

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

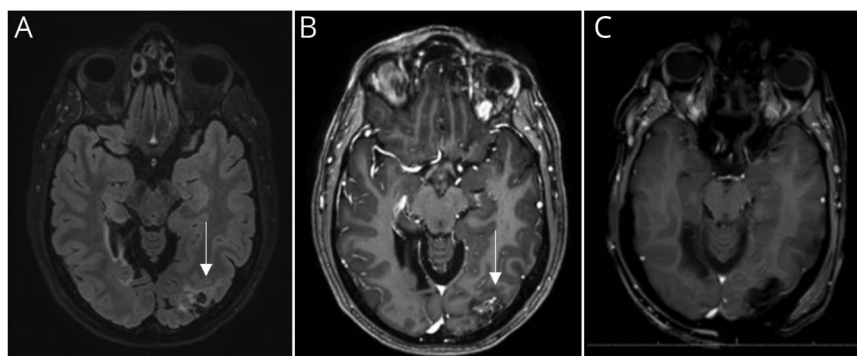
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Figure 1 MRI of the Brain



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.

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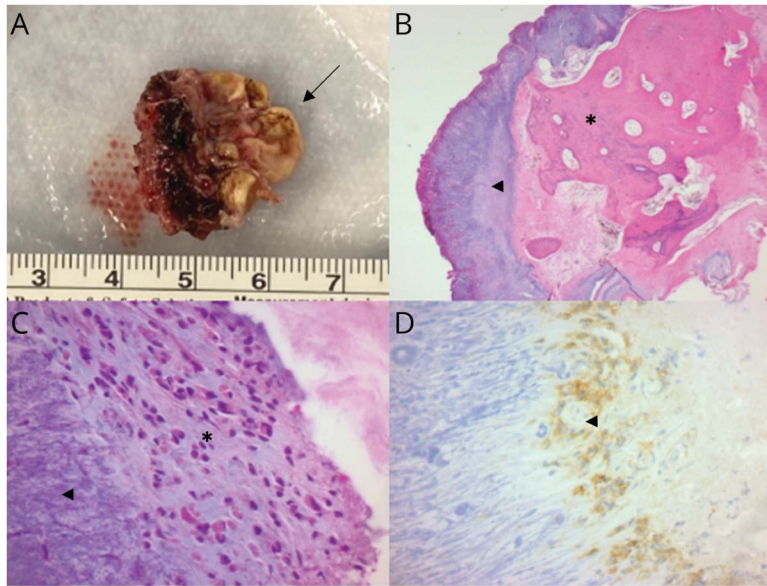
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Figure 2 Pathology



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.

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