Published Ahead of Print on May 2, 2023 as 10.1212/WNL.0000000000207385

Neurology®
The most widely read and highly cited peer-reviewed neurology journal
The Official Journal of the American Academy of Neurology

AMERICAN ACADEMY OF NEUROLOGY®

Neurology Publish Ahead of Print
DOI:10.1212/WNL.0000000000207385

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

Author(s):
Luca H Debs, MD[1]; Austin Helton, BS[1]; Sami Belakhlef, MD[2]; Suash Sharma, MD[3]; Scott Y Rahimi, MD[1]

Corresponding Author:
Luca H Debs, lucadebs@gmail.com

Affiliation Information for All Authors: 1. Department of Neurosurgery, Medical College of Georgia, Augusta, GA; 2. Department of Pathology, Medical College of Georgia, Augusta, GA; 3. Department of Pathology, College of Medicine, Memphis, TN.

Equal Author Contribution:

Contributions:
Luca 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
Austin Helton: Drafting/revision of the manuscript for content, including medical writing for content; Analysis or interpretation of data
Sami Belakhlef: Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design
Suash Sharma: Major role in the acquisition of data; Study concept or design
Scott Y Rahimi: Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data

Figure Count:
2

Table Count:
0

Search Terms:
[2] All Cerebrovascular disease/Stroke, [3] Arteriovenous malformation, [60] All Epilepsy/Seizures, [120] MRI

Acknowledgment:

Study Funding:
The authors report no targeted funding

Disclosures:
The authors report no relevant disclosures.

Preprint DOI:

Received Date:
2022-12-12

Accepted Date:
2023-03-21

Handling Editor Statement:
Submitted and externally peer reviewed. The handling editor was Resident & Fellow Section Deputy Editor Katherine Fu, MD.

We report a 50-year-old man presenting with grand-mal seizures. Workup included MRI brain demonstrating a heterogeneously enhancing focus, likely hemorrhagic, in the left occipital region (Figure 1). A vascular etiology was presumed due to patient's history of Hereditary Hemorrhagic Telangiectasia. Further, digital subtraction angiography revealed an irregular blush with absence of early venous drainage to suggest an arteriovenous malformation (AVM). Upon 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 two-fold; cerebral autoregulation is maintained and future surveillance angiograms are avoidable. CAPNONs have been observed following 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 neurological baseline.

WNL-2023-000236_slides --- http://links.lww.com/WNL/C791

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.

Figure 1: Brain MRI
Pre and post-operative magnetic resonance imaging (MRI). (A) Pre-operative axial T2 FLAIR and (B) pre-operative axial T1 with contrast illustrating enhancing lesion (arrow) in the left occipital lobe with surrounding edema. (C) Post-operative axial T1 with contrast demonstrating gross total resection.

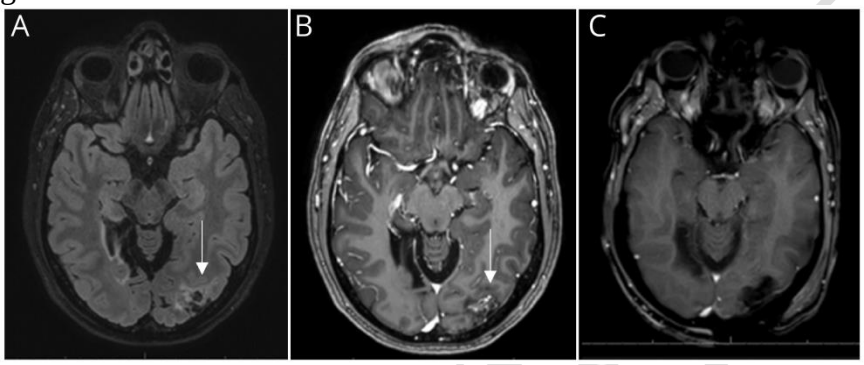
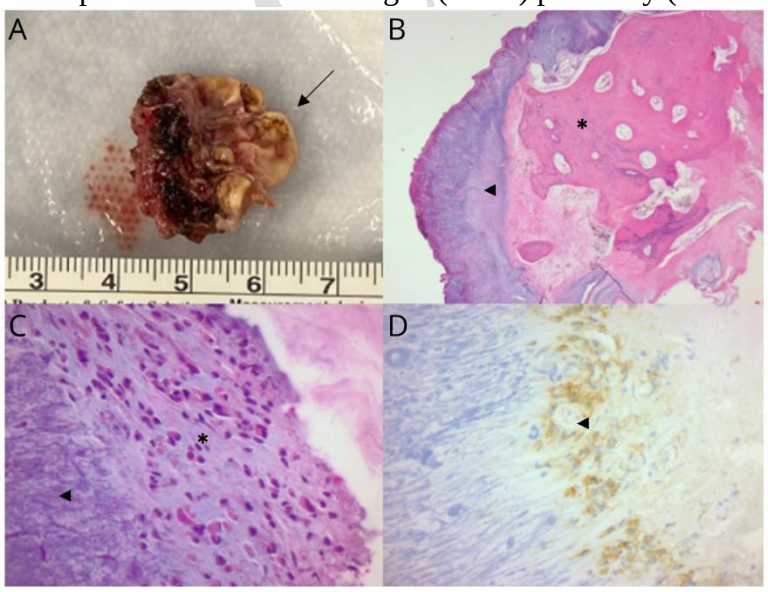


Figure 2: Pathology
Gross pathology (A, centimeters) showing calcifications (arrow). Histologic sections demonstrate a nodular lesion with hypocellular fibrillated basophilic material (arrowhead) with adjacent mature bone (*) (B, magnification 2X) and peripheral rim of surface spindle and epithelioid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



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 published online May 2, 2023
DOI 10.1212/WNL.0000000000207385