic manifestation in pituitary-involved granulomatosis with polyangiitis: a case report and literature review. *BMC Ophthalmol*. 2018 Nov 16;18(1):299. doi: 10.1186/s12886-018-0966-0. PMID: 30445952; PMCID: PMC6240177.

14: Barbieri FR, Novegno F, Iaquinandi A, Lunardi P. Hypertrophic Pachymeningitis and Hydrocephalus-The Role of Neuroendoscopy: Case Report and Review of the Literature. *World Neurosurg*. 2018 Nov;119:183-188. doi: 10.1016/j.wneu.2018.07.194. Epub 2018 Jul 31. PMID: 30075265.

15: Tsuda Y, Oguri T, Sakurai K, Watanabe T, Maeda N, Yuasa H. [Intrasellar xanthogranuloma with abscess formation in a patient with Rathke's cleft cyst]. *Rinsho Shinkeigaku*. 2018 Jun 27;58(6):411-413. Japanese. doi: 10.5692/clinicalneurology.001163. Epub 2018 Jun 1. PMID: 29863103.

16: Draghi R, Mantovani G, Runza L, Carrabba G, Fusco N, Rampini P, Costa A, Locatelli M. Rathke's cleft cyst associated with pituitary granulomatosis with polyangiitis: An unusual combination of hypothalamus-pituitary region pathologies. *Radiol Case Rep*. 2017 Oct 31;13(1):233-236. doi: 10.1016/j.radcr.2017.09.018. PMID: 29552263; PMCID: PMC5853151.

17: Maekawa K, Shibata M, Seguchi M, Kobayashi K, Naito Y, Miya F. Multiple Cerebral Infarctions Due to Patent Foramen Ovale in a Patient with Eosinophilic Granulomatosis with Polyangiitis. *J Stroke Cerebrovasc Dis*. 2018 Jun;27(6):e92-e94. doi: 10.1016/j.jstrokecerebrovasdis.2017.12.022. Epub 2018 Jan 17. PMID: 29373226.

18: Chen Z, Miao Y, Wu H, Wang R, Zhou Z, Zhang S, Chen L, Qiu Y. Anti- Neutrophil Cytoplasmic Antibody-Negative Central Nervous System Granulomatosis With Polyangiitis and Its Clinical

Characteristics. *J Craniofac Surg*. 2018 Mar;29(2):449-451. doi: 10.1097/SCS.00000000000004145. PMID: 29239918.

19: Itaya S, Ueda Y, Kobayashi Z, Tomimitsu H, Kobayashi D, Shintani S. Bilateral Frontal Lobe Vasogenic Edema Resulting from Hypertrophic Pachymeningitis due to Granulomatosis with Polyangiitis. *Intern Med*. 2017 Dec 15;56(24):3353-3355. doi: 10.2169/internalmedicine.8832-17. Epub 2017 Oct 11. PMID: 29021459; PMCID: PMC5790726.

20: Brasiliense LBC, Dumont TM. Alarming Internal Carotid Artery Aneurysm Eroding the Sphenoid Sinus. *World Neurosurg*. 2017 Dec;108:985.e13-985.e14. doi: 10.1016/j.wneu.2017.09.006. Epub 2017 Sep 30. PMID: 28974410.

21: Christou L, Thomas A, Dutta S, Shaw S, Jose B. Granulomatosis with polyangiitis presenting as a pituitary lesion. *Br J Hosp Med (Lond)*. 2017 Apr 2;78(4):234. doi: 10.12968/hmed.2017.78.4.234. PMID: 28398895.

22: Baird SM, Pratap U, McLean C, Law CP, Maartens N. Rare presentation of Wegener's granulomatosis in the pituitary gland: Case report and literature review. *Int J Surg Case Rep*. 2017;33:24-26. doi: 10.1016/j.ijscr.2017.02.014. Epub 2017 Feb 20. PMID: 28273603; PMCID: PMC5338910.

23: Roy D, Rao PJ, Phan K, Mobbs RJ, Selby M. Spinal cord compression from Wegener's granulomatosis: an unusual presentation. *J Spine Surg*. 2016 Dec;2(4):319-323. doi: 10.21037/jss.2016.12.03. PMID: 28097250; PMCID: PMC5233865.

24: Locatelli M, Di Cristofori A, Draghi R, Bertani G, Guastella C, Pignataro L, Mantovani G, Rampini P, Carrabba G. Is Complex Sphenoidal Sinus Anatomy a Contraindication to a Transsphenoidal Approach for Resection of Sellar Lesions? Case Series and Review of the Literature. *World Neurosurg*. 2017 Apr;100:173-179. doi: 10.1016/j.wneu.2016.12.123. Epub 2017 Jan 5. PMID: 28065874.

25: Harabuchi S, Bando N, Yasukawa R, Kono M, Goto T, Harabuchi Y, Ikeda H, Kamada H, Nishihara H. ANCA-Negative Granulomatosis with Polyangiitis Presenting with Hypertrophic Cranial Pachymeningitis, Abducens Nerve Palsy, and Stenosis of the Internal Carotid Artery. *Case Rep Otolaryngol*. 2017;2017:9687383. doi: 10.1155/2017/9687383. Epub 2017 Nov 30. PMID: 29527371; PMCID: PMC5733126.

26: Bernat AL, Lefevre E, Sène D, Herman P, Biassette HA, Froelich S. Management Scheme for Cerebral Wegener Granulomatosis: An Unusual Pseudotumoral Skull Base Pathology. *World Neurosurg*. 2016 Dec;96:608.e13-608.e16. doi: 10.1016/j.wneu.2016.09.061. Epub 2016 Sep 23. PMID: 27671887.

27: Hisahara S, Yamada M, Matsuura Y, Tsuda E, Akiyama Y, Saitoh M, Kawamata J, Mikuni N, Shimohama S. ANCA-negative granulomatosis with polyangiitis presenting with orbital apex syndrome and recurrent pachymeningitis: A case report. *J Neurol Sci*. 2016 Sep 15;368:175-7. doi: 10.1016/j.jns.2016.07.013. Epub 2016 Jul 9. PMID: 27538627.

28: Decker ML, Emery DJ, Smyth PS, Lu JQ, Lacson A, Yacyshyn E. Microscopic Polyangiitis with Spinal Cord Involvement: A Case Report and Review of the Literature. *J Stroke Cerebrovasc Dis*. 2016 Jul;25(7):1696-1704. doi: 10.1016/j.jstrokecerebrovasdis.2016.01.034. Epub 2016 Apr 8. PMID: 27068775.

29: Alotaibi NM, Ibrahim GM, Jalal H, Perez-Ordóñez B, Hodaie M, Kalia SK. Subdural Collection as Initial Presentation of Granulomatosis With Polyangiitis. *JAMA Neurol*. 2016 May 1;73(5):602-3. doi:

10.1001/jamaneurol.2015.4710. PMID: 26998710.

30: Horovitz Y, Lidar M. [CENTRAL NERVOUS SYSTEM INVOLVEMENT IN GRANULOMATOSIS WITH POLYANGIITIS (GPA)]. Harefuah. 2015 May;154(5):292-5, 340. Hebrew. PMID: 26168637.

31: Brembilla C, Lanterna LA, Risso A, Bombana E, Gritti P, Trezzi R, Bonaldi G, Biroli F. Craniovertebral junction instability as an extension of cocaine- induced midline destructive lesions: case report. J Neurosurg Spine. 2015 Aug;23(2):159-65. doi: 10.3171/2014.11.SPINE1471. Epub 2015 May 8. PMID: 25955800.

32: Mohapatra A, Holekamp TF, Diaz JA, Zebala L, Brasington R. Atlantoaxial subluxation and nasopharyngeal necrosis complicating suspected granulomatosis with polyangiitis. J Clin Rheumatol. 2015 Apr;21(3):156-9. doi: 10.1097/RHU.0000000000000236. PMID: 25807096.

33: de Hoog J, Volovici V, Dammers R. Successful surgical optic nerve decompression in a patient with hypertrophic pachymeningitis due to granulomatous polyangiitis. BMJ Case Rep. 2015 Jan 22;2015:bcr2014208110. doi: 10.1136/bcr-2014-208110. PMID: 25612758; PMCID: PMC4307056.

34: Caporali R, Monti S, Magrassi L, Montecucco C. Pneumocephalus due to granulomatosis with polyangiitis. Rheumatology (Oxford). 2014 Dec;53(12):2269. doi: 10.1093/rheumatology/keu347. Epub 2014 Sep 9. PMID: 25209069.

35: Sierra S, Luquin N, Tardáguila M, Olivé A. Choroid plexitis as a unique neurological manifestation in granulomatosis with polyangiitis (Wegener's disease). J Rheumatol. 2014 Jun;41(6):1192-3. doi: 10.3899/jrheum.131302. PMID: 24882849.

36: Taraschenko OD, Amory CF, Waldman J, Hanspal EK, Bernardini GL. Lateral medullary stroke in patient with granulomatous polyangiitis. J Stroke Cerebrovasc Dis. 2014 May-Jun;23(5):1259-61. doi: 10.1016/j.jstrokecerebrovasdis.2013.09.004. Epub 2013 Oct 12. PMID: 24128976.

37: Carr C, Diehn F, Kaufmann T. Nonenhancing epidural abscess and discitis in an immunocompromised patient: a case report. Clin Neurol Neurosurg. 2013 Oct;115(10):2274-6. doi: 10.1016/j.clineuro.2013.07.018. Epub 2013 Aug 9. PMID: 23937866.

38: Meilak L, Haliasos N, Narducci A, Powell M. More than meets the Wegener's eye. J Rheumatol. 2013 Jul;40(7):1226-7. doi: 10.3899/jrheum.121403. PMID: 23818723.

39: Nicolosi F, Nodari G, Spena G, Roca E, Migliorati K, Esposito G, Stefini R, Fontanella M, Panciani PP. Cerebral Wegener's granuloma: surgery mandatory for diagnosis and treatment. Case Rep Neurol Med. 2013;2013:750391. doi: 10.1155/2013/750391. Epub 2013 May 16. PMID: 23762680; PMCID: PMC3670469.

40: Kalra S, Yadav A, Agarwal S, Kumar S. Wegener's granulomatosis with subdural hematoma as the initial manifestation. Int J Crit Illn Inj Sci. 2013 Jan;3(1):88-90. doi: 10.4103/2229-5151.109430. PMID: 23724392; PMCID: PMC3665127.

41: Pereira EA, Plaha P, Hofer M, Karavitaki N, Cudlip SA. Hypophyseal Wegener's granulomatosis presenting by visual field constriction without hypopituitarism. Clin Neurol Neurosurg. 2013 Jun;115(6):762-4. doi: 10.1016/j.clineuro.2012.06.041. Epub 2012 Aug 13. PMID: 22898089.

42: Onodera H, Hiramoto J, Morishima H, Tanaka Y, Hashimoto T. Treatment of an unruptured fusiform aneurysm of the internal carotid artery associated with Wegener's granulomatosis by endovascular balloon occlusion. Case report. Neurol Med Chir (Tokyo). 2012;52(4):216-8. doi: 10.2176/nmc.52.216.

PMID: 22522334.

43: Rangel-Castilla L, Barber SM, Zhang YJ. Hydrocephalus in Wegener's granulomatosis: neuroendoscopic findings and management. *J Rheumatol*. 2011 Oct;38(10):2277-8. doi: 10.3899/jrheum.110458. PMID: 21965703.

44: Ceri M, Ortobozkoyun L, Unverdi S, Kirac M, Duranay M. Massive intracerebral hemorrhage associated with Wegener granulomatosis. *Rheumatol Int*. 2012 Jun;32(6):1813-4. doi: 10.1007/s00296-011-1954-3. Epub 2011 May 19. PMID: 21594720.

45: Guilfoyle MR, Khan S, Helmy A, Jalloh I, Trivedi S, Trivedi R, Kirkpatrick PJ. Spinal intradural haemorrhage in a patient with Wegener's Granulomatosis. *Clin Neurol Neurosurg*. 2010 May;112(4):341-3. doi: 10.1016/j.clineuro.2009.12.004. Epub 2010 Jan 6. PMID: 20056313.

46: Baaj AA, Vale FL, Carter JD, Rojiani AM. Granulomatosis with CNS involvement: a neuroimaging clinicopathologic correlation. *J Neuroimaging*. 2009 Apr;19(2):194-7. doi: 10.1111/j.1552-6569.2008.00251.x. Epub 2008 Apr 3. PMID: 18393953.

47: Thiryayi W, Donaldson MH, Border D, Tyagi A. An enhancing pituitary lesion in a young woman: a diagnostic dilemma. *J Clin Neurosci*. 2007 Mar;14(3):286-8. doi: 10.1016/j.jocn.2005.12.005. Epub 2006 Dec 22. PMID: 17188492.

48: Kupersmith MJ, Martin V, Heller G, Shah A, Mitnick HJ. Idiopathic hypertrophic pachymeningitis. *Neurology*. 2004 Mar 9;62(5):686-94. doi: 10.1212/01.wnl.0000113748.53023.b7. PMID: 15007115.

49: Yasuhara T, Fukuhara T, Nakagawa M, Terai Y, Yoshino K, Mizobuchi K, Fujimoto S. Wegener granulomatosis manifesting as meningitis. Case report. *J Neurosurg*. 2002 Nov;97(5):1229-32. doi: 10.3171/jns.2002.97.5.1229. PMID: 12450051.

50: McEvoy AW, Khan N, Kitchen ND. Bitemporal hemianopia caused by metastatic carcinoma in a patient with Wegener's granulomatosis: an unexpected finding. *Br J Neurosurg*. 1997 Dec;11(6):579-81. doi: 10.1080/02688699745781. PMID: 11013634.

51: Li KK, Teknos TN, Lai A, Lauretano A, Terrell J, Joseph MP. Extracranial optic nerve decompression: a 10-year review of 92 patients. *J Craniofac Surg*. 1999 Sep;10(5):454-9. PMID: 10726518.

52: Scarrow AM, Segal R, Medsger TA Jr, Wasko MC. Communicating hydrocephalus secondary to diffuse meningeal spread of Wegener's granulomatosis: case report and literature review. *Neurosurgery*. 1998 Dec;43(6):1470-3. doi: 10.1097/00006123-199812000-00126. PMID: 9848863.

53: Yokote H, Terada T, Nakai K, Itakura T. Subdural and meaningful involvement related to Wegener's granulomatosis: case report. *Neurosurgery*. 1997 May;40(5):1071-3; discussion 1073-4. doi: 10.1097/00006123-199705000-00036. PMID: 9149267.

54: Cheng TM, O'Neill BP, Scheithauer BW, Piepgras DG. Chronic meningitis: the role of meningeal or cortical biopsy. *Neurosurgery*. 1994 Apr;34(4):590-5; discussion 596. doi: 10.1227/00006123-199404000-00004. PMID: 8008155.

55: Masago A, Fukuoka H, Yoshida T, Majima K, Tada T, Nagai H. Intracranial mycotic aneurysm caused by *Aspergillus*-case report. *Neurol Med Chir (Tokyo)*. 1992 Nov;32(12):904-7. doi: 10.2176/nmc.32.904. PMID: 1282683.

56: POTOTSCHNIG B. LA SIMPATICECTOMIA GANGLIARE CERVICALE NEL GRANULOMA

GANGRAENESCENS. CRITICA DI UN CASO CLINICO [CERVICAL GANGLIONIC SYMPATHECTOMY IN GRANULOMA GANGRAENESCENS. CRITICAL STUDY OF A CLINICAL CASE]. Otorinolaringol Ital. 1963;32:402-12. Italian. PMID: 14154946.

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Baird SM, Pratap U, McLean C, Law CP, Maartens N. Rare presentation of Wegener's granulomatosis in the pituitary gland: Case report and literature review. Int J Surg Case Rep. 2017 Feb 20;33:24-26. doi: 10.1016/j.ijscr.2017.02.014. [Epub ahead of print] PubMed PMID: 28273603.

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