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

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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<sup>1</sup>. 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<sup>2</sup>. Complete resection was confirmed by intraoperative angiogram and postoperative MRI. The patient had no complications and returned to his seizure-free neurological baseline.

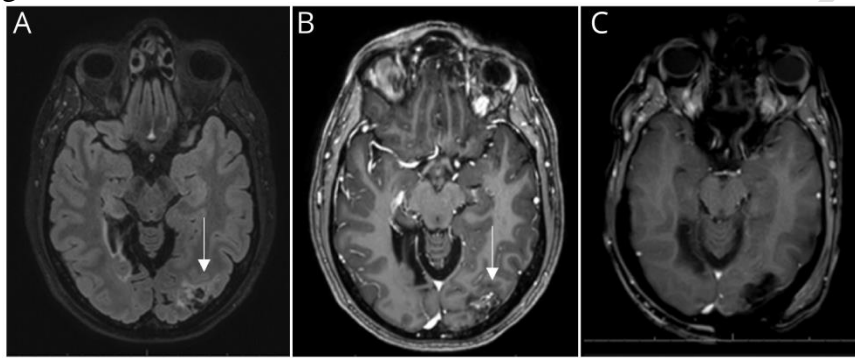
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References:

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.

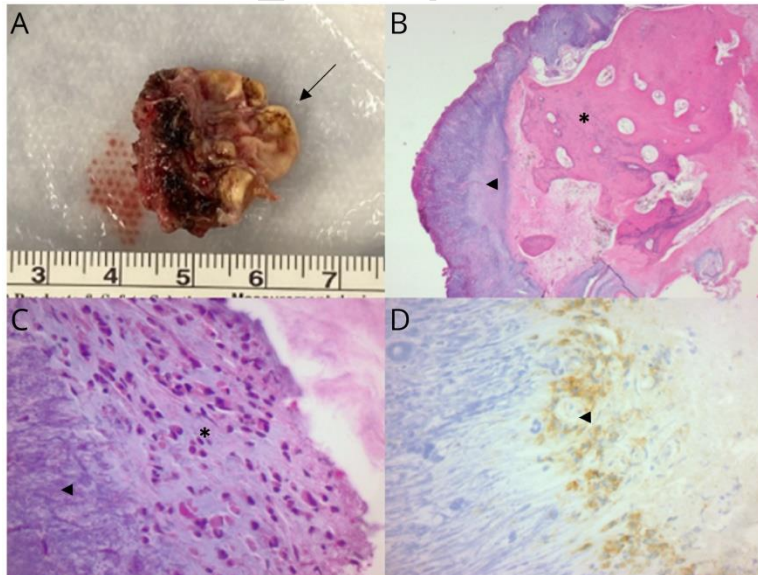
**Figure 1: Brain MRI**

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).



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