

Teaching NeuroImage: Calcifying Pseudoneoplasm of the Neuraxis in the Setting of Hereditary Hemorrhagic Telangiectasia and Seizures

Luca H. Debs, MD, Austin Helton, BS, Sami Belakhlef, MD, Suash Sharma, MD, and Scott Y. Rahimi, MD

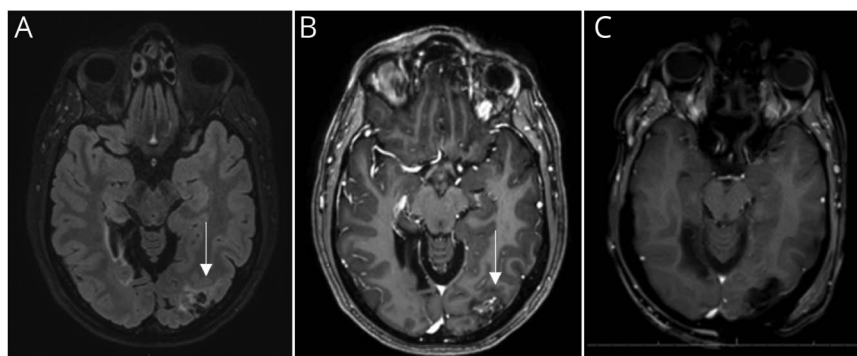
Neurology® 2023;101:e982-e983. doi:10.1212/WNL.0000000000207385

Correspondence

Dr. Debs

lucadebs@gmail.com

Figure 1 MRI of the Brain



Preoperative and postoperative MRI. (A) Preoperative axial T2 fluid-attenuated inversion recovery and (B) preoperative axial T1 with contrast illustrating enhancing lesion (arrow) in the left occipital lobe with surrounding edema. (C) Postoperative axial T1 with contrast demonstrating gross total resection.

We report a 50-year-old man presenting with grand-mal seizures. Workup included MRI of the brain demonstrating a heterogeneously enhancing focus, likely hemorrhagic, in the left occipital region (Figure 1). A vascular etiology was presumed due to the patient's history of Hereditary Hemorrhagic Telangiectasia. Furthermore, digital subtraction angiography revealed an irregular blush with the absence of early venous drainage to suggest an arteriovenous malformation (AVM). On surgical resection, the specimen showed substantial calcifications (Figure 2). Final pathology diagnosed a calcifying pseudoneoplasm of the neuraxis (CAPNON), presumably arising from an AVM remnant.¹ They are rare, slow-growing lesions believed to form secondary to tissue insult. The benefits of this finding over an AVM are 2-fold; cerebral autoregulation is maintained, and future surveillance angiograms are avoidable. CAPNONs have been observed after trauma, infections, neoplasms, and inflammation.² Complete resection was confirmed by intraoperative angiogram and postoperative MRI. The patient had no complications and returned to his seizure-free neurologic baseline.

Author Contributions

L.H. Debs: drafting/revision of the manuscript for content, including medical writing for content; major role in the acquisition of data; study concept or design; analysis or interpretation of data. A. Helton: drafting/revision of the manuscript for content, including medical writing for content; analysis or interpretation of data. S. Belakhlef: drafting/revision of the manuscript for content, including medical writing for content; study concept or design. S. Sharma: major role in the acquisition of data; study concept or design. S.Y.

MORE ONLINE

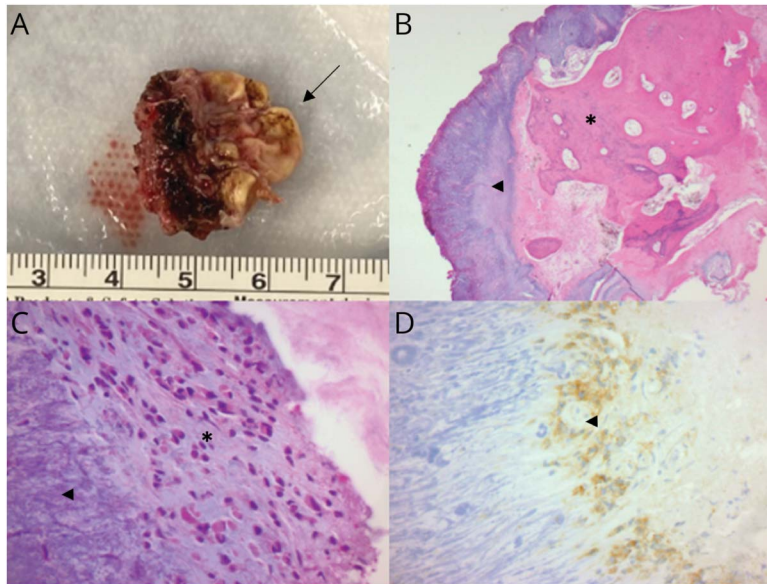
Teaching slides

links.lww.com/WNL/C791

From the Department of Neurosurgery (L.H.D., A.H., S.Y.R.), and Department of Pathology (S.B.), Medical College of Georgia, Augusta; and Department of Pathology (S.S.), College of Medicine, Memphis, TN.

Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Figure 2 Pathology



Gross pathology (A, centimeters) showing calcifications (arrow). Histologic sections demonstrate a nodular lesion with hypocellular fibrillated basophilic material (arrowhead) with adjacent mature bone (*) (B, magnification 2 \times) and peripheral rim of surface spindle and epithelioid cells (*) embedded in a chondromyxoid matrix (arrowhead) (C, magnification 40 \times) with epithelial membrane antigen positivity (arrowhead) (D, magnification 40 \times).

Rahimi: major role in the acquisition of data; study concept or design; analysis or interpretation of data.

Study Funding

No targeted funding reported.

Disclosure

The authors report no relevant disclosures. Go to [Neurology.org/N](https://www.neurology.org/N) for full disclosures.

Publication History

Received by *Neurology* December 12, 2022. Accepted in final form March 21, 2023. Submitted and externally peer reviewed. The handling editor was Resident & Fellow Section Deputy Editor Katherine Fu, MD.

References

1. Florian IA, Popovici L, Timis TL, Florian IS, Berindan-Neagoe I. Intracranial gorgon: surgical case report of a large calcified brain arteriovenous malformation. *Am J Case Rep.* 2020;21:e922872.
2. Pithon RFA, Bahia PRV, Marcondes J, Canedo N, Marchiori E. Calcifying pseudoneoplasm of the neuraxis. *Radiol Bras.* 2019;52(5):342-343.

Neurology[®]

Teaching NeuroImage: Calcifying Pseudoneoplasm of the Neuraxis in the Setting of Hereditary Hemorrhagic Telangiectasia and Seizures

Luca H. Debs, Austin Helton, Sami Belakhlef, et al.

Neurology 2023;101:e982-e983 Published Online before print May 2, 2023

DOI 10.1212/WNL.0000000000207385

This information is current as of May 2, 2023

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/101/9/e982.full
References	This article cites 2 articles, 0 of which you can access for free at: http://n.neurology.org/content/101/9/e982.full#ref-list-1
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): All Cerebrovascular disease/Stroke http://n.neurology.org/cgi/collection/all_cerebrovascular_disease_stroke All Epilepsy/Seizures http://n.neurology.org/cgi/collection/all_epilepsy_seizures Arteriovenous malformation http://n.neurology.org/cgi/collection/arteriovenous_malformation MRI http://n.neurology.org/cgi/collection/mri
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright © 2023 American Academy of Neurology. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

